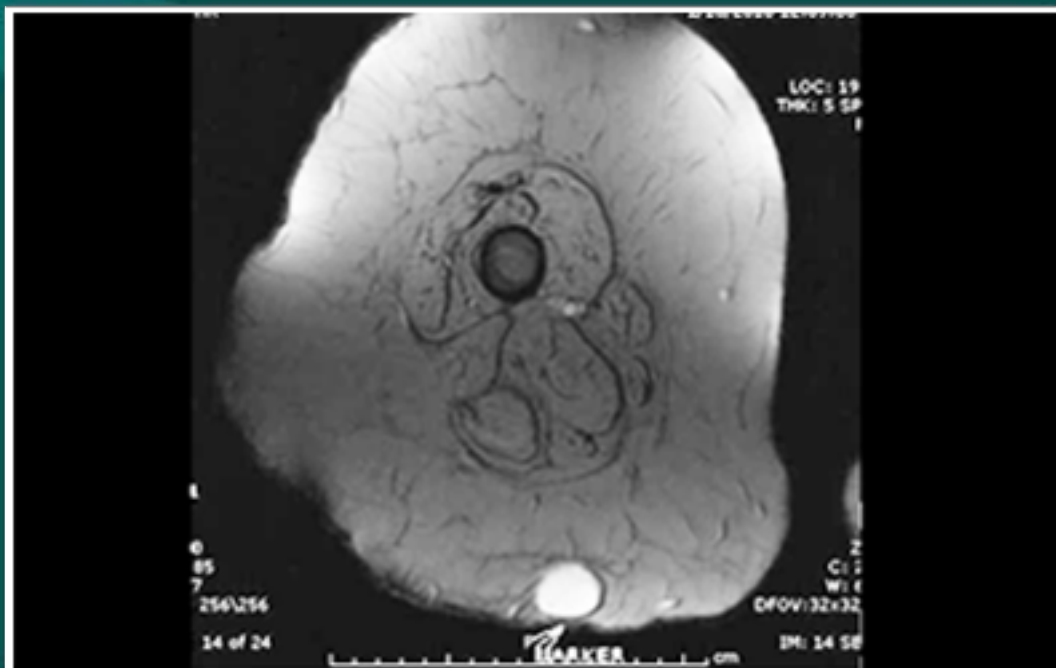


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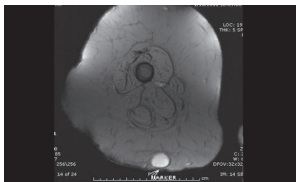
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CASE SERIES

OPEN ACCESS

Granulicatella adiacens isolated from sterile body fluids: A case series from India

Sushma Krishna, Kavitha Dinesh, Deepa Harichandran,
Neeba Jayasurya, Shamsul Karim

ABSTRACT

Introduction: The genera *Abiotrophia* and *Granulicatella* spp. (previously known as nutritionally variant *Streptococcus*) are infrequently isolated from clinical specimens. Literature quotes that they account for about 5–6% of the infective endocarditis and bacteremia, and lesser in central nervous system infections (post instrumentation) and others. The objective of the study was to assess the clinical significance and outcome of the patients with laboratory isolations of *Granulicatella adiacens*. **Case Series:** We reviewed the clinical records from 2011–12 noting down the demographic details, identifiable risk factors, management of patients in whom *Granulicatella adiacens* was isolated. Seven cases of *Granulicatella adiacens* were reported in which five were children (<2 years) and two were male adults. Six strains were from blood and one was isolated from cerebrospinal

fluid shunt fluid, and were regarded as clinically significant. Pre-existing co-morbidities like nephrotic syndrome, premature birth and dysmorphism were noted in almost all the children. One of the patients had undergone invasive ventriculoperitoneal shunt insertion. All the patients except one (discharged against medical advice) recovered. **Conclusion:** The study describes the spectrum of infections by *Granulicatella adiacens*. *G. adiacens* can grow on routine sheep blood agar without pyridoxal supplementation in CO₂ incubator when sub-cultured from automated blood culture bottles. This is one of the largest study from India.

Keywords: *Abiotrophia*, Blood, India, *Granulicatella adiacens*, Shunt infectious, Streptococcus

How to cite this article

Krishna S, Dinesh K, Harichandran D, Jayasurya N, Karim S. *Granulicatella adiacens* isolated from sterile body fluids: A case series from India. Int J Case Rep Images 2014;5(12):802–807.

doi:10.5348/ijcri-201464-CS-10050

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Received: 06 October 2014
Accepted: 17 October 2014
Published: 01 December 2014

INTRODUCTION

Granulicatella species form a part normal oral, genitourinary and intestinal tract flora. Along with the genus *Abiotrophia*, they were originally known as nutritionally variant streptococci (NVS) because of their requirement for pyridoxal as additional agents to be incorporated into standard media for successful laboratory isolation. Three species of *Granulicatella* have now been described viz, *G. adiacens*, *G. elegans* and *G. balaenopterae* [1]. They are uncommon clinical isolates and are implicated in causing invasive infections such as infective endocarditis, bacteremia, and shunt infections [2–4]. Nutritionally

Patient 1: A one-year-old premature baby with duodenal atresia and pelvic pseudocyst was admitted with posthemorrhagic hydrocephalus with ventriculoperitoneal shunt. Shunt infection was suspected and cerebrospinal fluid sent for culture. *G. adiacens*, sensitive to penicillin, erythromycin, cefotaxime, ofloxacin, and resistant to azithromycin (Table 1) was grown in culture and the child was started on vancomycin for one week and rifampicin (one/sixth of 300 mg) for two weeks. Clinical condition improved and shunt was removed later.

Patient 2: A two-year-old girl with nephrotic syndrome on steroids, was admitted with spiking temperatures. Two consecutive blood cultures isolated *G. adiacens*, sensitive to penicillin, erythromycin, cefotaxime, ofloxacin, and resistant to azithromycin. Bacteremia was confirmed and the patient was started on ceftriaxone for 10 days and repeat culture was sterile.

Patient 3: A two-month-old dysmorphic male neonate with global developmental delay, a case of DPT vaccine induced encephalopathy, aspiration pneumonia, failure to thrive, presented with fever of seven days duration. Blood culture set grew pan sensitive *G. adiacens*, Piperacillin-tazobactam was started and on request, child was discharged against medical advice.

Patient 4: A one-year-old female child was admitted with convulsions and fever. Seizure workup was not contributory and a diagnosis of simple febrile seizures was made. While on antiepileptic, blood cultures grew erythromycin and azithromycin resistant *G. adiacens*, she was treated with cefixime for seven days and improved.

Patient 5: A 58-year-old male, a known case of chronic renal failure and multiple myeloma (on thalidomide) with joint effusion and leucopenia was admitted for pyrexia of unknown origin. Two out of six blood cultures received grew *G. adiacens*, erythromycin and azithromycin resistant. With the characteristic mitral valve vegetation on echo, a diagnosis of infective endocarditis was made. He was treated with IV penicillin for forty days and gentamicin for two weeks. Repeat blood cultures on follow-up were negative.

Patient 6: A 43-year-old male presented with severe joint pains and fever. He was a known case of type 2 diabetes mellitus, hypertension and dyslipidemia. Dengue serology (IgM) was positive. Blood cultures grew *G. adiacens*, which was erythromycin and azithromycin resistant. The patient was started on ceftriaxone for seven days with platelet transfusion after which he improved. Repeat blood cultures were negative.

Patient 7: A one-year-old boy with nephrotic syndrome (on steroids), presented with high-grade fever and wheeze from three days. Two blood cultures grew *G. adiacens*, resistant to penicillin, erythromycin and azithromycin resistant. A diagnosis of lower respiratory tract infection was made, was treated with cefotaxime for seven days and the boy improved.

DISCUSSION

Identification of nutritionally variant streptococci is difficult at the laboratory bench. Gram stain may show pleomorphism and morphology depends upon the conditions of growth. They appear in chains including cocci, coccobacilli in chains and occasionally rod-shaped cells when it is grown in cysteine- or pyridoxal-supplemented broth. Some tendency towards rod formation may be observed in the stationary phase which may lead to a misidentification of gram-positive bacilli group (like *Lactobacillus*, Diphtheroids, etc.). Small ovoid cocci occur singly, in pairs or in chains of variable length in CDMT semi-synthetic medium. On culture, they are generally fail to grow on routine culture media. However, the recent automated culture bottles have pyridoxal supplementation in the required concentration (0.001%) which is specifically required for the growth of nutritionally variant streptococci. All the isolates in our study grew well on SBA with alpha-hemolysis in CO₂ incubator without further additional pyridoxal supplementation by 48 hours. The colonies of *G. adiacens* are alpha-hemolytic or non-hemolytic (gamma hemolytic) on SBA [6] and needs to be differentiated from other phenotypically related look alike catalase-negative gram-positive cocci such as Enterococci, Lactococci, Leuconostoc, Vagococcus, Weissella, etc. by biochemical tests, some of which are not routinely available and needs commercial kit systems (like API Rapid Step or Vitek) to identify them. All isolates turned out to be clinically significant and patients were treated with culture sensitive antibiotics and recovered, except one (discharged against medical advice). Five of the study patients with primary diagnosis of other diseases had episodes of bacteraemia and striking pre-disposing factors and recovered with the prompt antibiotic therapy.

About 5–15% of patients with endocarditis have negative blood cultures; in one-third to half of these cases, cultures are negative because of prior antibiotic exposure. The remainder of these is due to fastidious organisms, such as nutritionally variant organisms, HACEK organisms, and *Bartonella* species. *Granulicatella* spp. is known to cause sepsis, bacteremia and infective endocarditis in 5% of cases. The NVS endocarditis has been considered to have a high relapse rate and relapses following treatment have been reported for *Granulicatella* endocarditis and have to be treated in the same way as enterococcal endocarditis. The patient five of infective endocarditis with typical vegetations had no episodes of relapse and was believed to be cured with penicillin and gentamicin for a six-week duration. The need for routine antimicrobial susceptibility testing is not clear as majority of the isolates remain sensitive to penicillin. However, occasional reports of beta-lactam (as in patient seven in the series) and macrolide resistance (most of the isolates in the series were) have been reported where they pose a challenge to treat invasive

infections limiting the available choice [8] and hence, testing should be done even if it is by non-standardized disk diffusion method.

G. adiacens also has been documented to cause central nervous system infections like meningitis, epidural abscess in association with prior neurosurgical procedures including craniotomy, ventriculoperitoneal shunt placement, CT-guided myelography and tumor resection [9]. Patient 1 had a prior shunt placement antecedent to the cerebrospinal fluid isolation. Besides the above, isolation of NVS as likely pathogens has been reported in a diverse list of infections that can be caused by other streptococci, including peritonitis, prosthetic joint infections, breast implant infections and osteomyelitis [10, 11]. In this series, *G. adiacens* was not isolated from any samples other than blood and cerebrospinal fluid. Reporting of more such cases throws light on the clinical spectrum and provides insight about the pathogenesis of these rare listed organisms as definite pathogens, which in turn will allow better and adequate antibiotic therapy for the treatment of invasive infections. Awareness of NVS and willingness to look for them, more so in apparently negative cultures, may unveil them as potential pathogens in other infections too. The report highlights the large series of *G. adiacens* isolations from India conveying that the isolate cannot be disregarded as insignificant commensal and is worth alerting the physician to rule out possible bacteremia, infective endocarditis and shunt infections.

CONCLUSION

Study adds on to the spectrum of infections by *Granulicatella adiacens* from India. *G. adiacens* can grow on sheep blood agar without pyridoxal supplementation in a CO₂ incubator when sub-cultured from automated blood culture bottles containing pyridoxal HCl. We suggest that *Granulicatella* (and *Abiotrophia*) species should be considered in patients where slow-growing α-hemolytic or non-hemolytic streptococci are isolated from blood cultures or other sterile sites in device-associated, in immunocompromised and in the infective endocarditis patients.

Author Contributions

Sushma Krishna – Substantial contributions to conception and design, Drafting the article, Final approval of the version to be published

Kavitha Dinesh – Acquisition of data, Revising it critically for important intellectual content, Final approval of the version to be published

Deepa Harichandran – Substantial contributions to conception and design, Acquisition of data, Revising it critically for important intellectual content, Final approval of the version to be published

Neeba Jayasurya – Acquisition of data, Revising it critically for important intellectual content, Final approval of the version to be published

Shamsul Karim – Acquisition of data, Revising it critically for important intellectual content, Final approval of the version to be published

Guarantor

The corresponding author is the guarantor of submission.

Conflict of Interest

Authors declare no conflict of interest.

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Article citation: Krishna S, Dinesh K, Harichandran D, Jayasurya N, Karim S. *Granulicatella adiacens* isolated from sterile body fluids: A case series from India. Int J Case Rep Images 2014;5(12):802–807.



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CASE SERIES

OPEN ACCESS

Isolated polyostotic fibrous dysplasia of the spine: A diagnostic challenge

Virinder Mohan, Krishnan K. Unni, Nimisha Batra, Ajit Ambekar

ABSTRACT

Introduction: Fibrous dysplasia is a non-malignant fibro-osseous disease of the bone can occur in monostotic or polyostotic form. The polyostotic form is more common and involves pelvis, femur, tibia, ribs, calvarium and the facial bones. Spinal involvement in fibrous dysplasia is extremely uncommon both in monostotic as well as in polyostotic variety. Isolated involvement of multiple vertebrae with normal rest of the skeleton is extremely rare and only one such case has been reported earlier [1].
Case Series: In this study, we are reporting two cases of isolated fibrous dysplasia of the spine with multiple vertebral involvement and without any clinical, endocrinal as well as radiological

evidence of the disease in the axial skeleton. The patients presented in 2nd –4th decade, both with complaints of low back pain. The radiological, clinical and biochemical evaluation have been discussed in details. The diagnosis of the disease was made in both cases on the basis of clinico-radiological workup and confirmed by histopathology. **Conclusion:** Fibrous dysplasia is not commonly seen to occur in spinal column. When seen, vertebral lesions show almost the same features on conventional radiographs as seen in appendicular skeleton. Patients usually present with minor clinical symptoms which are disproportionate to the imaging findings. A high index of suspicion on the conventional radiography with proper clinical workup will help in the diagnosis.

Keywords: Fibrous dysplasia, Fibrous dysplasia of spine, Isolated spinal fibrous dysplasia

How to cite this article

Mohan V, Unni KK, Batra N, Ambekar A. Isolated polyostotic fibrous dysplasia of the spine: A diagnostic challenge. Int J Case Rep Images 2014;5(12):808–812.

doi:10.5348/ijcri-201465-CS-10051

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Received: 18 September 2014
Accepted: 16 October 2014
Published: 01 December 2014

INTRODUCTION

Fibrous dysplasia is an interesting non-malignant fibro-osseous disease of the bone accounting for about 2.5% of all the bony lesions. Polyostotic form of the disease occurs more frequently than the monostotic form and usually involves bones of the pelvis, femur, tibia, ribs, calvarium and the facial bones. Vertebral involvement in both types is extremely uncommon [1–5].

In a typical case of polyostotic disease involving multiple bones of the axial skeleton with or without clinical stigmata of the disease, the presence of disease in a single or multiple vertebrae may not pose a diagnostic problem. However, selective involvement of single or multiple vertebrae with normal rest of the skeleton and without any cutaneous stigmata or endocrinal involvement, the diagnosis becomes a real challenge.

CASE SERIES

Case 1: A 41-year-old male, a serving army soldier, was X-rayed for low back pain of nearly four years duration, at a peripheral medical unit of the Royal Army of Oman, and the radiographs were sent to the senior author for his opinion. The anteroposterior and lateral radiographs of the dorsolumbar and sacral spine (Figure 1A–B) revealed different degree of collapse of multiple lower dorsal, lumbar and upper sacral vertebrae with evidence of expansion, complete effacement of the trabecular pattern and compromise of the spinal canal at multiple levels.

Possibilities of multiple myeloma, metastatic malignancy and hyperparathyroidism were entertained on the basis of above radiographic findings and the patient was asked to report to the senior author for full clinico-radiological and biochemical workup.

Clinical examination revealed a healthy young soldier in sound health. General physical examination was unremarkable. Local examination revealed mild tenderness in the lumbosacral region. Spinal movements were restricted but were pain free. Skeletal survey including chest skiagram did not reveal any positive finding. Ultrasound examination of the abdomen was normal. All the laboratory investigations were reported normal.

On enquiry, the patient produced an old set of Lumbar spine radiographs taken three years back for the same complaints, which revealed almost the same radiographic findings (Figure 2).

Considering the clinical status and young age of the patient, the radiological findings of multiple vertebral collapse without any history of trauma, effacement of the trabecular pattern with evidence of bony expansion and ground glass attenuation with no significant change in the radiographic findings during three years interval and with no evidence of compressive myelopathy in spite of the advanced spinal changes, a firm clinico-radiological diagnosis of a benign pathology was made with fibrous dysplasia as the possible diagnosis. A nuclear bone scan revealed high uptake of the tracer in the involved vertebrae (Figure 3). Computed tomography (CT) scan of the lumbosacral spine revealed expansile destructive lesions of multiple vertebrae (Figure 4) but these findings did not help in confirming or refuting the diagnosis of fibrous dysplasia.

Considering the advanced radiographic findings in multiple vertebrae and possibility of impending

paraplegia, the patient was referred to UK for confirmation of the diagnosis and for prophylactic surgical fusion, to prevent compressive myelopathy.

The patient had a repeat CT scan, MRI scan (Figure 5) and bone scan followed by one close and one open biopsy. However, although the slides were reviewed by multiple pathologists, no definitive conclusions could be made and the possibilities of Paget's disease and fibrous dysplasia were entertained with majority favoring Paget's disease. No surgical intervention was undertaken and the patient was called for review after six months.

Meantime, histopathological slides were obtained from the London hospital and were sent to Mayo's clinic, along with full clinico-radiological workup. The slides were reviewed by the second author (K.K.Unni) along with his team of skeletal pathologists, who confirmed the diagnosis of polyostotic fibrous dysplasia. Three years follow-up did not reveal any progress of the disease.

Case 2: A 20-year-old female was seen at another hospital for low back pain of two years duration and had X-rays of lumbosacral spine which were reported as metastatic malignancy. She was referred to this department. For CT scan of chest and abdomen to find out the possible source of primary malignancy.

Review of the radiographs revealed variable degrees of collapse of multiple dorsolumbar vertebrae with evidence of expansion and complete effacement of the trabecular pattern (Figure 6). The bony changes were almost similar as seen in the X-rays of the Case 1. Clinically, the patient was in good health with no neurological deficit inspite of the advanced radiological findings. On the basis of good general condition of the patient, young age and sex of the patient and advanced spinal changes without any compressive myelopathy and our experience with the first case, a confident clinico-radiological diagnosis of fibrous dysplasia of the spine was made. Skeletal survey revealed normal skull, pelvic and normal limb bones. However, the chest radiograph revealed, an expansile lesion of



Figure 1: (A) Anteroposterior, and (B) Lateral radiographs of dorsolumbar and sacral spine showing collapse of multiple lower dorsal, lumbar and sacral vertebral bodies with expansion, complete effacement of the trabecular pattern and spinal canal compromise at multiple levels.



Figure 2: X-ray dorsolumbar spine lateral view taken three years back showing the same radiographic features without much change.

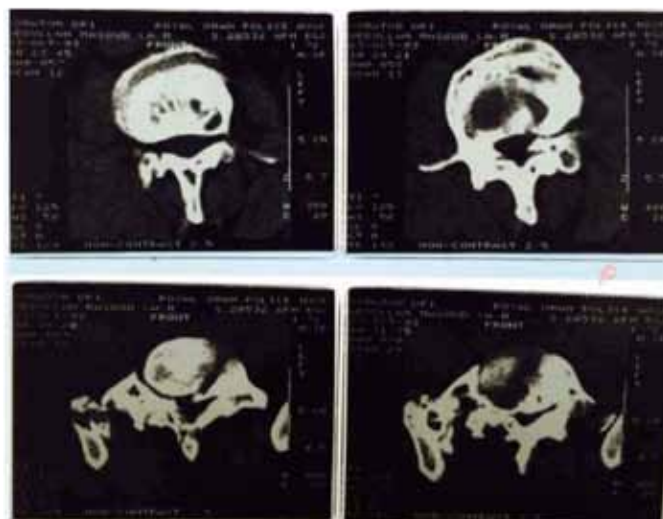


Figure 4: Computed tomography scan of lumbosacral spine showing expansile destructive lesion of multiple lumbar vertebrae.



Figure 3: Nuclear bone scan show increased uptake of the radiotracer by the involved vertebral bodies and the sacrum. Rest of the bones appear normal.

right sided sixth rib with ground glass trabecular pattern, typical of fibrous dysplasia (Figure 7).

The clinico-radiological diagnosis was confirmed by biopsy from the rib. The patient was advised prophylactic orthodesis of the spine, which was refused. She is receiving symptomatic treatment and is doing well.



Figure 5: Magnetic resonance imaging T2-weighted image sagittal section of lumbar spine showing collapse of multiple vertebrae with altered signal intensity in both lumbar and sacral vertebral bodies. There is also narrowing of the spinal canal at multiple lower lumbar levels.



Figure 6: Lateral radiograph of dorsolumbar spine showing changes similar to that of the first case. There is collapse of multiple vertebral bodies with expansion and complete effacement of the trabecular pattern.



Figure 7: Chest skiagram anteroposterior view showing an expansile lesion of the sixth rib on the right side, with ground glass trabecular pattern.

DISCUSSION

Fibrous dysplasia of bone is a developmental disorder characterized by fibrous replacement of the normal medullary bone with poorly organized spicules of immature bone in a fibrous connective tissue. The disorder was first characterized by Lichenstein in 1938 and was subsequently found to have monostotic and polyostotic varieties, the latter of which may be coupled with cutaneous and or endocrinal abnormalities [6].

Fibrous dysplasia affecting the vertebrae is very unusual [1, 3, 4, 7, 8]. Dahlin and Unni found only two cases of vertebral involvement out of 418 cases reviewed [3]. Even in the cases with polyostotic disease showing typical lesions in multiple bones of the skeleton, one may come across only an odd case either on skeletal survey or on nuclear bone scan, and the diagnosis of vertebral fibrous dysplasia in these cases may not be difficult in the light of typical bony findings elsewhere in the skeleton. In the monostotic form, the diagnosis is seldom possible on Imaging and is always histopathological [7, 9, 10]. The diagnosis of fibrous dysplasia was really a challenge in our cases as the disease was involving multiple vertebrae with normal appendicular skeleton.

Fortunately in both of our cases, we could make the diagnosis on the basis of clinico-radiological findings, although it took lot of time and efforts in getting the diagnosis confirmed histopathologically in the Case 1. Only one similar case showing multiple vertebral involvement with normal appendicular skeleton has been found reported in scanned literature [1].

There is no predilection for any part of the spinal column for fibrous dysplasia, though sacral and coccygeal involvement is distinctly rare. Lumbar spine has been found to be involved in majority of case [4, 11] followed by cervical and thoracic spine [5, 7, 9, 10, 12].

CONCLUSION

No pathognomic radiologic findings have been described in spinal fibrous dysplasia. The vertebral lesions in fibrous dysplasia show almost the same features on conventional radiographs as seen in the appendicular skeleton, including expansion of the bone with thinning of the cortex and ground glass trabecular pattern. The lesions may present with minor symptoms and the clinical signs and symptoms are disproportionate to the imaging findings which are quite advanced. Spinal cord compression is quite rare [8]. The discrepancy in the clinical and imaging findings is quite helpful in arriving at a clinico-radiological diagnosis. A high index of suspicion on the plain X-rays, with proper clinical workup as in our cases will help in the diagnosis less short of biopsy.

Acknowledgements

We are grateful to the Director, Armed Forces Medical Services, Royal Army of Oman Ministry of Defence, Sultanate of Oman for all the help rendered in getting the diagnosis conformed in Case 1.

Author Contributions

Virinder Mohan – Substantial contribution to concept and design, Acquisition of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published

Krishnan K. Unni – Substantial contribution to concept and design, Acquisition of data, Revising it critically for important intellectual content, Final approval of the version to be published

Nimisha Batra – Substantial contribution to concept and design, Acquisition of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published

Ajit Ambekar – Substantial contribution to concept and design, Acquisition of data, Revising it critically for important intellectual content, Final approval of the version to be published

Guarantor

The corresponding author is the guarantor of submission.

Conflict of Interest

Authors declare no conflict of interest.

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CASE REPORT

OPEN ACCESS

Human metapneumovirus treated with inhaled ribavirin: A case report

Monica Khunger, Ellen F. Eaton, Craig Hoesley

ABSTRACT

Introduction: Human metapneumovirus is a new pulmonary pathogen in the Paramyxoviridae family. It is increasingly associated with respiratory illnesses particularly in immunocompromised patients. We report a case of a multiple myeloma patient diagnosed with human metapneumovirus pneumonia and successfully treated with inhaled ribavirin. **Case Report:** A 56-year-old white female previously diagnosed with multiple myeloma presented with fever, non-productive cough, dyspnea and new onset ground glass opacities on computed tomography scan. Human metapneumovirus was detected in bronchoalveolar lavage specimen with direct fluorescent antibody testing. Marked clinical improvement was observed following a five-day course of inhaled ribavirin. **Conclusion:** Human metapneumovirus can cause community acquired pneumonia in immunocompromised patients presenting with lower respiratory tract illness. It is associated with significant morbidity and mortality in this population. Early diagnosis and prompt treatment with inhaled ribavirin may improve outcomes in immunocompromised patients.

Keywords: Bone marrow transplant, Human metapneumovirus, Immunocompromised, Ribavirin

How to cite this article

Khunger M, Eaton EF, Hoesley C. Human metapneumovirus treated with inhaled ribavirin: A case report. Int J Case Rep Images 2014;5(12):813–817.

doi:10.5348/ijcri-2014139-CR-10450

INTRODUCTION

Human metapneumovirus (hMPV) was first identified in 2001 from 28 nasopharyngeal isolates from Dutch children with upper respiratory infections symptom. Its electron micrographic structure, biochemical properties and cytopathic effects position hMPV in the Paramyxoviridae family [1]. No similar virus was identified in 400 children without respiratory symptoms suggesting its role in symptomatic upper respiratory tract infection (URI). Reverse transcriptase PCR (RT-PCR) and direct fluorescent antibody (DFA-Abs) testing proved this was a novel virus. Its RNA did not amplify with primers associated with other members of Paramyxoviridae family. In addition, anti-sera of animals exposed to hMPV did not react with viral particles of known members of Paramyxoviridae family. A year after its discovery, Boivin et al. studied 11 nasopharyngeal and bronchoalveolar lavage (BAL) specimens and isolated hMPV for the first time in adults [2]. This study also demonstrated the first evidence of re-infection when the investigators found a single child had two different hMPV strains in two different winter seasons.

The clinical manifestations relate to hMPV-infected dendritic cells (DCs) releasing soluble matrix proteins from virus. These matrix protein bind activated DCs

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Received: 31 July 2014
Accepted: 18 August 2014
Published: 01 December 2014

inducing their maturation and production of cytokines and subsequent activation of T lymphocytes. In addition, hMPV causes hyperplasia of the respiratory epithelium resulting in airway obstruction and hyper responsiveness to methacholine challenge. This suggests the possibility of asthma exacerbation with hMPV infection similar to other respiratory tract viral infections [3]. The clinical manifestations of hMPV infection in adults and children are identical. The infection usually presents with cough, nasal congestion, rhinorrhea, dyspnea, hoarseness and wheezing. Immunocompromised hosts present similarly but may also have fever [1].

There are two major modalities for the diagnosis of hMPV: Reverse transcription (RT-PCR) and (Direct fluorescent antibody assay). Though RT-PCR is the most sensitive method of diagnosis, it has limitations including the possibility of false positive results in asymptomatic carriers. The other method for diagnosis is antibody assay which detects hMPV antigens by immunofluorescence on nasopharyngeal or bronchoalveolar lavage specimen. This assay uses a blend of three fluorescein labeled murine monoclonal antibodies (MAbs). Though not successful in detecting new antigenic variants, this test has a very high specificity and can be performed rapidly [1].

Supportive therapy is the main stay of treatment for hMPV. To date, there are no FDA-approved treatment options for individuals with hMPV infection. Hamelin et al. compared intraperitoneal ribavirin and glucocorticoids in hMPV infected mice and found ribavirin significantly decreased both hMPV replication in lungs and pulmonary inflammation on postinfection day five whereas glucocorticoids only reduced alveolar and interstitial inflammation [4].

There is little data on successful treatment of human metapneumovirus with ribavirin in the immunocompromised patients, including multiple myeloma patients. We report a case of human metapneumovirus pneumonia with successful symptom resolution after a five-day course of inhaled ribavirin.

CASE REPORT

A 56-year-old white female with multiple myeloma presented to outpatient clinic with low grade fever, non-productive cough and chest congestion. She also had progressive dyspnea on exertion with minimal exertion. She denied chest pain, dysuria, orthopnea, paroxysmal nocturnal dyspnea, lower extremity edema, rash and oral lesions. She had no recent travel or sick contacts. She had a past medical history of lambda light chain multiple myeloma complicated by acute kidney injury, diffuse bone involvement and plasmacytoma of the right iliac crest one year prior to arrival. This was treated with successful autologous bone marrow transplant one year ago, and since then she had been managed on prednisone 20 mg/day. She received levofloxacin at an outside hospital

before presentation with slight improvement in her fever curve but not complete resolution. Her temperature was 100.6°F, heart rate was 120 beats per minute, blood pressure was 110/70 mmHg, and oxygen saturation was 93% on 2L nasal cannula. On examination she was in no acute respiratory distress. Fine bibasilar crackles were heard, and her examination was otherwise unremarkable. Her laboratory were significant for absolute neutrophil count of 143. The hemoglobin levels, platelet count, renal and liver function tests were within normal limits. Conventional radiographic findings (Figure 1) and subsequent computed tomography (CT) scan (Figure 2) on admission and hospital day-2, respectively, revealed bilateral patchy lung infiltrates, right lower lobe atelectasis with discrete ground-glass opacification. On admission blood and sputum samples were obtained for routine microscopy and culture, including bacterial, fungal and mycobacterial cultures, and were unrevealing. Nasal washings were also negative for respiratory syncytial virus (RSV), influenza, para-influenza and fungal assay were all negative. She underwent a bronchoscopy with bronchoalveolar lavage (BAL). The BAL fluid was also cultured for bacteria, fungi and mycobacteria and all were negative. Additional diagnostic testing of the BAL fluid for *Pneumocystis jirovecii* and other respiratory viruses (influenza A and B, RSV, parainfluenza virus, adenovirus, and enterovirus) remained negative but hMPV type B was detected using direct fluorescent antibody testing.

The patient remained symptomatic and febrile even on broadspectrum anti-bacterial agents. Here initial antibiotic regimen included cefepime 2 g IV BID, vancomycin 1 g IV q18 hours, trimethoprim/sulfamethoxazole 25 mL IV q8 hours and azithromycin 250 mg IV daily. Based on the results of BAL fluid testing a five-day course of inhaled ribavirin, 2 g q8 hours, was initiated. Her neutropenia resolved with administration of filgrastim at a dose of 5 µg/kg/day. With ribavirin treatment, her shortness of breath and respiratory distress gradually improved, and her oxygen was weaned off. She also showed marked improvement in her physical examination within 48 hours. As a result she was discharged home within three days of completion of her inhaled ribavirin treatment. A computed tomography scan of thorax performed two months after her discharge showed complete resolution of ground glass opacities (Figure 3). Incidentally, she was noted to have elevated lambda light chain and beta-2 microglobulin levels, with a low free lambda/kappa light chain concerning for multiple myeloma recurrence. Bone marrow biopsy confirmed a relapse of her multiple myeloma. This, in addition to her acute illness, was a probable reason for her neutropenia at presentation. The patient developed further complications of recurrent multiple myeloma. She presented four months later with acute renal failure and cord compression related to spinal metastases. When she became uremic and obtunded, her family decided to pursue palliative care measures and withdrew aggressive resuscitation. She died five months after metapneumovirus diagnosis.

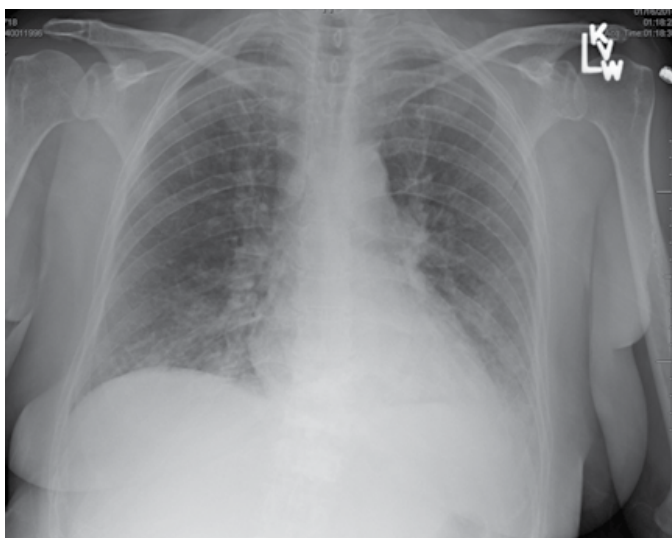


Figure 1: Conventional radiographic imaging on admission revealed bilateral patchy lung infiltrates, right lower lobe atelectasis with discrete ground-glass opacification.



Figure 2: Computed tomography imaging on hospital day-2, revealed bilateral patchy lung infiltrates, right lower lobe atelectasis with discrete ground-glass opacification.



Figure 3: Computed tomography scan of chest performed two months after treatment with inhaled ribavirin showing resolution of ground glass opacities.

DISCUSSION

The development of fever with pulmonary infiltrates is a frequent life-threatening complication in immunocompromised patients, requiring early diagnosis and treatment. Our patient's acute respiratory illness was consistent with hMPV infection based on her clinical presentation, hMPV direct fluorescent antibody testing result, and the absence of alternate explanation for her symptoms. She had an indolent but progressive respiratory illness consistent with hMPV. Her history of multiple myeloma and corticosteroid treatment resulted in immunosuppression and likely contributed to her prolonged course. Respiratory infections due to hMPV, like RSV are more severe in immunocompromised patients [1]. CT scan also showed bilateral ground glass opacities not present on CT scan performed five months prior to presentation consistent with a viral etiology.

An hMPV associated respiratory disease occurs in all age groups. It is found in 3.4% of adult population with respiratory tract infection [1, 5]. The hMPV infections may be more severe and the course more prolonged in immunocompromised patients due to poor clearance of virus.

In a study by William et al., 251 episodes of respiratory tract infections were studied in patients with hematological malignancies [6]. Twenty-two (9%) of these episodes were found to be associated with hMPV infection. Of these, 16 occurred in hematopoietic cell transplant recipients and three of nine patients with lower respiratory tract disease died.

Ribavirin is a synthetic guanosine nucleoside analog with in vitro activity against hMPV [7]. Its mechanism of action involves inhibition of viral RNA polymerase. Data on ribavirin treatment of hMPV in immunocompromised hosts are limited. Englund et al. detected hMPV in bronchoalveolar lavage specimens from 5 of 163 (3.0%) HSCTs patients [8]. Four of the five patients died with acute respiratory failure. The one patient treated with inhaled ribavirin had a fatal outcome. Another case report by Kamble et al. reported successful treatment in a hematopoietic stem cell transplant patient with a four-day course of intravenous ribavirin and immunoglobulins [9]. In another case series by Shahda et al., nine immunocompromised patients with hMPV pneumonia were studied—two were successfully treated with oral and aerosolized ribavirin along with intravenous immunoglobulins (IVIg) [10]. In another case series, Egli et al., described two multiple myeloma patients with hMPV pneumonia [11]. Of these two, one was treated with IVIg and oral ribavirin while the other was treated with IVIg alone.

We report the first case of successful outcome and improved respiratory distress with inhaled ribavirin alone without oral antiviral therapy or IVIg in a multiple myeloma patient. In the absence of data on hMPV treatment outcomes, experience with inhaled ribavirin in immunocompromised RSV patients led us to select inhaled

rather than oral or IV administration [12]. Likewise, there are no studies of optimal ribavirin dosing in human metapneumovirus pneumonia. However, randomized controlled multicenter trials of inhaled ribavirin in RSV infection suggested the use of 6 g/24 h of aerosolized ribavirin at a rate of 2 g/8h, the dose selected for our patient [13]. We believe the early recognition of hMPV pneumonia and timely therapy with inhaled ribavirin contributed to the successful outcome. Inhaled ribavirin is an expensive and cumbersome treatment modality but may be appropriate in heavily immunocompromised population like current patient. However, multi-center case series are needed to shed light on the long-term safety and efficacy of inhaled ribavirin. From the time our patient was diagnosed and treated new research has suggested that oral ribavirin may be an option for patients with RSV and moderate to severe immunosuppression, a finding that may hold promise in hMPV [14].

CONCLUSION

The goals of this case report are:

- (1) to alert clinicians to the possibility of hMPV as a cause of severe community acquired pneumonia in immunocompromised hosts
- (2) to highlight the importance of early suspicion of viral infection and rapid diagnostic testing
- (3) to suggest the use of inhaled ribavirin as a possible treatment modality. Prompt administration of inhaled ribavirin should be considered in immunocompromised patient population with established hMPV infection.

Author Contributions

Monica Khunger – Data acquisition, Analysis and interpretation of data, Drafting of the manuscript, Final approval of the version to be published

Ellen F. Eaton – Data acquisition, Analysis and interpretation of data, Drafting of the manuscript; Final approval of the version to be published

Craig Hoesley – Case report concept and design, Analysis and interpretation of data, Critical revision of the manuscript for important intellectual content, Administrative, technical, or material support; case report supervision, Final approval of the version to be published

Guarantor

The corresponding author is the guarantor of submission.

Conflict of Interest

Authors declare no conflict of interest.

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CASE REPORT

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A rare case of glomus tumor of the thigh: Malignant or not?

Chukwuemeka Ezeoke, Dong Xiang, Nishant Poddar

ABSTRACT

Introduction: This is the case of a patient with a symptomatic painful mass on right thigh. The patient was found to have a very rare case of glomus tumor of the thigh, which has been seen in only 2% of cases since this tumor was first discovered in 1924. A glomus tumor should be considered in the differential diagnosis of a painful, benign mass in a limb. **Case Report:** A 48-year-old African-American morbidly obese female presented with a tender mass on the back of her right thigh. The mass progressively increased in size for three months prior to presentation. She denied having a mass under the nail bed, hands, palms, or any breast lumps. Her yearly mammograms were negative. Magnetic resonance imaging (MRI) scan showed a well-circumscribed mass measuring 3.1x2.2 cm. She was referred to a vascular surgeon for excisional biopsy, which revealed a glomus tumor of uncertain malignant potential measuring 2.7 cm extending to the inked surgical margin. She underwent re-excision with no evidence of residual mass. **Conclusion:** Glomus tumor of the thigh is very rare. Pain, tenderness, and cold

hypersensitivity should raise the suspicion for glomus tumor.

Keywords: Glomus tumor of thigh, Glomus tumor with uncertain malignant potential, Glomus tumor

How to cite this article

Ezeoke C, Xiang D, Poddar N. A rare case of glomus tumor of the thigh: Malignant or not? Int J Case Rep Images 2014;5(12):818–821.

doi:10.5348/ijcri-2014140-CR-10451

INTRODUCTION

This is a case of a 48-year-old female who initially presented with a painful progressive enlarging mass on the back of her right thigh. The mass progressively increased in size with more discomfort at rest. She denied any pruritus, fever, erythema or other systemic symptoms. She was found to have a rare case of glomus tumor of the thigh, which has been seen in 2% of cases since this tumor was discovered in 1924. Often, missed at presentation, a glomus tumor should be considered in any painful, benign mass in a limb. The likelihood of the tumor progressing to malignancy is identified by a number of factors: size >2 cm, atypia, increased mitotic activity >2 per 50/hpf, high nuclear grade, and depth of involvement in relation to muscle fascia or visceral location [1].

CASE REPORT

A 48-year-old African-American female with a history significant for asthma, hypertension, and osteoarthritis, initially presented with a painful mass on the posterior right thigh. The mass progressively increased in size

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Received: 17 June 2014

Accepted: 10 July 2014

Published: 01 December 2014

causing the patient to seek medical evaluation. Patient denied ever having a mass under the nail bed, hands, palms, or any breast lumps on yearly mammogram.

On presentation, her vital signs were normal. Physical examination was remarkable for an immobile, non-indurated, non-erythematous, 1x1.5 cm mass on the right posterior mid-thigh region with tenderness to palpation, and without any palpable lymph nodes. No abnormalities were seen on laboratory studies. Chest X-ray and electrocardiogram were without abnormalities.

Magnetic resonance imaging (MRI) scan of the right thigh which showed a well-circumscribed mass measuring 3.1x2.2 cm (Figures 1 and 2). She underwent resection of the painful mass with immediate relief of pain. Excisional biopsy grossly showed a 2.7x2.5x1.0 cm tumor surrounded by subcutaneous adipose tissue. It was well circumscribed and partially surrounded by a thin fibrotic capsule. Serial sectioning of the tumor showed a yellowish-tan, lobulated surface. There was no necrosis or hemorrhage identified grossly. The tumor appeared to be completely excised with grossly negative margins. Under low magnification, the tumor appeared to be well circumscribed and surrounded by a fibrous capsule (Figure 3). The lesion contained numerous irregular dilated vascular channels lined by bland single layer endothelial cells. Nests of neoplastic epithelioid cells surrounded the vascular channels. Tumor cells were present at the inked margin.

Under high magnification, epithelioid cells were mostly uniform in size with small round nuclei and inconspicuous nucleoli. The tumor cells contained moderate amount of eosinophilic cytoplasm. Mitotic rate was low (less than 2 per 50 high power fields). There was no cytological atypia or atypical mitosis identified (Figure 4). Immunohistochemical stains demonstrated that the tumor cells were positive for smooth muscle antibody (SMA), caldesmon, Desmin and CD34 and negative for S100 and AE1/AE3 (Figures 5–6). Ki-67 showed a mitotic index less than 1%. Five months later, she underwent re-excision as the inked margin of initial excisional biopsy was positive for tumor cells. She has been disease free with no clinical evidence of recurrence.

DISCUSSION

Glomus tumor is a well-circumscribed mass composed of vessels surrounded by epithelioid cells with uniform, round nuclei. It is very rare and comprises 2% of soft tissue tumors [2, 3]. The glomus body, a thermoregulator, is an arteriovenous anastomosis localized in dermal, precoccygeal soft tissue and in areas of the skin that are rich in glomus bodies (for example, the subungual regions of digits or the deep dermis of the palm, wrist, forearm, and foot) [2, 4].

Glomus tumor is a rare, benign neoplasm composed of cells resembling smooth muscle cells of the normal glomus body. Its clinical differential diagnosis includes:

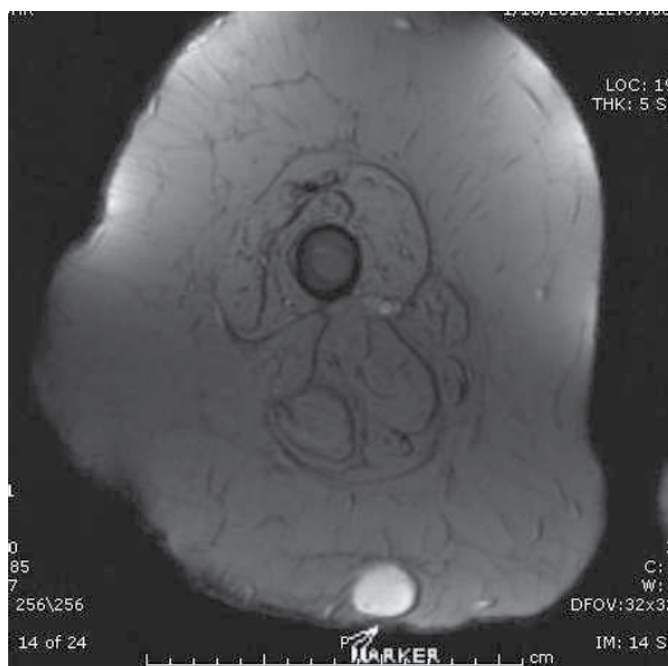


Figure 1: MRI right femur: GRE Axial view of the glomus tumor.

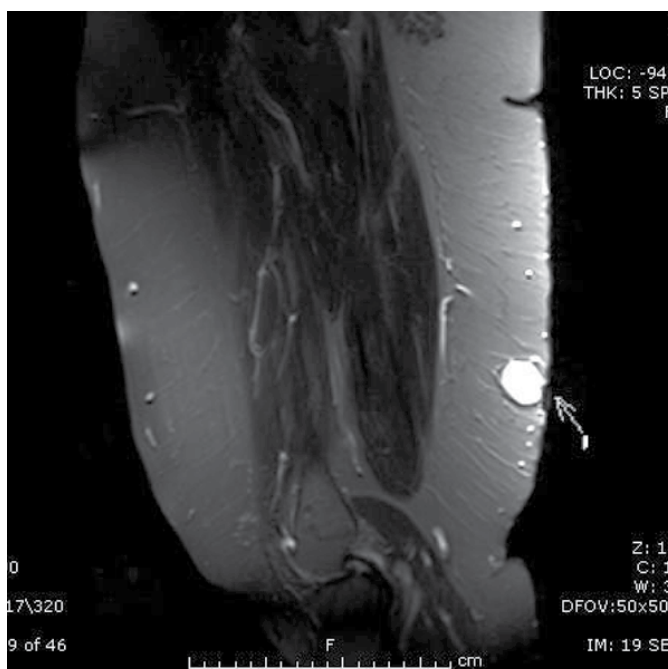


Figure 2: MRI right femur: STIR Sagittal view of the glomus tumor.

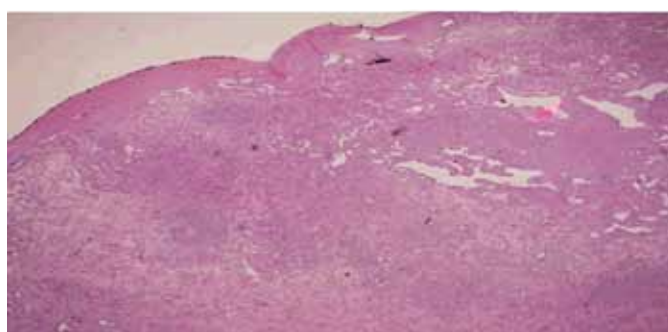


Figure 3: A low power view showing that the tumor is well circumscribed with a fibrotic capsule.

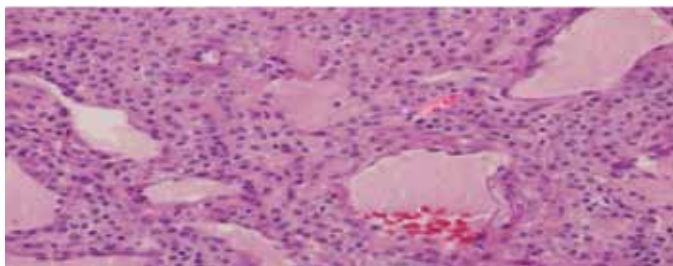


Figure 4: A high power view showing the tumor contains numerous irregular vascular channels surrounded by nests of epithelioid cells with uniform nuclei, inconspicuous nucleoli, and eosinophilic cytoplasm.

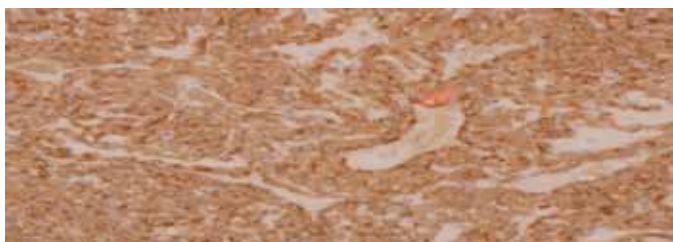


Figure 5: Immunostain showing the tumor cells are diffusely positive for alpha smooth muscle actin.

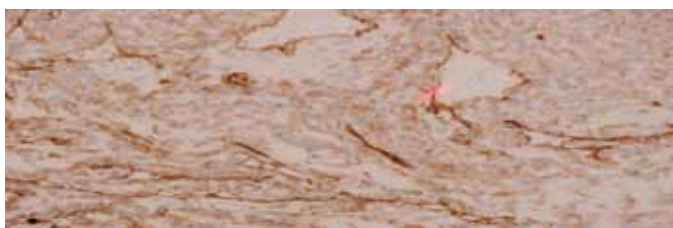


Figure 6: Immunostain showing the tumor cells are positive for CD34.

abscess, epidermal inclusion cysts, hemangioma, arteriovenous malformations. Glomus tumor may be observed at any age. In most instances, it occurs in the fourth or fifth decades of life. 73% of glomus tumor occurs in the upper extremities; 23% in the lower extremities, of which only 2% occurs in the thigh [2, 4]. Presentation is classically a triad of pain, pinpoint tenderness, and hypersensitivity to cold. Localized pain and tenderness can be detected in 86% of these patients, but cold intolerance occurs in less than 2% [5, 6]. Vascular resistance through muscle contractions because of their intramuscular proximity decreases blood flow to the tumor. In essence, glomus tumors originate from neuro-myoarterial bodies; therefore, temperature changes will affect vascular resistance because of its thermoregulatory properties via skin blood flow, and cold simulations.

Ultrasound is useful in diagnosing cases in the outpatient clinic. MRI scan is used for accurate guidance and evaluation of tumor for excisional biopsy. Biopsy confirms the diagnosis. Histologically, glomus tumor is a well-circumscribed dermal nodule composed of glomus cells, vasculature, and smooth muscle cells. Solid glomus tumor, with scarce vasculature and scant muscle component, is the most common variant. Less common

variants include glomangioma, with prominent vascular component, and glomangiomyoma, with prominent vascular and smooth muscle components [1, 2, 3].

Glomus tumors are classified as: malignant with a 30–100% risk of metastasis, symplastic, glomangiomas, and tumor of uncertain malignant potential. Tumor of uncertain malignant potential is defined per College of American Pathologists as follows: superficial location and high mitotic activity or size >2 cm only, or deep location only. Our patient's tumor is classified as tumor of uncertain malignant potential because it is >2 cm, but superficial, with low mitotic activity [1, 2]. No case of glomus tumor of uncertain malignant potential has been reported to become metastatic or malignant after excision despite 7–10 years of follow-up; although one case did require re-excision [2]. Therefore, its benign nature is still uncertain. Close follow-up is advised. The most definitive treatment so far with the limited data available is surgical excision [2]. The risk of progression to malignancy is unknown, however, more cases need to be reported for better understanding of the disease and developing effective treatment to prevent recurrence.

CONCLUSION

Glomus tumor of the thigh is very rare. Pain, tenderness, and cold hypersensitivity should raise suspicion for glomus tumor.

Author Contributions

Chukwuemeka Ezeoke – Substantial contributions to conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article, Revising it critically for important intellectual content; Final approval of the version to be published.

Dong Xiang – Substantial contributions to conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published

Nishant Poddar – Substantial contributions to conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published.

Guarantor

The corresponding author is the guarantor of submission.

Conflict of Interest

Authors declare no conflict of interest.

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CASE REPORT

OPEN ACCESS

Generalized chloromas with multiple cranial nerves palsies in a patient with chronic myeloid leukemia in a tertiary institution in South-south Nigeria: A case report

Mabel Ino-Ekanem, Timothy Amos Ekwere

ABSTRACT

Introduction: Chloromas or granulocytic sarcomas are extramedullary granulocytic tumors which occur in acute myeloid leukemia (AML), myeloproliferative disorders and myelodysplastic syndromes. They are composed of myeloid blast and herald the onset of systemic relapse. **Case Report:** We report a rare case of 49-year-old male with generalized chloromas in chronic myeloid leukemia (CML) associated with multiple cranial nerves deficit suggesting an intracranial involvement. The central nervous system (CNS) is considered to be an uncommon site for chloromas. Treatment with hydroxyurea was unremarkable as the chloroma became more widespread and neurological deficit worsen. This study brings to fore this rare presentation and the apparent limitation of hydroxyurea (cytoreductive therapy) in patient management. **Conclusion:** Widespread florid chloromas presenting in chronic phase of CML with associated CNS manifestation is rare. The response to Initial therapy with hydroxyurea was unremarkable. The patient was discharged against medical advice and thus lost to follow-up.

Keywords: Chloromas (myeloid sarcoma), Chronic myeloid leukemia, Hydroxyurea, Granulocytic sarcoma

How to cite this article

Ino-Ekanem, M, Ekwere TA. Generalized chloromas with multiple cranial nerves palsies in a patient with chronic myeloid leukemia in a tertiary institution in South-south Nigeria: A case report. Int J Case Rep Images 2014;5(12):822–825.

doi:10.5348/ijcri-2014141-CR-10452

INTRODUCTION

Chronic myeloid leukemia (CML) is a malignant tumor of the pluripotential hematopoietic stem cell. It is characterized by marked increase in granulocytes and more than 95% of cases are associated with the presence of Philadelphia chromosomes [1]. Chloromas are tumors composed of immature granulocytic cells and reported in 2–5% of cases affected with CML [1]. They are often localized, very rarely generalized and are regarded as an early sign of systemic relapse or blastic transformation [2].

CASE REPORT

A 49-year-old male artisan referred from a private hospital with two months history of left sided abdominal pain, progressive weakness and easy fatigability. He had a persistently low hematocrit ranging from 0.18–0.20 L/L despite repeated transfusion with three units of blood in the private hospital. His physical examination findings were as follows; middle aged man afebrile (temperature 36.8°C), moderately pale, anicteric, not cyanosed, not dehydrated, no significant peripheral lymphadenopathy, no pedal edema, no purpura or ecchymotic lesions. His

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Received: 08 September 2014

Accepted: 13 October 2014

Published: 01 December 2014

abdomen was asymmetrically distended, moved with respiration. The liver was not palpably enlarged and had a span of 12 cm. The spleen was enlarged and measured 17 cm below the left costal margin, it was firm and smooth with a blunt edge. The kidneys were not ballotable and there was no ascites. Rectal examination was normal; other systems were essentially normal. Hematological investigations of the patient are hematocrit 0.20 L/L, total white blood cell count $135.5 \times 10^9/L$ and platelet count $171 \times 10^9/L$.

Peripheral blood film (PBF) examination showed a complete spectrum of myeloid cells at different stages of differentiation and basophilia- WBC differential: myeloblasts 02%, promyelocytes 05%, myelocytes 22%, metamyelocyte 07%, band forms 20%, neutrophils 33%, eosinophils 03%, basophils 05%, and lymphocyte 03%. Platelets appeared normal on the film (Figure 1A). Bone marrow aspiration (BMA) cytology showed hyperactive myelopoiesis with complete spectrum of myeloid series with peaks at the myelocytes stage of differentiation. Myelogram was as follows: Myeloblast 05%, promyelocytes 6%, myelocytes 24% metamyelocytes 6%, band forms 20%, neutrophils 30%, eosinophils 04%, Basophils 03%, and lymphocytes 02% (Figure 1A). The PBF and BMA examinations were in keeping with chronic myeloid leukemia (CML) in chronic phase. Therefore, patient was counsel on the course of the disease.

Also, 10 mL of EDTA anticoagulated blood sample was used for BCR-ABL transcript quantization. BCR-ABL 1 major (e14a2) transcript type was detected using multiplex PCR method. ABL quantization was 1.20×10^4 copies per micro liter of cDNA, BCR-ABL quantity was 6490 copies per micro liter of cDNA using Real time PCR. BCR-ABL ratio was 54.083%.

The patient was given appropriate counseling on the disease with assistance from Social and Counseling unit of the hospital. Supportive treatments including; optimization of hematocrit, hydration and allopurinol to prevent tumor lysis syndrome were instituted prior to commencement of hydroxyurea at a dose of 1 g daily. On day-3 of admission, patient complained of difficulty in swallowing, hoarseness of voice, slurred speech and facial deviation to the left. There was no headache or limb weakness. He was reviewed by the neurologist who made an impression of multiple cranial nerve palsies secondary to infiltration of cranial nerves V, VI, VII, IX and XII. On day-6 of admission a repeat hemogram was done which showed a hematocrit of 0.29L/L, white blood cell count $191.7 \times 10^9/L$ and platelet count of $415 \times 10^9/L$. Myelogram were: Myeloblast 18%, Promyelocyte 10%, Myelocyte 20%, metamyelocyte 15%, Band form 10%, Neutrophils 15%, Lymphocyte 03%, Eosinophils 02% and Basophils 07% (Figure 1B). Also, multiple subcutaneous nodules involving the scalp, face, trunk and extremities were noticed (Figure 2). The following differential diagnoses were considered; Non-Hodgkin's lymphomas of the lymphoblastic type, large-cell lymphoma and small round cell tumors.

Excision biopsy of a subcutaneous nodule was done. The histologic sections showed diffuse infiltration of fibro connective tissue by sheets of rounds to oval immature cells (blasts) with scant cytoplasm, vesicular to hyperchromatic nuclei and frequent mitotic figures. In many foci, there was also numerous tingible body macrophages scattered within the lesion (Figure 3A–B). Based on this a histological diagnosis of granulocytic sarcoma (chloroma) was made. The dose of hydroxyurea was increased to 2 g daily, however, response both clinical and hematological were minimal. The chloromas became more widespread and the neurological deficits worsen. The patient insisted on being discharged, declining further treatment and referral to another tertiary health facility in South-West Nigeria for further treatment with Imatinib. This chemotherapeutic agent is given free to patients through the support of an international donor agency in this centre. Since then, the patient was lost to follow-up.

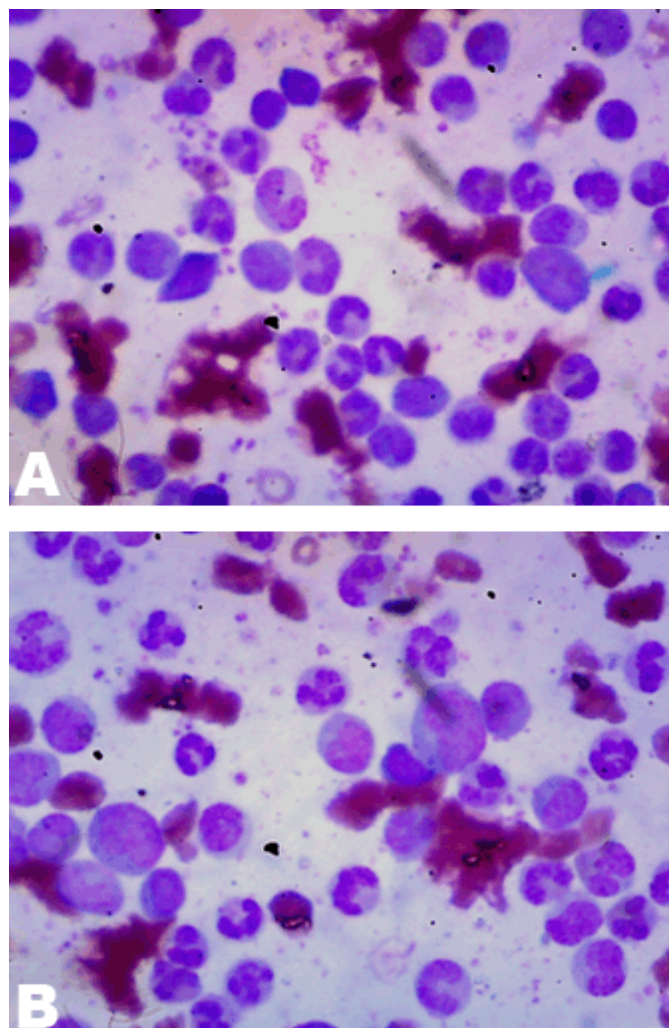


Figure 1: (A) Peripheral blood film of subject, showing complete spectrum of the myeloid cells at different stages of maturation (Leishman stain, x100). (B) Peripheral blood film showing increasing number of blasts (Leishman stain, x100).



Figure 2: Generalized chloromas on different parts of the body of the index patients.

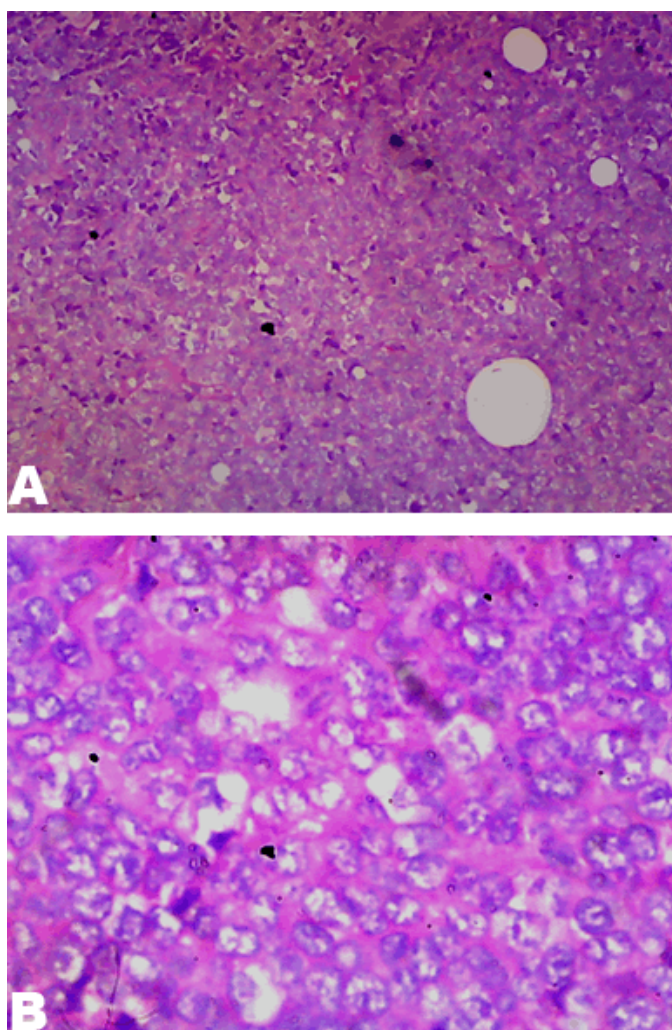


Figure 3: (A & B) Photomicrographs of fibro-connective tissue showed chloroma consisting of round-to-oval immature tumor cells (blasts) with vesicular to hyperchromatic nuclei and scanty cytoplasm admixture with numerous tangible body macrophages (H&E stain, x100, x400 respectively).

DISCUSSION

Granulocytic sarcomas or chloromas are rare extramedullary tumors [3], they are localized tumor

masses usually arising de novo or associated with other hematologic disorders such as acute myeloid leukemia, myeloproliferative disorders or myelodysplastic syndromes [3].

The incidence of chloromas in patients with CML ranges from 2–4% [3–5]. They can present in various organs or tissues. The commonest sites of involvement in CML patients are bone (57%), lymph nodes (29%) and skin or soft tissue (21%) [6]. Involvement of the central nervous system is rare [7]. Both solitary [6] and multiple lesions have been reported [8]. Commonly, chloromas presents in the late stage of the disease or during a relapse and its occurrence is associated with a poor disease outcome [5, 6].

In our index case, the patient developed chloromas while on hydroxyurea, a cytoreductive chemotherapy. Also, the associated neurological deficit involving some cranial nerves may suggest intracranial deposits of these malignant myeloid cells. Intracranial chloromas are seen more commonly in AML or following a relapse case of AML. In CML, it is associated with the onset of blastic crisis [2]. Smidt et al., reported a case of intracranial chloroma which was initially missed-diagnosed as chronic subdural hematoma in a 45 years male with relapsing AML [9]. The diagnosis was, however, made radiologically using gadolinium enhanced magnetic resonance imaging (MRI) scan of the brain and confirmed by biopsy and histology examination of the biopsied tissue. This imaging technique is able to determine the identity of an intracranial mass, particularly when differentiating between intracranial chloromas and hematoma in patients with hematologic malignancies [9]. However, the intracranial involvement in this patient could not be diagnosed radiologically because of financial constraint as the patient was unable to afford the cost of MRI scan of the brain. Healthcare cost is largely out of pocket payment borne entirely by patients in our environment.

Currently, no therapeutic strategies including; chemotherapy, radiotherapy and/or hemopoietic stem cell transplantation have been considered best in the management of this condition [10]. The initial therapy with hydroxyurea used in this case was apparently ineffective as the chloroma persisted and the neurological deficit worsened. Perhaps early introduction of imatinib may have slowed down the progression of the disease. The agent was not available to us and also the patient could not afford it. Hence, the need to refer him to another health facility where he could access the drugs free, but patient declined. Unfortunately, the final outcome of the patient is unknown as he requested to be discharged against medical advice.

CONCLUSION

Widespread florid chloromas presenting in chronic phase of chronic myeloid leukemia (CML) with associated central nervous system manifestation is rare. The patient

response to hydroxyurea was unremarkable. However, the final outcome of this case is unknown as patient was discharged against medical advice and thus lost to follow-up.

Author Contributions

Mabel Ino-Ekanem – Substantial contributions to conception and design, Acquisition of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published
Timothy Amos Ekwere – Substantial contributions to conception and design, Acquisition of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published

Guarantor

The corresponding author is the guarantor of submission.

Conflict of Interest

Authors declare no conflict of interest.

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CASE REPORT

OPEN ACCESS

A case of pediatric paraparesis secondary to an idiopathic acute transverse myelitis

Joana Teixeira, Susana Carvalho, Sofia Martins, Teresa Pontes,
Álvaro Machado, Henedina Antunes

ABSTRACT

Introduction: Acute transverse myelitis (ATM) refers to a frequently idiopathic, segmental spinal cord inflammation. It is a rare condition, in particular in children, and not previously reported in a family retinitis pigmentosa (RP) clinical setting. **Case Report:** An 11-year-old previously healthy girl, with a family history of RP, presented with a subacute flaccid paraparesis, with bilateral, up to the fourth dorsal level, mixed sensory hypoesthesia and autonomic dysfunction. Brain and spinal cord magnetic resonance imaging (MRI) showed an extensive, T2-hyperintense, non-contrast enhancing lesion from the second to fifth dorsal levels. Cerebrospinal fluid (CSF) and lab studies were normal, as the ophthalmologic observation. Treated with high-dose corticosteroids and intensive physical therapy, a significant recovery could be seen. **Conclusion:**

Early pharmacological and physical treatment is fundamental and may indeed change the prognosis of this disease ATM. The family history of RP, although probably incidental, brings nevertheless the issue of a possible etiological contribution, or pathologic common pathways.

Keywords: Transverse myelitis, Retinitis pigmentosa, Paraparesis, Neurogenic urinary bladder

How to cite this article

Teixeira J, Carvalho S, Martins S, Pontes T, Machado Á, Antunes H. A case of pediatric paraparesis secondary to an idiopathic acute transverse myelitis. Int J Case Rep Images 2014;5(12):826–830.

doi:10.5348/ijcri-2014142-CR-10453

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Received: 10 September 2014

Accepted: 18 September 2014

Published: 01 December 2014

INTRODUCTION

Acute transverse myelitis (ATM) refers to a multiple-level segmental spinal cord injury, caused by an acute inflammatory process.

It is very rare, with an estimated incidence of 1–5 cases per million per year [1]. Of these, only 1/5 occur in children, mainly before the age of two (a bimodal incidence can be seen, with a low number of cases between two and five years) [1, 2].

Although commonly idiopathic, an autoimmune disturbance is frequently suspected, and a polyphasic demyelinating disorder can only be disregarded after a reasonable follow-up period.

Clinically, it is characterized by acute to subacute onset of a variable signs of motor, sensory and autonomic dysfunction, which can be localized to a certain level (commonly a series of adjacent levels) of the spinal cord.

It can have major consequences, with residual sensitive, autonomic and motor dysfunction in up to 20% of cases [3].

Retinitis pigmentosa (RP) refers to a heterogeneous group of inherited ocular diseases resulting in a progressive retinal degeneration. It affects 1 in 3,000–5,000 people and occurs in isolation or in a syndromal manner [4, 5].

CASE REPORT

An 11-year-old previously healthy gymnastics practitioner girl, with anisometropia and family history of RP (mother and maternal aunt), was seen for a 4-day-evolving lower-limb loss of strength and sensitivity, combined with dorsal pain and sphincter dysfunction.

There was no fever or recent relevant traumatic injury.

Neurological examination revealed a left-predominant flaccid paraparesis with normal myotactic and superficial reflexes, a mixed sensory disturbance with algic hypoesthesia up to the fourth dorsal level, and proprioceptive distal loss.

Magnetic resonance imaging (MRI) of the medulla showed slight dorsal high intensity signal in all T2-weighted sequences. Laboratory examination, including all virologies, relevant serologies and immunity screening, were found to be normal. Cerebrospinal fluid (CSF) was also completely normal (including absent oligoclonal bands), as it was the computed tomography (CT) angiography of chest and neck vessels.

Assuming the most likely diagnosis of ATM, she was admitted and started methylprednisolone bolus (30 mg/kg/day).

MRI was repeated at day-12 showing that the signal change extended from the first to fifth dorsal level, was bright in all T2-weighted sequences, did not uptake administered gadolinium, and was more clearly localized in the lateral and anterior columns (Figure 1).

At ophthalmologic evaluation there was no evidence of changes in visual acuity or ocular fundus.

Clinical improvement started at day-5 of methylprednisolone. There was increase muscle strength sufficient for autonomous, although limited, deambulation. It was decided to keep corticosteroid therapy (oral prednisolone 1 mg/kg/day). She also started physiatrist treatment with further muscle strength improvement and gait control. Bladder catheterization was needed because of high post-voiding residual volume.

She was discharged at day-23, with residual paraparesis (Medical Research Council Scale grade 4+), maintaining prednisolone, physiatrist treatment and intermittent bladder catheterization.

Five months later MRI was repeated, showing a reduced dorsal medulla thickness (Figure 2).

Currently, six months after, she maintains paraparesis, being capable of walking for short distances, but needing help for longer distances, personal hygiene and clothing. Neurogenic bladder persists, with secondary nocturnal enuresis, but with spontaneous daytime voiding.



Figure 1: MRI (sagittal section), T2-weighted sequences, showing a first to second dorsal level hyperintense signal, extending to the fourth to fifth dorsal level, more clearly localized in the lateral and anterior columns, with no uptake of administered gadolinium.

DISCUSSION

In ATM, symptoms are grouped on physiological ground as motor, sensory or autonomic. They greatly vary because of the topographical variability (the level, extension and localization) of the disease and, whenever secondary, of the pathology of the underlying cause. As so, clinical onset can be rapidly progressive or more slowly-evolving over a few weeks, and symptoms can affect all limbs, only the lower ones, with or without symmetrical impairment, and predominantly affect one of the three above-mentioned motor, sensory and autonomic systems [2]. MRI is the fundamental study to carry in an emergency setting so to exclude compressive lesions. In ATM,



Figure 2: MRI (sagittal section), T2-weighted sequences, showing a hyperintense signal of anterior dominance in the middle dorsal region, associated with a reduced dorsal medulla thickness.

lesions are found mainly in the white matter surrounding the central medullar channel, usually involving several adjacent medullar segments, and are more easily seen in T2-weighted sequences, where the edema appears bright. This usually precedes the latter medullar atrophy, the sole imagiological evidence of a past ATM [6–8].

In the majority cases, CSF has increased protein content as well as mild lymphocytosis. However, as spinal cord inflammation may not be evident at the beginning, some authors suggest that lumbar puncture should be repeated between second day and seventh day of the disease [6].

Oligoclonal bands should always be sought, because if they are found in the CSF and not in the blood, they raise the risk for multiple sclerosis [9]. An ophthalmologic evaluation is also recommended for all patients, as an additional finding of optic neuritis has a major implication on the diagnosis (multiple sclerosis or optic neuromyelitis) [6, 9].

Treatment is not consensual. The first line therapy is methylprednisolone for 5–7 days, followed by oral prednisolone (1 mg/kg/day) for 3–4 weeks. Non-pharmacological treatment includes intermittent bladder catheterization and physiotherapy [2, 9].

Etiological considerations in ATM should include viral/bacterial infections, autoimmune and connective tissue diseases, demyelinating diseases (multiple sclerosis, neuromyelitis optica), intra or extra-axial tumors and vascular diseases. Regarding the last etiological group idiopathic spontaneous dorsal spinal cord infarction is another possible unusual etiology of acute paraparesis in children. When it is possible to exclude all of these causes, ATM is then classified as idiopathic [1, 2, 6].

No etiologic cause of the ATM was found. Given the family history of (RP), a possible association was raised, although both the absence of prior descriptions and of RP signs in our patient, strongly reduce this possibility. There are few descriptions of RP associated with central nervous system infections. It was related to Creutzfeldt-Jakob disease and with congenital toxoplasmosis [4, 10]. There is also been described a close association between Human T-lymphotropic virus type I (HTLV-I) infection associated myelopathy and RP although the pathogenesis remains to be defined [5]. On this ground, we raise the possibility of an etiological contribution, or pathologic common pathways between ATM and RP.

CONCLUSION

Early pharmacological and physical treatment is fundamental and may indeed change the prognosis of this disease ATM. The family history of RP, although probably incidental, brings nevertheless the issue of a possible etiological contribution, or pathologic common pathways.

Author Contributions

Joana Teixeira – Substantial contributions to conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published

Susana Carvalho – Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published

Sofia Martins – Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published

Teresa Pontes – Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published

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Guarantor

The corresponding author is the guarantor of submission.

Conflict of Interest

Authors declare no conflict of interest.

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Article citation: Teixeira J, Carvalho S, Martins S, Pontes T, Machado Á, Antunes H. A case of pediatric paraparesis secondary to an idiopathic acute transverse myelitis. *Int J Case Rep Images* 2014;5(12):826–830.



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CASE REPORT

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Cystic hygroma of arm treated with OK-432: A case report

Chin Aun Low, Foad Agus Iwan

ABSTRACT

Introduction: Cystic hygroma is a type of lymphangioma and is located most frequently in the head and neck region, followed by axilla, superior mediastinum, and mesentery and retroperitoneal. Its occurrence in upper extremity is rarely reported in literature. **Case Report:** We report a newborn baby with Down's syndrome presented with cystic hygroma in his left arm and forearm. Patient was treated with OK-432 for the swelling of the left arm. However, patient presented at five months of age with severe sepsis secondary to gangrene of left arm and concomitant meningitis which subsequently led to his death. **Conclusion:** Picibanil aka OK-432 has been increasingly used for the last two decades for the treatment of cystic hygroma with much success. However, few literatures report of serious morbidity with its use. More case controlled studies should be carried out to further understand the side effects of sclerosant therapy, as it has the potential to be ideal treatment for cystic hygroma in the future.

Keywords: Cystic hygroma, Lymphangioma, Picibanil, Neonatal gangrene

How to cite this article

Low CA, Iwan FA. Cystic hygroma of arm treated with OK-432: A case report. Int J Case Rep Images 2014;5(12):831–834.

doi:10.5348/ijcri-2014143-CR-10454

INTRODUCTION

Cystic hygroma is the outcome of congenital developmental failure of lymphatic system in the body.

Most cases of cystic hygroma reside in the head and neck region. Other sites are superior mediastinum, axilla, retroperitoneum, mesentery, pelvis and lower limbs. Its occurrence in upper extremity is scarcely reported in literature. Approximately, 50% of cases are diagnosed at birth [1]. Since the introduction of sclerosant therapy, a non-surgical method of treatment has been favored over surgical treatment. We report a newborn with Down's syndrome presented with cystic hygroma in the arm and forearm. He was subsequently treated with a sclerosant known as Picibanil (OK-432) which is a mixture originating from group A Streptococcus pyogenes.

CASE REPORT

A newborn male neonate with features of Down's syndrome presented to the special care nursery (SCN) to our institution with complaint of swelling of left arm and left forearm on day-1 of life. There were no other complaints. Clinical examination revealed swelling of the left neck extending to the left arm until proximal forearm measuring 12x8x7 cm. The swelling showed no sign of inflammation. On examination, the swelling appeared to have multiple cystic areas, which were easily compressible and brilliantly transilluminant without any bruit.

Magnetic resonance imaging (MRI) scan revealed cystic lesion extending from C2 proximally to left

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Received: 24 September 2014

Accepted: 15 October 2014

Published: 01 December 2014

proximal forearm distally. The upper neck lesion appeared subcutaneous and lower neck lesion extended into the intermuscular plane of left sternocleidomastoid, infraspinatus and deltoid. At the chest level, it lied between left pectoralis major and pectoralis minor anteriorly and extended up to left teres major and latissimus dorsi posteriorly. At the arm level, it compressed the left coracobrachialis muscle (Figure 1). Neurovascular structures were intact and normal bone structures were shown.

Our diagnosis was cystic hygroma of left arm and left forearm that was evidently supported by MRI. The patient was then referred to a tertiary center and was treated with OK-432 sclerotherapy. Fine-needle aspiration was not done to prevent introduction of infection to the upper limb and due to high recurrence rates of lymphangioma after aspiration as a therapeutic procedure.

The baby started the OK-432 therapy at third months of life. The patient was given a single dose of 0.2 mg (20 mL) of OK-432 over the left deltoid where the swelling was most prominent. There was noticeable reduction in the size of the swelling for the first two months from time of injection to the size of 8x5.5x4 cm. However, he was admitted to SCN in our institution at fifth month with complaints of fever, fits and blackish discoloration of the left upper limb. The parents denied any traditional medication applied to the baby's left arm and forearm. On examination, patient has pyrexia and there were gangrenous patches over the left arm which was the site of previous injections of OK-432 with an open wound measuring 6x2 cm over the lateral aspect of the left arm (Figure 2). The patient was then intubated in view of severe metabolic acidosis with pH of 6.73 and was supplemented with intravenous bicarbonate. The patient was treated as severe sepsis secondary to gangrene of left upper limb and concomitant meningitis. The patient was started on dual antibiotic therapy with intravenous ampicillin and cefotaxime given based on weight-based dosage. On day two of admission, patient was diagnosed with disseminated intravascular coagulation and was transfused with fresh frozen plasma, platelet and packed cells. 500 cm³ of hemoserous fluid was aspirated from the left upper limb as a therapeutic procedure but the swelling recurred within the following two days. Unfortunately, the baby's condition deteriorated over the following five days and died due to unresolved severe sepsis.

DISCUSSION

Lymphangiomas may be classified histologically into three types namely simple, cavernous, and cystic. Simple lymphangiomas are composed of minute lymphatic channels communicating with its stroma components whereas cavernous lymphangiomas consists of dilated lymphatic components surrounded by stroma. Cystic lymphangioma are larger cystic lesions which poorly communicate with the lymphatic elements.



Figure 1: Magnetic resonance imaging (MRI) scan showing cystic lesion extending from C2 proximally to left proximal forearm distally.



Figure 2: Severe infection of left upper limb with blistering and necrotic patches seen.

Although cystic hygroma has been associated with a nuchal lymphangioma, Turner's syndrome, and Noonan syndrome, there is evidence that cystic hygroma occurs more frequently in Down's syndrome. It has been postulated significant correlation with Down's syndrome as patients with Down's syndrome have potentially enlarged jugular lymphatic dilatations [2].

Traditionally, cystic hygroma has been treated with surgical excision. However, this mode of treatment was often accompanied with serious complications, namely infection, recurrence, bleeding and injury to major nerve

and vessels. Surgical excision was only accompanied with a one-third possibility of successfully preserving all vital structures [3]. The recurrence rates are high even after apparent complete excision of the lesion.

Sclerotherapy as the primary modality of treatment is under trial in many centers. The sclerosant agents currently under researches are: OK-432 (Picibanil), bleomycin, doxycycline, alcohol, alpha interferon-2 and fibrin sealant [4]. Most of the studies documented complete resolution of the lesions with OK-432 in about 60–80% of cases.

OK-432 (Picibanil) is a lyophilized incubation mixture of group A *Streptococcus pyogenes* of human origin. Since its introduction as the main therapy for lymphatic malformations by Ogita in 1987, it claims better results with fewer complications. OK-432 acts by provoking inflammatory responses towards inactive bacteria, leading to increased endothelial permeability, which increases lymphatic drainage of that selected cystic region. In the later stage, fibrosis of cystic spaces predominates and will lead to shrinkage of the tumor [5].

Poldervaart's study showed that microcystic variety of lymphatic malformation treated with OK-432 alone had 27% of excellent result, 33% good result and 40% poor result, while of macrocystic variety of lymphatic malformation; 88% had excellent result. Since cystic hygroma is of macrocystic variant, the expectation of successful therapy with OK-432 alone is above 80%. In their data, the recurrence rates vary from 5–8%. The adverse effects are mostly mild, such as mild edema, erythema, pyrexia, induration and wrinkling of skin at site of injection [6]. Almost all of the adverse effects disappear after a week.

We presume the open wound and gangrene was due to unresolved infection at the site of injection, but we cannot be certain whether it was attained at time of injection or subsequent poor wound care or secondary infection caused by underlying tissue inflammation. It is obvious that the death of the patient was due to severe sepsis. Whether there is any correlation with the injection of OK-432 cannot be clarified. Most of the trials of OK-432 have short follow-up. There are uncertainties when it comes to cure and regression. Furthermore, mortality rate associated with the use of OK-432 has never been reported.

CONCLUSION

Cystic hygroma of left arm and left forearm is rarely reported. Picibanil aka OK-432 has been increasingly used for the past 2 decades for the treatment of cystic hygroma with much success. However, few literatures report of serious morbidity with its use. More case controlled studies should be carried out to further understand the side effects of sclerosant therapy, as it has the potential to be the ideal treatment for cystic hygroma in the future.

Author Contributions

Chin Aun Low – Substantial contributions to conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published

Foad Agus Iwan – Substantial contributions to conception and design, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published

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The corresponding author is the guarantor of submission.

Conflict of Interest

Authors declare no conflict of interest.

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CASE REPORT

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Management of dentoalveolar fracture with multiple avulsions: A case report with three years follow-up

Sangeetha K.M., Poornima Surendra, Roshan N.M., V.V. Subba Reddy, Rashmi G. Chaur, Sagar B. Srinivasa

ABSTRACT

Introduction: Traumatic dental injuries are common in children and adolescents and may cause dramatic episodes. Avulsion management associated with dentoalveolar fracture may pose significant difficulty to the clinician. Acute dental treatment is an important requisite after such injuries. Immediate management and continuous follow-up is necessary especially with ever long-term changing treatment protocols. **Case Report:** Herein we describe a 10 year-old-boy, referred to our hospital for multiple teeth avulsions, extrusions including molar which is very rare and dentoalveolar fracture as he met with an accident while playing. Timely treatment was done with cap splint appliance. Patient is doing well with three years of follow-up. **Conclusion:** We highlight the

significance of storage media for carrying the avulsed teeth, and cap splint for stabilization of complicated dentoalveolar fracture which resulted in excellent treatment and a long-time prognosis

Keywords: Avulsion, Reimplantation, Dentoalveolar, Fracture

How to cite this article

Sangeetha KM, Surendra P, Roshan NM, Subba Reddy VV, Chaur RG, Srinivasa SB. Management of dentoalveolar fracture with multiple avulsions: A case report with three years followup. Int J Case Rep Images 2014;5(12):835–838.

doi:10.5348/ijcri-2014144-CR-10455

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Received: 02 August 2014
Accepted: 03 September 2014
Published: 01 December 2014

INTRODUCTION

Dynamic state of the alveolar and dental development pose a challenge to the practitioner for the management of dentoalveolar injuries in an adolescent. Traumatic dental injuries comprises 5% of all the injuries in preschool, school and young adults for which people seek treatment [1]. Of all dental injuries, avulsion of permanent teeth comprise 0.5–3% [2]. Numerous studies have shown that after the avulsion, prognosis is very much dependent on the actions which are taken at the place of accident [1–4]. Also replantation is not indicated in all the situations like severe caries or periodontal diseases, severe cardiac conditions, non-cooperative patient and severe medical conditions. Even though replantation can save tooth, it is important to realize that there are lower chances of long-term survival and even may be lost at a later stage, total extra oral time and storage media/transport media used for carrying avulsed permanent tooth helps treatment planning [3].

CASE REPORT

A 10-year-old boy reported to the Department of Pedodontics and Preventive Dentistry with a traumatic injury to oral and perioral region because of fall when the child was playing cricket in the school. History suggests that the trauma resulted in avulsion of several teeth. By the advice of local dentist, parents of the injured child had carried the avulsed tooth in milk as it is a good storage media and also readily available. Two hours were elapsed by the time the patient reported to the Department of Pedodontics.

On examination child had bruises and swelling of upper lip. Intraoral examination revealed dentoalveolar fracture involving premaxilla and upper right posterior segment with palatal displacement of posterior teeth. There was severe palatal gingival laceration with bleeding. There was an avulsion of 21 and 24 which were carried in milk, 22 was palatally displaced and there was an extrusion of 25, 26, 31, 32 and 41 (Figure 1). The premolar and molar were completely out of their sockets but hanging with little tissue attachment and remained intraorally. Radiographic examination revealed fracture of interdental bone between 25 and 26 (Figure 1). Premolars were immature with only two-third of root formation. Occlusion was deranged because of palatal displacement of left maxillary posterior segment.

Treatment

There was no history suggestive of head injury, child had received first aid medical treatment for minor bodily injuries by school medical officer who had administered tetanus toxoid injection.

Emergency management of dental traumatic injury was planned after thorough history, radiographic (orthopantomograph) and hematological (bleeding time and clotting time) investigations. Under local anesthesia the palatal gingiva was sutured and bleeding was arrested. The avulsed teeth (21 and 24) were reimplanted successfully, after rinsing with saline. Widened dental socket and dentoalveolar fracture in the region of 25 and 26 were reduced with a blunt instrument and digital pressure. Premolars were repositioned and stabilized with interdental sutures. Since it was dentoalveolar fracture, for immobilizing displaced maxillary segment cap splint was planned. Alginate dental impression was made and cast were poured. Displaced maxillary segment was cut, rearranged on the cast and checked with occlusion of the lower jaw and modified cap splint (covering occlusal surface) was fabricated.

Under local anesthesia the displaced palatal segment was reduced and modified cap splint was cemented using zinc phosphate cement (Figure 2). Extruded mandibular anterior teeth were stabilized with flexible wire splint. Amoxicillin 250 mg 8 hourly and analgesics were

prescribed and patient was advised to take soft diet for 10 days.

The child was followed-up the next day for checking the stability of the splint. Although the child was comfortable, showed slight inflammatory swelling of the left side of the face. After ensuring the stability of splint, the child was advised to continue medication and soft diet. Patient was followed-up after one week and found that facial swelling has subsided and child coping up with treatment procedure.

Two weeks later, the modified cap splint was removed carefully and occlusion was ascertained. Occlusion was found to be satisfactory. Radiographic evaluation was done to evaluate present status of avulsed teeth. Flexible wire splint was placed for dentoalveolar stabilization. Root canal treatment was initiated for all the affected teeth. Intermediate calcium hydroxide dressing (ApexCal) was given for premolars as there was beginning of external inflammatory root resorption (Figure 3).

Two months later, radiographs revealed continued external inflammatory resorption with 24 and 25 so decision was made to obturate with mineral trioxide aggregate (Figure 4).

Three years follow-up revealed that the child was asymptomatic with normal function. On examination clinically, the child was asymptomatic showing



Figure 1: Orthopantomograph revealing empty sockets with 21, 24. Extrusion of 25, 26, 31, 32 and 42 with interdental bone fracture between 25 and 26.



Figure 2: Modified cap splint cemented.

infraocclusion with 21 but normal occlusion posteriorly. Radiographically, 21 and 24 revealed complete root replacement resorption even though clinically asymptomatic. However, 22, 26, 31, 32 and 42 were asymptomatic clinically as well radiographically revealed intact lamina dura (Figures 4 and 5).

DISCUSSION

Fractured alveolar process requires reduction, immobilization followed by stabilization for 2–4 weeks for its treatment. Arch bars are not suitable in children due to the size of teeth in mixed dentition and newly erupted permanent teeth have immature roots. Since in our case all the posteriors and central incisor were avulsed, modified acrylic splint was considered in order to stabilize both dentoalveolar fragment and the avulsed teeth. Although the avulsed teeth should not be splinted for more than 7–10 days, since there was associated dentoalveolar fracture, the cap splint was extended for two weeks [4].

The present case showed successful replantation as inflammatory resorption was arrested, although the avulsed teeth were carried in milk few minutes after the trauma, replacement resorption was continuous. But the lower incisors and the upper molar were replaced successfully.

Inflammatory process and cell resorption activity should be eliminated for treatment of root resorption. calcium hydroxide (CaOH) known to be bactericidal and osteogenic potential which is widely used in endodontics [5]. Inhibition of osteoclastic activity results in formation of hard tissue as it creates an alkaline environment in and around the tissue. The reason being CaOH used in the present case. The diffusion of calcium and hydroxide ions through dentinal tubules to the root surface [6]. A change in the concentration of hydroxide ions disturbs the pH gradient at the cell membrane of bacteria thus disrupts the energy supply of the organism. Also its high pH causes dematuration of the cell membrane proteins and intracellular toxins [7].

Mineral trioxide aggregate consisting of calcium and phosphorus, calcium hydroxide is formed when it reacts with tissue fluids. Tavolet et al. have suggested that after the material release calcium, mineralization gets stimulated, which forms calcium carbonate by reacting with tissue carbonic gas [8]. But Ozdemis et al. showed mineral trioxide aggregate did not produce an alkaline shift in the immersion media and by virtue of its high pH mineral trioxide aggregate should not be expected to heal the lesion [9]. Ginger Koshy George has shown that, calcium release by apexcal is greater than Mineral trioxide aggregate, with significant increase with time and advocated CaOH may be potentially used in cases of root resorption [10]. Hence in this, case we used CaOH in the beginning to stop the resorptive process and later Mineral trioxide aggregate obturation was done for the premolars.

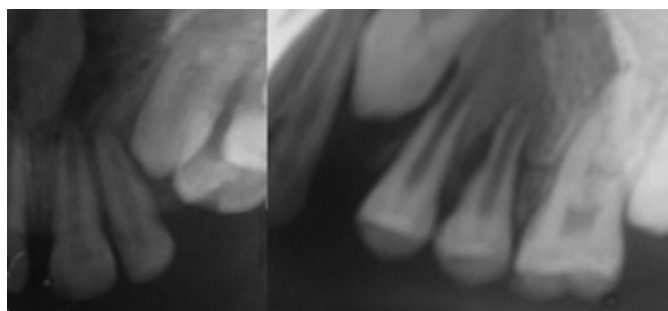


Figure 3: External root resorption with 24 and 25.

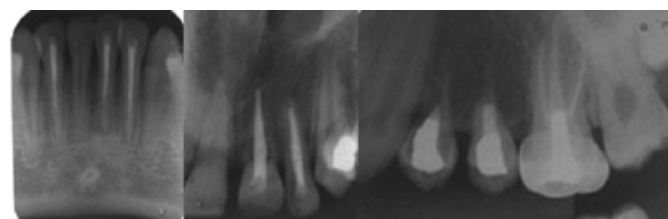


Figure 4: After three years follow-up showing intact lamina dura with 22,26, 31,32 and 42 and replacement resorption with 21,24 and 25.



Figure 5: Orthopantomograph after three years.

Even though there was a replacement resorption, we could achieve the retention of teeth for three years and still functioning clinically also maintenance of bone height and width for future implant procedures.

The three-year period of retention of permanent traumatized teeth reveals successful clinical techniques utilized to treat this child both in terms of esthetics as well function. Although there was a resorption, the decision to retain the premolars with mineral trioxide aggregate obturation was shown to be satisfactory. Hence in the present case we used CaOH in the beginning to stop the resorptive process and later mineral trioxide aggregate obturation was done. However, the first molar and the incisors showed no resorption with intact periodontal ligament space.

CONCLUSION

Awareness about storage media for avulsed teeth among common people help in better prognosis of replanted teeth. Modified cap splint can be used successfully for stabilization of complicated dentoalveolar fracture along with multiple teeth avulsions and extrusions.

Author Contributions

Sangeetha K.M. – Substantial contributions to conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article, Final approval of the version to be published, guarantor and

Poornima Surendra – Substantial contributions to conception and design, Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published

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Conflict of Interest

Authors declare no conflict of interest.

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CASE REPORT

OPEN ACCESS

De Garengeot's Hernia: A rare presentation of an ischemic appendix within a strangulated femoral hernia in an elderly male

Wahid Abdul, Charlotte Thomas, Keshav Swarnkar

ABSTRACT

Introduction: De Garengeot's hernia; the presence of a vermiform appendix in the femoral hernia sac, is a rare occurrence. Due to the paucity of cases and no large case series yet published, the incidence of De Garengeot's hernia is reported between 0.5–5%. Femoral hernias account for less than 5% of all groin hernias and are three times more common amongst females and amongst the elderly. We report an unusual case of an 84-year-old male patient with a strangulated femoral hernia sac containing an ischemic appendix. **Case Report:** An 84-year-old male presented to the emergency department with a 48 hour history of an increasingly painful right-sided groin swelling. A clinical diagnosis of an incarcerated right inguinal hernia was made and the area surgically explored. At exploration, an incarcerated femoral hernia sac containing an ischemic appendix was discovered. Appendicectomy was performed and the hernia defect repaired with nylon sutures. Post-operatively no surgical complications were

encountered and histological report confirmed clinical findings. **Conclusion:** The presence of a vermiform appendix in the femoral hernia sac is a rare occurrence, and has been more commonly reported amongst female patients. Due to atypical presentation, preoperative diagnosis of De Garengeot's hernia is difficult. Early surgical intervention avoids potential complications.

Keywords: De Garengeot's Hernia, Ischemic appendix, Groin hernia, Appendicectomy

How to cite this article

Abdul W, Thomas C, Swarnkar K. De Garengeot's Hernia: A rare presentation of an ischemic appendix within a strangulated femoral hernia in an elderly male. Int J Case Rep Images 2014;5(12):839–843.

doi:10.5348/ijcri-2014145-CR-10456

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Received: 26 August 2014

Accepted: 25 September 2014

Published: 01 December 2014

INTRODUCTION

The incidence of acute appendicitis in the general population during their lifetime is approximately 7% [1, 2]. In the United States, 96% of all groin hernias are inguinal and 4% are femoral with approximately 27,000 cases of femoral hernias encountered per year [3, 4]. The reported female-to-male ratio of femoral hernias varies between 3:1 [3] to 6:1 [5] and are more commonly observed in elderly women.

In the early 18th century, Rene Jacques Croissant De Garengeot reported the presence of a vermiform appendix in a femoral hernia sac and this was coined De Garengeot's Hernia [6, 7]. However, the first appendicectomy in a femoral hernia sac was only performed fifty-four years later by Herin [6].

The presence of a vermiform appendix in a femoral hernia sac is very uncommon with an estimated incidence between 0.5–5%, whilst appendicitis in a femoral hernia

is a far more uncommon finding [6, 8]. Due to this infrequent occurrence, it is only mentioned sparingly in literature, predominantly in the form of case reports.

We report an unusual case of an 84-year-old male patient with a strangulated femoral hernia sac containing an ischemic appendix.

CASE REPORT

An 84-year-old male was presented to accident and emergency department complaining of a two-day history of increasing, constant right groin pain and swelling. The patient underwent bilateral inguinal hernia repair 10 years ago and suffered a recurrence of right inguinal hernia during 2012; subsequently managed conservatively as he was deemed not medically fit for operative repair and the hernia had rarely concerned the patient.

The patient began to experience nausea, vomiting and poor appetite prior to the onset of pain. Furthermore, he noticed he had not opened his bowels for three days and was unsure whether he was passing flatus which was unusual for him.

His past medical history included NSTEMI, congestive cardiac failure, hypertension, hypothyroidism, COPD, transitional cell carcinoma of bladder and Parkinson's Disease. Medications included tamsulosin, aspirin, nicorandil, levothyroxine, simvastatin, bisoprolol, bumetanide, diazepam and ramipril.

On clinical examination, the patient was in moderate distress. However, vital signs were stable and he was afebrile. Abdominal examination revealed severe tenderness located over the right groin lump; the lump was tender and irreducible and bowel sounds were present. There were no palpable masses and digital rectal examination was normal.

Blood tests revealed an elevated C-reactive protein (CRP) of 43.3 whilst an abdominal X-ray was unremarkable. A working diagnosis of an incarcerated right inguinal hernia was made and due to the urgency of clinical presentation he was listed for emergency exploration of the right groin lump with a view to proceeding, if necessary.

An incision was made across the groin crease directly overlying the groin swelling. Exploration of the inguinal area revealed no hernia sac. The femoral canal was explored and incidentally an incarcerated femoral hernia sac containing an ischemic appendix was identified (Figure 1). The appendix base was easily delivered into the wound. As the cecum could not be delivered into the wound, the appendix was ligated and divided proximally close to the cecum; visible at the widened femoral ring. The base of the appendix was transfixed and an appendectomy performed through the same incision. A lower midline laparotomy was contemplated. However, due to patients' co-morbidities this less invasive procedure was persevered with. The hernia defect was closed with 2-0 nylon sutures. The fascia was closed with

vicryl sutures and monocryl sutures were used for the subcuticular layer. The resected appendix specimen was sent for histopathology (Figure 2).

Postoperatively, the patient developed a chest infection which was treated with intravenous antibiotics. Histopathology report confirmed atrophic and ischemic changes of the appendix.

DISCUSSION

Femoral hernias predominantly occur in women with a female to male ratio varying between 3:1 [3] and 6:1 [5]. The incidence of femoral hernia in men is around 2% [5, 9]. Due to the narrowness and rigidity of the femoral canal, femoral hernias have a higher rate of incarceration compared to inguinal hernias (56% compared to 6–10%) [10].

Factors increasing intra-abdominal pressure, and thereby predisposing to femoral hernias, include pregnancy, obesity, pelvic mass, urinary retention and constipation. Furthermore, weakening of transversalis fascia may be implicated. Our patient presented with a three-day history of constipation which may have contributed to the femoral hernia.

In 1731, Rene Jacques Croissant De Garengeot first

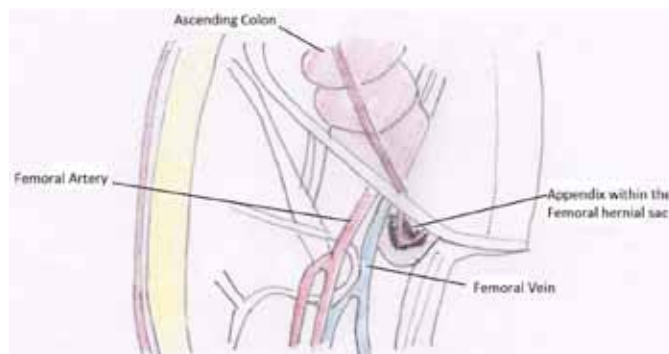


Figure 1: Anatomy of the femoral canal illustrating the position of the De Garengeot's Hernia.



Figure 2: Excised appendix specimen present in the femoral hernia sac.

reported the presence of an appendix in a femoral hernia sac. Fifty-four years later, Herin performed the first appendectomy in a femoral hernia sac [6]. The term De Garengeot's Hernia includes either a normal, inflamed, perforated or gangrenous appendix in a femoral hernia sac. De Garengeot's Hernia is a rare occurrence, often presenting with a painful lump inferior to the inguinal ligament [11]. Our patient presented with a painful lump in his right groin which was clinically diagnosed as an incarcerated right inguinal hernia.

Wakeley reported a 1% incidence of vermiform appendix in a femoral hernia in a personal case series on 655 patients [12] whilst Ryan reported a 0.13% incidence of acute appendicitis in an external hernia amongst 8692 cases [13]. However, due to the paucity of cases and no large case series yet published, the incidence of De Garengeot's Hernia is reported to be between 0.5–5% [6, 8, 14].

The majority of De Garengeot's Hernia reported in literature involved elderly women with the mean age of 69 years [4]. Our patient was an elderly male, thereby demonstrating uncommon presentation in this population group.

Pathogenesis

The hernia sac can often contain preperitoneal fat, omentum, colon or small bowel. The pathogenesis of De Garengeot's hernia is controversial. One of the theories; the congenital theory, proposes that abnormal attachment of appendix onto the cecum predisposes to pelvic appendix which can enter the hernia sac of the pelvic peritoneum [4, 9]. Other theories propose either primary or secondary obstructive events, with the latter as a results of constriction of appendix by a tight hernia neck-sac as a cause of appendicitis in femoral hernia. The latter theory may have contributed to the presentation in our patient, as he was constipated for three days without passing flatus and was vomiting.

Clinical Features

Diagnosing femoral hernias preoperatively is challenging due to the difficulty in palpating the hernia in asymptomatic patients and the resemblance to inguinal hernia when hernia is swollen and inflamed. Furthermore, they can present as thigh or groin swellings. Pain from appendiceal hernia is usually of a cramping nature rather than constant [15]. Our patient described a constant groin pain which would have made the clinical diagnosis of femoral hernia difficult.

Preoperative diagnosis of De Garengeot's hernia is difficult, and is frequently made at surgery with only one case reported in literature of diagnosis pre-operatively, with the aid of a CT scan [16]. As with our case, the initial clinical diagnosis of an incarcerated right inguinal hernia

was disproved upon surgery and a De Garengeot's hernia was identified.

Appendicitis in the elderly is often difficult to diagnose with approximately 50% cases being perforated during surgery compared to only 20% in young adults. Furthermore only 20% of patients aged 60 years and above have classical features such as fever, right lower quadrant pain and leukocytosis [17]. Our patient was 84-year-old and only presented with right groin pain with an elevated CRP. The absence of these classical features may provide a possible explanation as to why an ischemic appendix was identified at exploration rather than appendicitis.

Repair

Due to the paucity of De Garengeot's hernia cases there are no standard treatment for De Garengeot's hernia. Management includes either; incision and drainage with a delayed appendectomy or an immediate appendectomy and repair of hernia sac using McVays's repair [4]. McVay's repair employs non-absorbable interrupted sutures to bring the conjoint tendon to Cooper's ligament from the pubic tubercle to the femoral vein [18]. This technique was employed in our patient at exploration.

Early surgical treatment is vital to prevent potential complications. Postoperative monitoring of patients is vital as approximately one-third of the patients with De Garengeot's Hernia repair have an infection post-operatively [11]. Due to early surgical intervention our patient may have avoided these potential complications.

The majority of De Garengeot's Hernia cases observed and described in the literature were observed women. De Garengeot's Hernia in males are less common and therefore less frequently mentioned in literature. Cases described in literature commonly involved the right side of the groin with Scepi et al. (1993) reporting a case presenting in the left groin region [19].

Shum J and Croome K (2012) described a case of a 72-year-old male with a previous right sided non-mesh inguinal hernia repair, presenting with a three-day history of swollen, tender mass in the right inguinal region. During exploration, the appendix was inflamed within the femoral hernia sac and an appendectomy followed by hernia sac repair using McVay's technique with interrupted non-absorbable sutures was performed [18].

Pitchaimuthu and Dace (2009) reported the youngest male patient with De Garengeot's Hernia; a 40-year-old male who had presented with a week-long history of a painful right-sided groin swelling. During exploration, a mildly inflamed appendix was identified as an incarcerated femoral hernia. An appendectomy, followed by Lichtenstein hernia repair, was performed [20].

Wiszniewski et al. (2008) reported a 55-year-old male who had presented with a four-day history of a painful

right-sided groin swelling. On examination a 10x5cm painful mass was present in the right groin and a working diagnosis of an incarcerated right inguinal hernia was suspected. Intra-operatively, the femoral hernia sac contained a gangrenous, inflamed, non-perforated appendix which was excised and the femoral hernia was repaired [21].

In a case series, Sharma et al. retrospectively analyzed 457 femoral hernia repairs performed during January 1991 to September 2006 and identified seven patients with De Garengot's hernia. Six of these patients had an acute presentation with an average of 24–48 hours history of symptoms. All seven patients had normal white cell counts and were approached using an infra-inguinal incision. Three of these patients had an inflamed appendix, one was perforated with the remainder being normal. In all of these cases the femoral hernia was repaired using prolene mesh in normal appendix, whilst abnormal appendixes were excised and the femoral canal was closed with interrupted prolene sutures [11].

In another case report, an 88-year-old male presented with a two-day history of a painful right-sided groin mass. Clinically, a 6 cm tender non-reducible right inguinal mass was palpated and a presumptive diagnosis, of a strangulated inguinal hernia, was made. During surgery, a femoral hernia sac containing a perforated appendix was identified. Following appendicectomy, the femoral hernia was repaired. Histopathological studies subsequently reported an acute-on-chronic appendicitis with perforation of appendix [4].

Isaacs and Felsenstein (2002) described a 76-year-old man with a 6-year history of right groin hernia presented with a two-day history of right groin pain and increased swelling. During operative repair the hernia sac containing an inflamed appendix was found within the femoral canal. The appendix was excised and the hernia sac repaired using McVay's technique [7].

CONCLUSION

De Garengot's Hernia; the presence of a vermiform appendix in a femoral hernia sac, is an unusual occurrence which is commonly observed in women and in elderly patients. De Garengot's hernia is commonly encountered on the right side as a groin swelling. Preoperative diagnosis is challenging and, without radiological investigations, it is often made intra-operatively. Early surgical intervention is imperative to prevent potential complications. This is a case of an elderly male with a two-day history of right groin pain and swelling with an elevated C-reactive protein. Upon surgical exploration De Garengot's hernia was identified. Our case represents another rare case of De Garengot's Hernia, in an uncommon population group.

Author Contributions

Wahid Abdul – Substantial contributions to conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published

Charlotte Thomas – Substantial contributions to conception and design, Revising it critically for important intellectual content, Final approval of the version to be published

Keshav Swarnkar – Substantial contributions to conception and design, Revising it critically for important intellectual content, Final approval of the version to be published

Guarantor

The corresponding author is the guarantor of submission.

Conflict of Interest

Authors declare no conflict of interest.

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CASE REPORT

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Obstructive jaundice secondary to postsurgical persistent residual hydatid ectocyst of left lobe of liver

Maulana M. Ansari, Shahla Haleem, Wasif M. Ali, Leonard Enzeung, Sheikh Sarfraz Ali, Sunder K. Meet

ABSTRACT

Introduction: Liver is the most commonly affected by cystic echinococcosis and surgical treatment is usually curative. Although recurrence is not uncommon but obstructive jaundice secondary to persistent large residual ectocyst with compression at porta hepatis is an unusual complication not yet reported in literature. **Case Report:** A 30-year-old female complained of persistent abdominal pain for eight months following surgical treatment of the liver hydatid cyst and increasing jaundice for one month. Imaging revealed a cyst in the same area adjacent to left lobe of liver. Abdominal exploration revealed hydatid ectocyst under tension, compressing the porta hepatis, common bile duct and neck of the gallbladder. Subtotal excision was performed. Postoperative period was uneventful and jaundice resolved in two weeks' time. **Conclusion:** Follow-up longer than

six months is advisable after surgical treatment of liver hydatid cyst for early detection of complications in the residual ectocyst. Fine-needle aspiration under image guidance appears as a reasonable option in the recurrent liver cyst following primary surgical treatment before embarking on re-laparotomy.

Keywords: Hydatid, Liver hydatid, Obstructive jaundice, Postsurgical ectocyst, Residual ectocyst

How to cite this article

Ansari MM, Haleem S, Ali WM, Enzeung L, Ali SS, Meet SK. Obstructive jaundice secondary to postsurgical persistent residual hydatid ectocyst of left lobe of liver. Int J Case Rep Images 2014;5(12):844–848.

doi:10.5348/ijcri-2014146-CR-10457

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Received: 30 May 2013
Accepted: 23 July 2013
Published: 01 December 2014

INTRODUCTION

Tapeworm *Echinococcus granulosus* is a common cause of hydatid disease that may affect any part of the body primarily or secondarily. Its wide prevalence have been reported from cattle, and sheep breeding countries such as Middle-East, Mediterranean, Australia, New Zealand, North and South America [1, 2]. Liver is the most commonly affected organ (70%), followed by the lung (20%) and other organs such as brain, thyroid, spleen, pancreas, gallbladder, etc. (10%) [1–4].

Recently, we encountered an unusual case of liver hydatid cyst that got complicated by persistent symptomatic large residual ectocyst with compression of porta hepatis and obstructive jaundice following primary surgical treatment. Surprisingly, on web search, we did not find anything related to the problem in our patient although recurrence of the liver hydatid cyst has been cited from 1.1–22% of cases [5] and hence we present this case report.

CASE REPORT

A 30-year old female was referred to us after detection of jaundice and a large cyst abutting the left lobe of the liver on check abdominal ultrasound (USG) done for abdominal pain after eight months of symptom-free period following the uneventful recovery from laparotomy for hydatid cyst of left lobe of the liver at other institution. There was no history of close contact with cattle or pet animals in the patient's house. Repeat abdominal USG showed a cyst located posterior to the stomach and abutting the left lobe of the liver, raising the suspicion of a recurrent hydatid cyst, postsurgical residual cavity or pseudocyst of the pancreas. Common bile duct was compressed by the cyst and there was mild dilatation of the intrahepatic bile ducts. Chest X-ray was clear. Hemoglobin was 12.2 g/dL, white blood cell count 8500/mm³ (N48, L45, E4, M3), and an absolute eosinophil count 290/mm³ (Biological Ref.: 50–450 mm³). Serum bilirubin was 2.5 mg/dL (Direct 1.90 mg/dL and Indirect 0.60 mg/dL), SGOT/AST 120.0 IU/L, SGPT/ALT 180.0 IU/L, and serum alkaline phosphatase 244.0 U/L, suggestive of an obstructive jaundice. Contrast-enhanced computed tomography (CECT) of the abdomen revealed a large thick walled cyst (10.3x8.5x10.0 cm) arising from the left lobe of the liver (Figure 1A–B). The cyst was compressing the porta hepatis and common bile duct and the gallbladder (Figure 1C) but the intra-hepatic bile ducts were not dilated. The patient was reviewed on high definition ultrasound machine by a senior radiologist that showed dilated intra-hepatic bile ducts, confirming the obstructive jaundice but unfortunately the ultrasound films could not be taken due to financial constraints. Albendazole 400 mg twice a day was started and re-laparotomy was planned.

Abdominal exploration through the previous midline scar revealed a large thick walled residual tense cyst which measured about 10.5 cm in diameter and was attached with a wide base to the under surface of left lobe of the liver, extending to and compressing the porta hepatis, the common bile duct and even the neck of the gallbladder. There was dense fibrosis and adhesions around the cyst, but the cyst did not have any connection to the pancreas as was suspected in a few cuts of CT (Figure 1D). Aspiration (10 mL) revealed non-bilious non-watery slightly turbid serous fluid not suggestive of hydatid fluid, and 10 mL of 10% povidone-iodine was still instilled and kept for 10 minutes as a precautionary measure. The cyst was guarded with povidone-iodine soaked abdominal sponges and then opened up. There was no element of the live or dead hydatid endocyst, and the cyst contained only fluid (~150 mL), suggestive of persistent previous ectocyst under tension. Subtotal excision of the ectocyst using the monopolar cautery hook was done, leaving behind a 1-cm rim of the cyst wall attached to the liver. There was no other cyst in the rest of the abdomen. The abdomen was closed after thorough lavage with saline

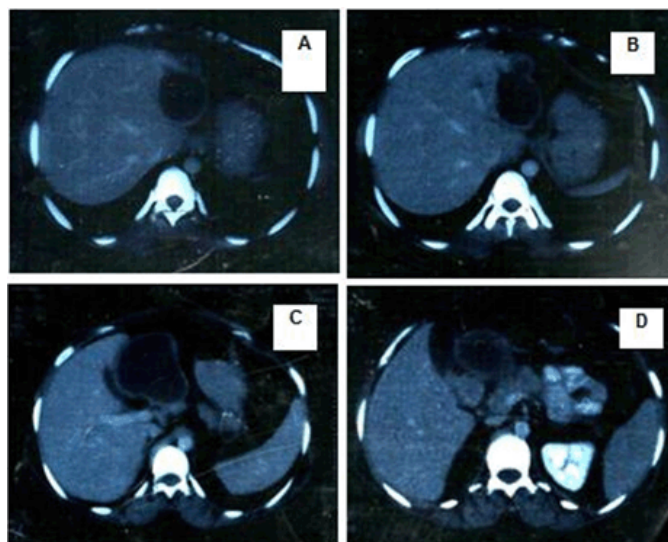


Figure 1: Computed tomography of abdomen: (A, B) Residual cyst present within left lobe of liver, (C) Cyst compressing common bile duct, (D) Cyst abutting and appearing to arise from pancreatic head.

with a suction tube drain in the hepatorenal pouch. Postoperative period was uneventful.

The drain was removed on third postoperative day. The patient was discharged on eighth postoperative day with Albendazole 400 mg twice a day. Liver functions were normalized in 2 weeks' time: serum bilirubin 1.0 mg/dL, SGOT 12.0 IU/L, SGPT 18.0 IU/L, and serum alkaline phosphatase 24.4 U/L, and check abdominal ultrasound was within normal limits. The patient was asymptomatic at four weeks of follow-up when anti-helminthic therapy was stopped.

DISCUSSION

Hydatid disease is caused by *Echinococcus* larva (tapeworm). *Echinococcus granulosus* is the most common causative parasite infesting the humans that produces unilocular hydatid cyst (cystic echinococcosis); uncommonly, *Echinococcus multilocularis* and *Echinococcus vogeli* may infect the humans, producing alveolar echinococcosis and polycystic echinococcosis, respectively [6].

Highest incidence of cystic echinococcosis has been reported from the temperate countries, including North America and South America, Australia, New Zealand, Mediterranean countries, the southern and central parts of the former Soviet Union, Central Asia, Middle-East Countries, China, and parts of Africa [2, 6]. It is endemic in sheep-breeding countries, posing a serious health problem [4].

Several species of carnivorous animals may act as the definitive host, the most important being the dog. Most important intermediate hosts are cows and sheep globally, but sometimes, humans get infested by consuming the ova of the parasite.

Adult *E. granulosus* is 3–6 mm long parasite that lives in the bowels of dogs, cats, wolves, foxes and other carnivorous animals, and health of these definitive hosts are not affected by the parasite [4]. Released eggs get scattered throughout the environment by their feces and may pass to the humans via contaminated vegetables and food. The ingested egg liberates an embryo in the duodenum. The embryo penetrates the intestinal mucosa and enters the portal venous circulation [7, 8].

Liver, acting as the first filter, stops about 75% of the embryos; Lung, acting as the second filter, stops only about 10% of these embryos; and about 15% of the embryos cross both the filters and enter the systemic circulation that may affect any part of the body, producing unilocular hydatid cyst [9]. If a cyst ruptures, it may lead to the development of many new hydatid cysts inside or outside the parent organ [2].

Persistent residual cyst in our patient is intriguing, especially in the absence of biliary communication. The presence of dense adhesions between the cyst and the stomach, and presence of tension within the cyst possibly lead to its expansion towards the porta hepatis with resultant bile duct obstruction and jaundice. This is rather an unusual mechanism of obstructive jaundice due to external compression of a postsurgical persistent large tense cyst in the left lobe of liver, although daughter cyst(s) or ruptured membrane(s) secondary to intrahepatic rupture/communication of liver hydatid cyst do occur rarely and has been recently reported in 2 out of 391 patients (0.51%) by Bedioui et al., although these authors did not mention their clinical presentation [10].

The CT scan may not be very accurate as was observed in our patient, i.e., CT did not reveal the dilated intrahepatic bile ducts which were confirmed on high definition ultrasound, and a few CT slices showed the cyst appearing to arise from the pancreas, that was proved wrong on the operation table.

Although fine-needle aspiration cytology in diagnosis of echinococcosis is documented as a safe procedure in literature, that can demonstrate scoleces, hooklets or laminated membrane [3, 10–13], but it is not commonly used in the suspected cases as has happened in the present case. If it had been employed in our patient, the residual ectocyst might have resolved after 1–2 ultrasound-guided percutaneous aspirations and the patient might have been saved off a major operation.

CONCLUSION

Surgical treatment of the liver hydatid requires longer follow-up of more than six months for detection of the residual or recurrent cyst. The computed tomography scan is a valuable tool for abdominal hydatid cysts but is not always reliable. High definition ultrasound by an expert ultrasonologist is valuable in the difficult situations and is highly recommended when available. Fine-needle aspiration under image guidance appears as a reasonable

option in the recurrent liver cyst following primary surgical treatment before embarking on re-laparotomy.

Author Contributions

Maulana M. Ansari – Conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article, Critical revision of the article, Final approval of the version to be published

Shahla Haleem – Conception and design, Critical revision of the article, Final approval of the version to be published

Wasif M. Ali – Acquisition of data, Drafting the article, Final approval of the version to be published

Leonard J. Enzung – Acquisition of data, Drafting the article, Final approval of the version to be published

Sheikh Sarfraz Ali – Acquisition of data, Drafting the article, Final approval of the version to be published

Sunder K. Meet – Acquisition of data, Drafting the article, Final approval of the version to be published

Guarantor

The corresponding author is the guarantor of submission.

Conflict of Interest

Authors declare no conflict of interest.

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Article citation: Ansari MM, Haleem S, Ali WM, Enzeung L, Ali SS, Meet SK. Obstructive jaundice secondary to postsurgical persistent residual hydatid ectocyst of left lobe of liver. *Int J Case Rep Images* 2014;5(12):844–848.



Maulana Mohammed Ansari is Senior Faculty member with thrust areas in advanced laparoscopy and laparoscopic anatomy especially during TEP hernioplasty. He is credited with 112 published papers, 67 working papers, and 51 conferences/workshop. He coined nine new medical terms, described 31 rare lesions/conditions, successfully used 10 atypical treatments, designed two new surgical treatments and elucidated one surgical anatomy. Email: mma_amu@gmail.com; mmansari.amu@gmail.com



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CASE REPORT

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The management of a recurrent lymphocele following a brachiobasilic fistula superficialization

Ahmed Mohamed Elhassan Elfaki Osman,
Saif Eldin Mohammed Ali Ibrahim

ABSTRACT

Introduction: A lymphocele is defined as a collection of lymphatic fluid in a space within the body not bordered by epithelial linings. They usually occur following surgeries due to iatrogenic disruption of the lymphatic vessels or following an injury in which there is destruction of the lymphatic vessels. Lymphoceles frequently arise following extensive pelvic surgeries, especially gynecological oncological surgeries, and renal transplant surgery. Other surgeries associated with lymphocele formation include open abdominal aortic aneurysm repair, mediastinal and peripheral vascular surgery. **Case Report:** A 65-year-old male with end-stage renal disease (ESRD), presented with an upper arm lymphocele one month after basilic vein superficialization with no other complaints. On examination, all upper extremity pulses were intact (2+) and the arteriovenous fistula had a positive thrill. The swelling was cystic, measuring about 10x20 cm. On greyscale ultrasound, the swelling appeared hypochoic.

Doppler ultrasonography confirmed the patency of the fistula. His management included lymphatic fluid aspiration and povidone iodine sclerotherapy sessions. This regimen was carried out for four consecutive weeks; the same amount of lymphatic fluid was aspirated weekly, indicating the inadequacy of the procedure. On the fifth session, after aspirating the same amount of lymphatic fluid and sclerotherapy, external pressure was applied through a gauze stitched between two skin folds and was left in place for five days. There was no recurrence of the lymphocele after stitch removal. Therefore, we are reporting this case because we strongly believe that aspiration of lymphatic fluid with sclerotherapy complemented by fixed external pressure provided a definitive treatment for a recurrent upper-arm lymphocele. **Conclusion:** Complications arising after peripheral vascular surgeries are vast. Lymphoceles occur less frequently than thrombosis or aneurysms. However, the diagnosis should be kept in mind in any patient presenting with a swelling following recent vascular surgery. With regards to the studies conducted on management of postoperative lymphoceles, some authors advocate the approach of consecutive drain-and-sclerotize sessions while others support more conservative methods such as immobilization and pressure dressings.

Keywords: Basilic vein superficialization, Lymphocele, External pressure, Brachiobasilic fistula, Arteriovenous fistula

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Received: 14 August 2014
Accepted: 01 September 2014
Published: 01 December 2014

How to cite this article

Osman AMEE, Ibrahim SMA. The management of a recurrent lymphocele following a brachiobasilic fistula superficialization. Int J Case Rep Images 2014;5(12):849–853.

doi:10.5348/ijcri-2014147-CR-10458

INTRODUCTION

A lymphocele is defined as a collection of lymphatic fluid in a space within the body not bordered by epithelial linings. They usually occur following surgeries due to iatrogenic disruption of the lymphatic vessels [1, 2] or following an injury in which there is destruction of the lymphatic vessels [2]. Lymphoceles frequently arise following extensive pelvic surgeries, especially gynecological oncological surgeries, and renal transplant surgery. Other surgeries associated with lymphocele formation include open abdominal aortic aneurysm repair [3], mediastinal and peripheral vascular surgery.

CASE REPORT

A 65-year-old male with end-stage renal disease for eight months, presented one month after a right basilic vein superficialization (Figure 1) with a swollen upper arm (Figure 2). The swelling started two weeks after the operation and grew gradually over the course of two to three weeks. He did not complain of any pressure symptoms, nor was he experiencing pain distal to the swelling. The arteriovenous fistula fistula was intact.

On examination, the swelling was well confined, cystic, non-tender, located on the medial aspect of the right upper arm and was not attached to overlying skin. It measured about 10x20 cm. The patient's radial and ulnar pulses were intact and there were no signs of ischemia. A thrill was felt and a bruit was heard on the brachio basilic fistula which lied on the superior border of the swelling.

On greyscale ultrasonography the swelling represented a hypoechoic region (Figure 3). A Doppler ultrasound was performed, thus confirming the patency of the fistula.

In view of the above description, the differential diagnoses we put in mind were hematoma, lymphocele or a seroma. On aspiration of the swelling, the fluid was straw-colored and cytology confirmed features of lymphocele.

The patient was planned for weekly sessions of lymphatic fluid aspiration and sclerotherapy. This continued for four consecutive weeks. On the first session, a total of 130 mL of straw-colored fluid was drained (Figure 4), 7 cm³ of diluted povidone iodine were injected and mild pressure was applied with a bandage. On the subsequent sessions, the same amount of fluid was aspirated, indicating inadequacy of the procedure, and povidone iodine was used as the sclerotherapeutic agent. On the fifth session, following aspiration and sclerotherapy, a gauze was fixed externally between two skin folds using two simple interrupted sutures to apply high pressure on the cavity and was left in place for five days (Figure 5). After removal of the gauze, the



Figure 1: Basilic vein superficialization postoperative scar on the upper border of the lymphocele.



Figure 2: Upper arm postoperative lymphocele.



Figure 3: Greyscale ultrasound denoting the hypoechoic area of the swelling (arrow).

lymphocele has resolved completely (Figure 6) and on serial follow-up sessions there was no recurrence and the arteriovenous fistula was functioning well.

DISCUSSION

Lymphoceles commonly arise as a complication of surgeries, where the normal lymphatic vessels are dissected in the procedure or as a consequence following trauma. They regularly appear following major pelvic surgeries, gynecological oncological surgeries and renal transplantation. Other surgeries may also witness lymphocele formation, as open abdominal aortic aneurysm repair. Clinically, lymphoceles, hematomas and seromas may have similar presentations. Aspiration and cytology helps in confirming or excluding a diagnosis. Lymphoscintigraphy (radiological) helps visualize the course of the lymphatic vessels.



Figure 6: (A) Comparative view between the pre-procedural lymphocele (arrow), (B) The postprocedural resolution (arrow).



Figure 4: Aspirated lymphatic fluid.



Figure 5: External pressure applied following lymphatic fluid aspiration and sclerotherapy.

Regarding the management of postoperative lymphoceles by sclerotherapy, Mahrer et al. conducted a study with 38 patients, the success of sclerotherapy treatment was observed in 33 patients. The number of sclerotherapy sessions ranged from 1–14, with an average of four sessions. They stated that the patients with a lower amount of fluid aspirated in the initial drainage (206 mL) had a higher chance of success, while those with a higher initial aspirated volume (1,708 mL) were the group that failed to respond to treatment. They concluded that the lymphocele cavity size is directly related to the success of sclerotherapy treatment [4]. In contrast to the presentation of this case report, the amount of fluid aspirated was 130 mL and the patient has received four sclerotherapeutic sessions with recurrences of the same amount of fluid in the cavity of the lymphocele in all sessions. Resolution of the lymphocele only occurred after application of the external pressure.

In another study, treatment of a recurrent postoperative lymphocele was performed by drainage and alcohol ablation without complications, in one out of 13 patients. The rest were successfully treated via

drainage and sclerosis [5]. Porcellini et al. conducted a study which evaluated the management of postoperative lymphocele following lower limb arterial reconstructive surgery, in a conservative outpatient fashion. The patients were managed by ambulation limitation, elevation of the limbs and pressure dressings without serial aspirations. All the lymphoceles have resolved in an average period of 21 days [6].

In a study done by Weinberger et al., they discussed the management of postoperative lymphocele after gynecological surgeries. They stated that the methods prevailing for management tend to be the less invasive ones like sclerotherapy and catheter drainage, followed by surgery (laparotomy vs laparoscopy) for recurring lymphocele [7]. The use of isosulfan blue dye was also described in identifying and ligating a forearm lymphocele without prior drainage or sclerotherapy, which appeared as a complication of an interosseous artery pseudoaneurysm repair [8].

Fabrizio Sansone et al. described a technique in which they sutured pledgets over the skin on the groin to apply constant pressure to groin lymphocele that occurred as a complication of major cardiovascular and vascular surgery. Their study included 10 patients and the length of time required for resolution of the lymphocele spanned from 10 to 20 days [9]. The technique described here is almost similar to what we have done, focusing mainly on pressure applied externally to avoid filling of the lymphocele cavity.

CONCLUSION

The complications arising after peripheral vascular surgery are vast, some are common and others are rare, as is the case with postoperative lymphocele. Nonetheless, the diagnosis should be kept in mind in any patient presenting with a swelling arising as a complication of recent vascular, or any major surgery.

Many studies have been conducted regarding the treatment of postoperative lymphocele, some authors advocate the implication of consecutive drain and sclerotize sessions while others use a more conservative approach using immobilization and pressure dressings. After our implication of this method for application of external pressure, we think it is a more effective way of resolving postoperative lymphocele from the first session, i.e., drainage, sclerotherapy and external pressure. Larger studies have to be conducted in order to properly assess the advantage of this method over conventional ways and to determine the prevalence of any complications that may arise.

Author Contributions

Ahmed Mohamed Elhassan Elfaki Osman – Acquisition of data, Drafting the article, Final approval of the version to be published

Saif-Eldin Mohammed Ali Ibrahim – Substantial contributions to conception and design, Revising it critically for important intellectual content, Final approval of the version to be published

Guarantor

The corresponding author is the guarantor of submission.

Conflict of Interest

Authors declare no conflict of interest.

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Article citation: Osman AMEE, Ibrahim SMA. The management of a recurrent lymphocele following a brachiobasilar fistula superficialization. Int J Case Rep Images 2014;5(12):849–853.



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CASE REPORT

OPEN ACCESS

Proper diagnosis and treatment of renal abscess: A case report

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Wei-Jen Yao, Ming-Jui Wu

ABSTRACT

Introduction: Diagnosis and proper treatment of renal abscesses remain a challenge for physicians. Reports have illustrated that small renal abscesses could be effectively treated with a course of intravenous antibiotics. However, delay in diagnosis and treatment could lead to higher morbidity and mortality. **Case Report:** We present a 43-year-old female with a small renal abscess after incomplete treatment of acute pyelonephritis, which was associated with renal stone and *Escherichia coli* bacteremia. Patient was then treated with enough intravenous antibiotics without any classical surgical drainage, and came out to be fully healthy. **Conclusion:** This case highlights

the need for early identification of risk factors as well as the subtle feature of renal abscess by proper diagnosis and adequate treatment.

Keywords: Acute pyelonephritis, Antibiotics, Renal abscess, Renal stone

How to cite this article

Hao Lyh-Jyh, Wang Ray-Shyang, Chen Chien-Ta, Wu Shao-Wen, Yao Wei-Jen, Wu Ming-Jui. Proper diagnosis and treatment of renal abscess: A case report. Int J Case Rep Images 2014;5(12):854–858.

doi:10.5348/ijcri-2014148-CR-10459

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Received: 25 September 2014
Accepted: 17 October 2014
Published: 01 December 2014

INTRODUCTION

Renal and perirenal abscesses are uncommon diseases originated mainly from infections in or around the kidney. The former one is accounted for around 0.02% and the latter case is for about 0.2% of hospital admissions in Altemeier's series of 540 intra-abdominal abscesses [1]. A delay in renal abscess diagnosis may lead to higher morbidity and mortality, which has been reduced to 12% since the accessibility of computed tomography (CT) scan and magnetic resonance imaging (MRI) scan [2, 3]. Classical treatment for renal abscesses include surgical exploration, incision and drainage, or nephrectomy [4, 5]. In fact, simply invasive treatment appeared in early 1970s, and the trend towards conservative treatment is frequent due to the progress in imaging techniques and new antibiotics. Small renal abscesses could be effectively treated with the sufficient drainage and a course of intravenous antibiotics in the previous reports [4, 6–8]. Herein, we report a small renal abscess after incomplete treatment of acute pyelonephritis, which was completely restored to health by adequate antibiotic treatment.

CASE REPORT

A 43-year-old female, with left renal stone, presented to our hospital after two days of fever and left flank pain. The initial evaluation revealed high fever (body temperature 39.7°C), tachycardia (heart rate, 112 beats per minute), leukocytosis (white blood cell, 24,560 per micrometer) with a left shift of elevated C-reactive protein (18.98 mg/dL), and left flank tenderness, but no thrombocytopenia. There was no diabetes mellitus history of this female. However, bilateral calyceal renal stones and relative swelling of left kidney were noted on abdomen sonography. Moreover, the blood cultures yielded *Escherichia coli* and hematuria without pyuria were noted. Thus, left side acute pyelonephritis was impressed and intravenous antibiotics with cefazolin 1 g q8h and gentamycin 80 mg q12h were prescribed for 5 days. The patient requested discharge due to family problem when the fever was subsided for two days with follow-up white blood cell count 11690/ μm and C-reactive protein 6.11 mg/dL.

Oral ciprofloxacin (250 mg tablet twice per day) was prescribed to her after discharge and she was informed to follow-up at our outpatient department (OPD) one week later. Unfortunately, chillness, low grade fever, left flank pain, leukocytosis (white blood cell, 15620/ μm) and high C-reactive protein (13.39 mg/dL) recurred three days later after discharge. She was re-admitted to ward via OPD and followed-up abdomen sonography showing a 2.16 cm heterogenous hypoechoic nodule in lower pole of the left kidney, favor renal abscess (Figure 1). After admission, medical treatment without therapeutic drainage was suggested by infectious disease specialist and urologist. Intravenous antibiotics with ciprofloxacin 400 mg q12h and amikacin 250 mg q12h were prescribed for two weeks. Fever subsided and mild local left flank area knocking pain was noted. Normal white blood cell count (8540/ μm) and mild elevated C-reactive protein (1.09 mg/dL) were noted. Follow-up abdomen computed tomographic scan revealed a 1-cm abscess in lower pole of the left kidney with focal perirenal fatty blurring, indicating that partial resolution of the left renal abscess was considered (Figure 2). She was then discharged and oral ciprofloxacin (250 mg tablet twice per day) was continuously prescribed for four more weeks at OPD, and follow-up abdomen sonography revealed less than 8 mm renal stone without any more abscess (Figure 3). Extracorporeal shock wave lithotripsy (ESWL) of left renal stone was suggested by the urologist, but the patient refused. She was instructed to drink eight glasses of fluid daily to maintain adequate hydration and to decrease the chance of urinary supersaturation with stone-forming salts. Other dietary guidelines were suggested to avoid excessive salt and protein intake and moderation of calcium and oxalate intake. There was no more pyelonephritis or renal abscess recurrence of this patient three years later after discharge.



Figure 1: Sonography illustrating the 2.16 cm heterogenous hypoechoic nodule in lower pole of the left kidney in the female patient. Area surrounded by arrowheads and yellow cross signs indicating the renal abscess.



Figure 2: Follow-up abdomen computed tomography scan revealed the 1 cm abscess in lower pole of the left kidney with focal perirenal fatty blurring after intravenous antibiotics with ciprofloxacin and unikin treatments for two weeks in the female patient. Area pointed by arrowheads indicating the reduced renal abscess.

DISCUSSION

The diagnosis of perinephric or renal abscess, as well as splenic abscess, is frequently delayed, and the mortality rate in some cases is extensive. Thus, perinephric and renal abscesses should be seriously taken care when a patient presents with symptoms of pyelonephritis and remains feverish after four or five days of treatment [1]. Besides, diagnoses should be entertained when a urine



Figure 3: Oral ciprofloxacin was continuously prescribed for four more weeks, and follow-up abdomen sonography revealed renal stones (less than 8 mm) without any more abscess in lower pole of the left kidney. Pyelonephritis recurrence of the patient did not occur after discharge.

culture yields a polymicrobial flora; a patient is known to have renal stones; or fever and pyuria coexist with a sterile urine culture. Meanwhile, renal ultrasonography and abdominal CT should be exploited to confirm the authentic cause.

Report has suggested an algorithmic approach to manage renal abscesses, which illustrated that main management with antibiotics was recommended in <3 cm in diameter small abscesses, and drainage (percutaneous or surgical) was recommended in >5 cm large abscesses. Both approaches could be applied in medium-sized abscesses (3–5 cm) [6]. Another report further suggested avoiding the aggressive treatment on renal and perinephric abscesses with 5 cm in diameter or less, which could have complete decrease after antibiotic therapy [9]. However, study also illustrated that aggressive drainage is suitable in abscesses >3 cm [6]. In fact, additional study has demonstrated that percutaneous abscess drainage might have several complications [10].

The total duration of antibiotic treatment course is dependent on the patient's clinical response. The current recommendations are to continue parenteral antimicrobial therapy for at least one to two days after clinical improvement, and oral antibiotic therapy can then be administered for an additional two weeks [11]. In previous several studies, renal stones and urinary obstruction have been reported as common predisposed conditions with an incidence of 24–54% and 21–50% of renal abscess, respectively [12, 13]. More than 75% of perinephric and renal abscesses arise from a urinary tract infection, which ascends from the bladder to the kidney with pyelonephritis occurring prior to abscess development [1]. *E. coli*, *Proteus* spp., and *Klebsiella* spp. are the organisms most frequently encountered in perinephric and renal abscesses [1]. Patients with renal

abscesses have a higher rate of *E. coli* infection in urine cultures, and a female predominance (91.8%) could be observed [4]. This may be a result of the development of renal abscesses via an ascending infection by organisms already isolated within the urinary tract [7].

The intravenous antimicrobial therapy may be a good alternative treatment if therapeutic drainage is believed to have considerable risk. Large abscesses, obstructive uropathy, severe vesicoureteral reflux, diabetes, old age, and urosepsis with gas forming organisms are the factors associated with antimicrobial treatment failure [2]. Percutaneous nephrostomy should be considered when there is a large abscess or obstructive uropathy, and no clinical improvement occurs after 48 to 72 hours of appropriate antibiotic therapy [2]. An incision and drainage is preferred when the open drainage is required. Nephrectomy is reserved for patients whose renal parenchyma is diffusely damaged and for elderly patients whose survival depends upon urgent surgical intervention [7].

Renal stone is an important risk factor for our case and incomplete intravenous antibiotics treatment course of acute pyelonephritis resulted in the renal abscess formation. In fact, medium-sized as well as small-sized renal abscesses can be treated successfully with adequate IV antibiotics without surgical drainage [4]. Empirical therapy with broad-spectrum antibiotics (ampicillin or vancomycin in combination with an aminoglycoside or third-generation cephalosporin or a fluoroquinolone) is usually recommended because it is often very difficult to identify the correct causative organisms from the urine or blood. Percutaneous drainage under CT or ultrasound guidance is indicated if the patient does not respond within 48 hours of treatment [6]. The drained fluid should be cultured for the causative organisms. The total duration of the treatment was conditioned by the clinical response and is about one to two months in most patients. The healing of the abscess assessment criteria include absence of pain, reduction of fever, normalization of ESR or CRP, disappearance of the abscess on ultrasound or CT scan which usually reveals a cortical scar. The best indicator of healing is the absence of recurrence of clinical signs and infection symptoms. If the clinical and laboratory parameters come within normal limits, then the antibiotic treatment can be stopped 10 days later. The patient must be followed up over an interval of two weeks, two or three months after the end of the treatment [14]. Asymptomatic renal stones may be followed conservatively. However, patients can be advised that about 50% of small renal calculi become symptomatic within five years [15]. Some surgical procedures may be required for larger stones (i.e., ≥ 7 mm) that are unlikely to pass spontaneously. In some cases, hospitalizing a patient with a large stone to facilitate surgical stone intervention is reasonable. However, acute renal colic mostly can be treated on an ambulatory care [16]. General treatment of renal stones is with hydration to increase urine output and with analgesia. Renal calculi less than

2 cm in size can generally be treated with extra corporeal shock wave lithotripsy (ESWL) [17]. After passage of the stones, treatment is directed at prevention of recurrent stones formation. The foundation of renal stones therapy is maintenance of high urine output (2–3 L/day) with oral hydration and a low-salt diet (<2 g/day) [18].

CONCLUSION

This case report is an example of a small renal abscess after incomplete treatment of acute pyelonephritis which was associated with renal stone and *E. coli* bacteremia. Several reports observed that small renal abscesses were effectively treated with a course of intravenous antibiotics. The total duration of the treatment was conditioned by the clinical response. This case highlights the need for early identification of risk factors as well as the subtle features of renal abscess for appropriate diagnosis and adequate treatment.

Author Contributions

Lyh-Jyh Hao – Substantial contributions to conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published

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Shao-Wen Wu – Acquisition of data, Analysis and interpretation of data, Drafting the article, Final approval of the version to be published

Wei-Jen Yao – Acquisition of data, Analysis and interpretation of data, Drafting the article, Final approval of the version to be published

Ming-Jui Wu – Substantial contributions to conception and design, Acquisition of data, Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published

Guarantor

The corresponding author is the guarantor of submission.

Conflict of Interest

Authors declare no conflict of interest.

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CASE REPORT

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Isolated neurocutaneous peripheral T cell lymphoma, NOS

Talal Hilal

ABSTRACT

Introduction: Peripheral T cell lymphomas (PTCLs) are a heterogeneous group of diseases that are a relatively uncommon subtype of non-Hodgkin's lymphoma (NHL) with an overall poor prognosis. They are difficult to classify and targeted therapy does not exist. Patients are usually treated with B cell specific drugs that have not shown to be effective in most subtypes. **Case Report:** A 52-year-old male presented with a one-week history of confusion. He was found to have diffuse subcutaneous lumps on his extremities, back, and thorax, and a large swelling over his left face. Imaging revealed a right frontal lobe mass. Biopsy of the mass confirmed PTCL. Further histopathological analysis and imaging for staging diagnosed the subtype as not otherwise specified (NOS). The patient received radiation therapy to the brain followed by systemic chemotherapy with cyclophosphamide, doxorubicin, vincristine, prednisone (CHOP). He responded by the end of the first cycle, but his long-term clinical course remains to be seen. **Conclusion:** Treatment for PTCL is a realm requiring further research with efforts focusing on the development of T cell specific drugs.

Keywords: B-cell lymphoma, Lymphoma, Non-Hodgkin lymphoma, Peripheral T cell lymphoma

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Received: 13 August 2014

Accepted: 02 September 2014

Published: 01 December 2014

How to cite this article

Hilal T. Isolated neurocutaneous peripheral T cell lymphoma NOS. Int J Case Rep Images 2014;5(12):859–863.

doi:10.5348/ijcri-2014149-CR-10460

INTRODUCTION

Peripheral T cell lymphomas (PTCLs) derive from post-thymic T cells and generally arise in lymphoid tissue “peripheral” to the thymus such lymph nodes, spleen, gastrointestinal tract and skin. They have a mature T cell phenotype and are grouped, along with mature natural killer lymphomas, according to clinical presentation as leukemic, nodal or extranodal [1]. This case describes an atypical presentation of PTCL, not otherwise specified (NOS), and reviews the classification and current treatment options for this uncommon disease.

CASE REPORT

A 52-year-old male presented to the emergency department after being found confused at home. His family relayed that he had developed progressive left facial swelling and right scalp swelling over the past three months and had been becoming episodically confused over the past week. His past history was significant for poorly controlled type 2 diabetes mellitus, alcohol abuse and excision of a meningioma 15 years ago. On examination the patient was afebrile, his blood pressure 119/74 mmHg, heart rate 77 beats per minute, respiratory rate 18 breaths per minutes and his oxygen saturation was 97% on room air. He was drowsy and had delayed responses to questions. He had facial asymmetry with left sided swelling and facial droop. His scalp on the right side over the parietal area was swollen and mildly

tender. Inspection of his oral cavity revealed a necrotic growth protruding from his soft palate. He had multiple diffuse violaceous lesions on his skin, some of which were ulcerated with central necrosis (Figure 1). He also had multiple firm, mildly tender nodules present on left forearm, right upper chest and several smaller nodules on upper back.

Computed tomography (CT) scan revealed a right frontal mass. Further assessment with magnetic resonance imaging (MRI) of the brain showed a 4x3 cm enhancing mass in the right inferior frontal gyrus with hypercellularity, hemorrhage and surrounding edema (Figure 2A–B). Initial assessment focused on histologic diagnosis and a biopsy of the soft palate growth and one of the skin lesions, both of which revealed features consistent with PTCL (Figure 3). Computed tomography scans of the chest, abdomen and pelvis showed no



Figure 1: Left thigh and intertriginous area with infiltration of lymphoma into skin causing ulceration and sterile discharge.

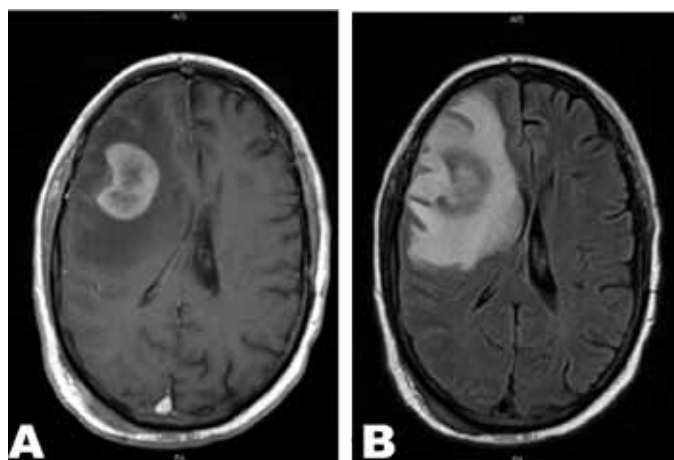


Figure 2: (A) T1-weighted magnetic resonance imaging of the brain depicts right hemispheric mass with midline shift. (B) FLAIR MRI of the brain depicts significant edema around the mass.

lymphadenopathy or evidence of primary neoplasm. Biopsy results of the brain mass were consistent with the same finding of PTCL (Figure 4A). Bone marrow biopsy and aspirate showed normal cellularity without involvement of lymphoma.

Based on the pattern of involvement and histologic findings of CD3 (Figure 4B) and CD7 positivity with CD4, CD8, CD30, CD56 negativity, the patient's disease fit the subtype of PTCL, NOS. The patient initially received intravenous dexamethasone and emergent radiation therapy to the brain to decrease mass effect and risk for subfalcine herniation. He received a total of 10 days of radiation. During that period the lesions on his skin were ulcerating, the soft palate growth was causing pain and trouble swallowing and subcutaneous nodules were enlarging. We then decided to stop radiation therapy and start systemic chemotherapy using cyclophosphamide, doxorubicin, vincristine, and prednisone (CHOP). The patient responded within a few days and all his nodules disappeared by the end of the first of four cycles of therapy.

DISCUSSION

PTCLs are a group of lymphomas that fall under the category of mature T cell lymphomas, and are an uncommon subtype of non-Hodgkin's lymphoma (NHL). The International Peripheral T cell and Natural Killer/T cell lymphoma study reported that PTCLs accounted for only 5–10% of all NHL cases in Western countries and about 10–20% in Asian countries [2]. The disease tends to occur in adults with 40% of cases occurring between the ages of 55 and 74, and only about 5% of cases occurring after the age of 85. The incidence rates among all races in males and females are approximately 2.3 and 1.4 per 100,000 individuals, respectively, in contrast to 24 and 16.5 cases per 100,000 males and females for non-Hodgkin's lymphoma. The disease is almost twice as frequent in males than females [3].

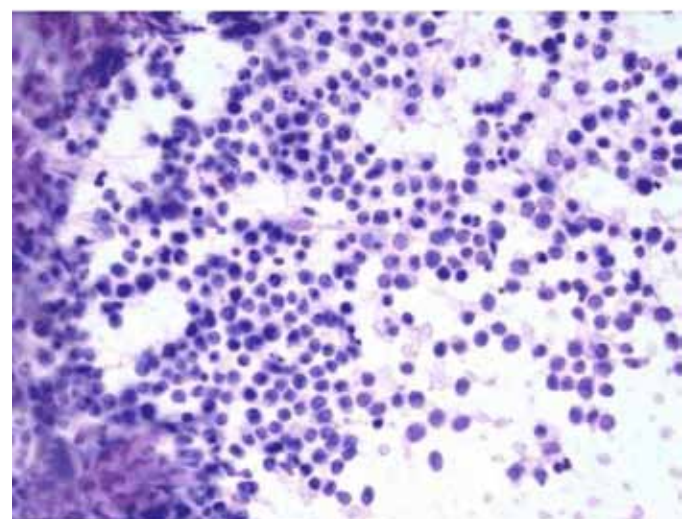


Figure 3: Histology depicts features consistent with mature T cell lymphoma. (H&E stain, x100).

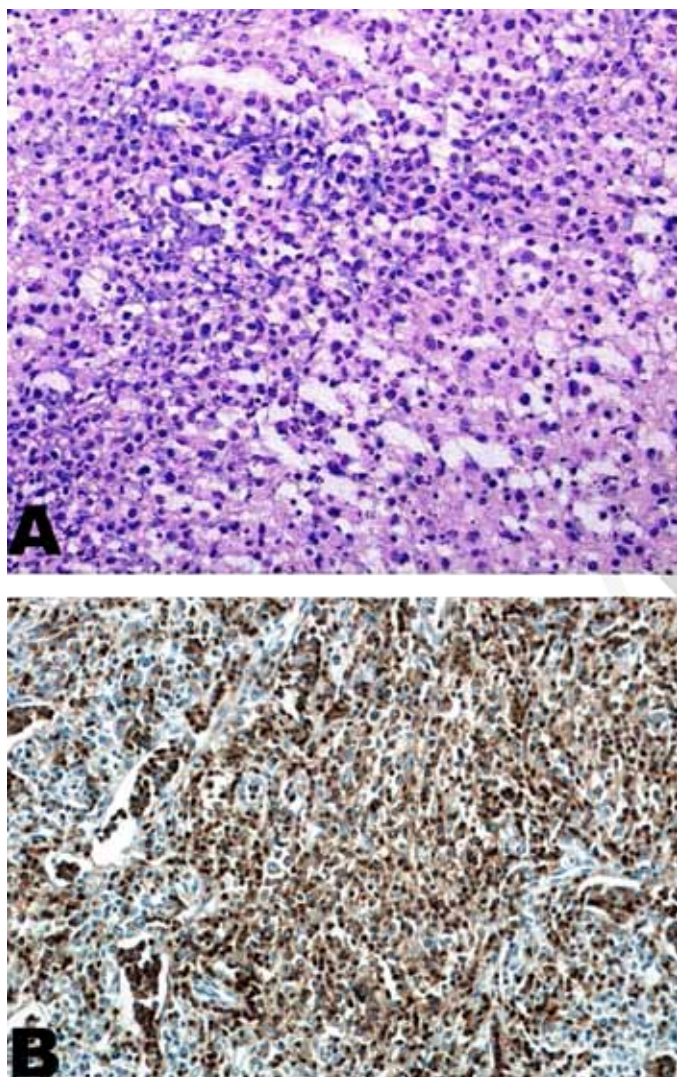


Figure 4: (A) Histology of brain biopsy consistent with mature T cell lymphoma (H&E stain, x100). (B) Immunohistochemistry for CD3 is positive in tumor cells.

The clinical presentation is variable and depends on the specific subtype of PTCL. Overall, patients present with more advanced disease and an increased incidence of B symptoms. Paraneoplastic features that are well described include eosinophilia, hemophagocytic syndrome, and autoimmune phenomena [4]. The incidence of brain metastasis in PTCLs is unknown with very few case reports demonstrating brain involvement at time of diagnosis and no cases that report isolated brain mass as the initial presentation of PTCL, NOS.

Definitive diagnosis is based on examination of tissue biopsy specimen for histologic features supplemented by detailed immunohistochemistry, flow cytometry, cytogenetics, and molecular genetics [5]. Historically, PTCLs have been difficult to classify given the heterogeneity of the condition. The most widely used classification system currently is that of the World Health Organization (WHO) [6] (Table 1).

The difficulty not only lies in diagnosing the subtype of PTCL, but also in treating it. Overall, the different subtypes all have a poor prognosis. The standard of care for all subtypes of PTCL has been combination chemotherapy using CHOP. This has been used based on trials that enrolled patients with diffuse large B cell lymphoma (DLBL) and has been shown to be effective only in the subtype anaplastic lymphoma kinase (ALK)-positive anaplastic large cell lymphoma (ALCL) [7, 8].

The outcomes of CHOP chemotherapy are variable. For PTCL, NOS, the reported complete response (CR) to treatment and overall survival (OS) is 50% and 30%, respectively [4]. Alternative first line therapies are currently under study with the focus being on the addition of a target-directed drug that takes on a role similar to that of rituximab in the treatment of mature B cell lymphomas. The addition of bortezomib, a selective proteasome 26S inhibitor, to CHOP has shown promising initial response rates in a phase II trial. The overall survival, however,

Table 1: Mature T and natural killer neoplasms as classified by the World Health Organization (WHO) 2008

Clinical Categories		Major Disorders		
Leukemic	T cell prolymphocytic leukemia (T-PLL)	T cell large granular lymphocytic leukemia (T-LGL)	Adult T cell leukemia/lymphoma	NK cell-related
Nodal	Peripheral T cell lymphoma, not otherwise specified (PTCL, NOS)	Angioimmunoblastic T cell lymphoma (AITL)	Anaplastic large cell lymphoma (ALCL), ALK positive	Anaplastic large cell lymphoma (ALCL), ALK negative
Extranodal	Extranodal NK/T cell lymphoma, nasal type	Enteropathy-associated T cell lymphoma (EATL)	Hepatosplenic T cell lymphoma (HSTL)	Subcutaneous panniculitis-like T cell lymphoma (SPLTL) – αβ only
Cutaneous	Mycosis fungoides		Sezary syndrome	

was unchanged compared to CHOP alone [9]. Another agent, alemtuzumab, a anti-CD52 monoclonal antibody is currently being studied in phase III trials as an add-on to CHOP. The results have yet to be published.

Another aspect of management is hematopoietic stem cell transplant (HSCT). Several studies have shown that first line high-dose chemotherapy with autologous HSCT had complete response rates ranging between 60–70% with the best studies showing a three-year survival ranging between 63–73%. Difficulties arise with toxicity and inability of patients to proceed with transplantation [10, 11].

Relapse is common with PTCLs and options at that point include salvage chemotherapy, autologous HSCT or allogeneic HSCT. Agents approved by the FDA for relapsed/refractory PTCL are pralatrexate, a folate analog, romidepsin, a histone deacetylase inhibitor and brentuximab vedotin (BV) used for ALCL. The first two showed response rates between 25–30%, a complete response between 10–15%. The overall survival for pralatrexate was 14 months and no overall survival was reported for romidepsin [12]. Autologous HSCT for relapsed disease showed poor results with a five-year overall survival <35% in most studies. Reduced intensity chemotherapy (RIC) followed by allogeneic HSCT shows promising results with problems arising from acute and chronic graft versus host disease (GvHD). Further prospective trials are warranted to address the role of RIC and allogeneic HSCT in PTCL, NOS [4].

CONCLUSION

Our understanding of the subcategory of peripheral T cell lymphomas (PTCLs) has improved over the years. What once was treated as a variant of mature B cell lymphomas is now seen under a different light. It is clear that patients who suffer from any of the PTCLs do worse than their mature B cell lymphoma counterparts. This understanding has led to further research into the molecular mechanisms that mediate disease. Clinical presentation is variable and histologic diagnosis is essential. Current efforts are focused on developing novel agents to target specific mutations that can alter the natural history of this poorly understood entity.

Acknowledgements

Mr. Frank Davis for providing support with a concise literature review on the topic. Dr. Melissa Kesler, Dr. Janna Neltner and Zane Staubach, from the Department of Pathology their valuable help with the pathology slides.

Author Contributions

Talal Hilal – Acquisition of data, Analysis and interpretation of data, Drafting the article, Final approval of the version to be published

Guarantor

The corresponding author is the guarantor of submission.

Conflict of Interest

Authors declare no conflict of interest.

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Article citation: Hilal T. Isolated neurocutaneous peripheral T cell lymphoma NOS. Int J Case Rep Images 2014;5(12):859–863.



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Thyrotoxicosis: An unusual presentation

Somnath Goptu, Gurjit Singh, Iqbal Ali, Siddharth Mishra

ABSTRACT

Introduction: In a thyrotoxic patient, Grave's disease, solitary toxic nodule and toxic nodular goitre are considered to be the possible diagnosis. However, in certain inflammatory conditions also present with similar features wherein thyroid hormones are released due to destruction of the gland. Such conditions as Hashimoto's thyroiditis may be missed on cytological examination and are diagnosed only on histopathological examination. The combination of Hashimoto's thyroiditis with toxicity is called hashitoxicosis. **Case Report:** A 37-year-old female presented with thyroid swelling and features of thyrotoxicosis which were confirmed by thyroid function tests. A clinical diagnosis of toxic nodular goitre was made. Euthyroid state was achieved after treatment with tab carbimazole and beta blockers. The patient underwent near total thyroidectomy. After surgery, patient developed hypocalcemia which was managed by intravenous and oral calcium supplementation. At postoperative estimation thyroid auto antibodies were found elevated and the patient was started on tab thyroxine. At sixth month follow-up serum calcium levels

returned to normal levels and hence calcium supplementation was stopped. **Conclusion:** Any patient presenting in thyrotoxic state, possibility of hashitoxicosis should be considered and confirmed by the estimation of thyroid antibodies and the use of ultrasonography guided fine-needle aspiration cytology.

Keywords: Grave's disease, Hashitoxicosis, Thyroid antibodies, Thyrotoxicosis

How to cite this article

Goptu S, Singh G, Ali I, Mishra S. Thyrotoxicosis: An unusual presentation. Int J Case Rep Images 2014;5(12):864–867.

doi:10.5348/ijcri-2014150-CR-10461

INTRODUCTION

In a patient of thyrotoxicosis possible etiologies considered are Grave's disease, toxic multi-nodular goitre and solitary toxic nodule. However, certain inflammatory conditions may lead to destruction and damage to thyroid gland resulting in leakage of hormones causing transient thyrotoxicosis. Such transient thyrotoxicosis in association with Hashimoto's thyroiditis is termed hashitoxicosis. Patients with this disorder are expected to remit or even develop hypothyroidism. Only correlation of histopathology with antibody testing may clinch the diagnosis.

CASE REPORT

A 37-year-old female presented with complaints of thyroid swelling for the last three months which was associated with tachycardia and positive Stellwag's sign with no previous history of thyrotoxicosis. Thyroid

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Received: 10 July 2014
Accepted: 16 August 2014
Published: 01 December 2014

function tests revealed normal levels of T₃ (2 nmol/L) and T₄ (14.8 nmol/L) but decreased levels of TSH (0.011 mU/L) hence diagnosis of thyrotoxicosis was made. Ultrasound of the neck revealed multinodular goitre. Fine-needle aspiration cytology (FNAC) was suggestive of colloid goitre (Figure 1).

The patient was started on antithyroid drugs and beta blockers. Euthyroid state was achieved in four weeks. Patient underwent near total thyroidectomy. Within 24 hours following surgery patient developed features of hypocalcemia. She had multiple episodes of carpopedal spasms over a period of next 40 days which were managed by I.V. injection calcium gluconate, and oral calcium and vitamin D₃. She was gradually weaned off the injection, and only oral calcium and vitamin D₃ supplementation were continued. Histopathology was suggestive of Hashimoto's thyroiditis (Figure 2).

The antithyroid antibody levels were estimated following surgery. Antimicrosomal and antithyroglobulin levels were high. The patient was started on thyroxine 50 mg supplementation and oral vitamin D supplementation was continued.

Thyroid function tests and serum calcium levels were within normal limits at six months following surgery, hence oral calcium was stopped. Thyroxine 50 mg had been continued.

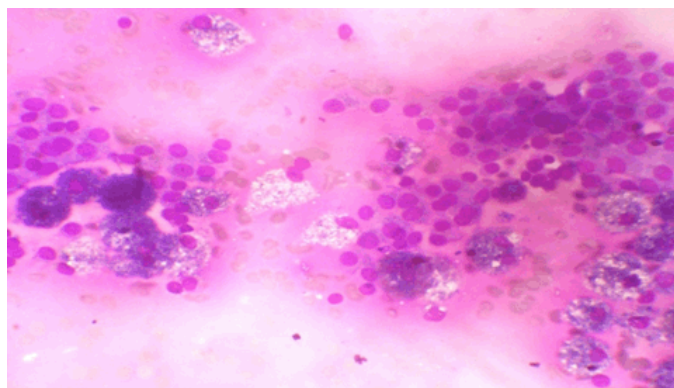


Figure 1: Fine-needle aspiration cytology suggestive of colloid goitre.

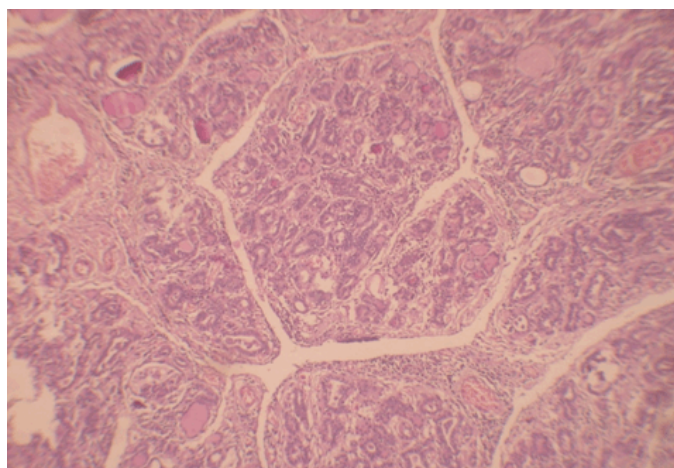


Figure 2: Histopathology showing presence of lymphocytes along with colloid goitre.

There has been no episodes of hypocalcemic attacks during ensuing three months following stoppage of oral calcium supplementation and serum calcium levels have remained within normal limits.

DISCUSSION

Grave's disease and toxic nodular goitre are always considered as the prime etiologies in a patient with thyrotoxicosis. However, certain destructive inflammatory conditions may damage the thyroid gland and cause the classical "leakage" of hormones into the blood resulting in transient thyrotoxicosis [1]. Such phenomenon can occur in Hashimoto's thyroiditis and called hashitoxicosis.

Hashitoxicosis has an incidence of 4.47%. Out of 69 patients with autoimmune thyroiditis studied by Nabhan et.al. only eight (11.69%) were diagnosed with hashitoxicosis [2]. Normal course of such condition is remission in due course of time or it develops into hypothyroidism.

Certain drugs like pegylated interferons $\alpha 2b$ (PEG-IFN α) and ribavirin can produce hashitoxicosis followed by type 1 diabetes [3].

It has multi-factorial etiology which has multiple genetic and environmental factors. Genetic factors include human leucocyte antigen, major histocompatibility complex and cytotoxic T lymphocyte association (CTLA) [4]. Environmental factors include infections, cytokine therapy, selenium, iodine uptake, smoking [5].

It is associated with other autoimmune diseases which includes type 1 diabetes mellitus, systemic lupus erythematosus, multiple sclerosis, rheumatoid arthritis, celiac disease, vitiligo, chronic urticaria [6]. However, our patient did not suffer from any other autoimmune diseases.

In hashitoxicosis, there is a loss of immune tolerance to the thyroid cells due to the production of autoantibodies which in turn leads to destruction of the gland. Hence, the patient presents in hyperthyroid state which is followed by a definite resolution or may go to hypothyroidism. There is a destruction of both follicular and the 'C' cells which are replaced by fibrosis [7].

Patients suffering from hashitoxicosis usually present initially with features of hyperthyroidism which later progresses to hypothyroidism [8].

Our patient presented with a short history of nodular goitre and features of toxicity. FNAC revealed colloid material and hence diagnosis of Hashimoto's thyroiditis was never entertained. The diagnosis of Hashimoto's thyroiditis can be missed in smears showing cytological evidence of hyperplasia or abundant colloid [9]. Later being the reason in our case. Therefore, it would be reasonable to advocate ultrasonography guided FNAC or multiple punctures in equivocal cases. Macdonald and Yazdi emphasised importance of accurate cytological interpretations in the diagnosis of Hashimoto's

thyroiditis to avoid false positive and false negative reports [10].

MacDonald and Yezdi emphasised the need for adequate sampling of the thyroid while performing fine-needle aspiration biopsy (FNAB) [11]. In their study of 184 aspirates diagnosed with Hashimoto's thyroiditis, 39 had corresponding surgical specimen taken from 31 patients. Amongst these 12 (31%) FNABs from nine patients, the cytological diagnosis was not confirmed histologically. The diagnosis of Hashimoto's thyroiditis is likely to be missed in smears showing cytological evidence of hyperplasia or abundant colloid [11]. In our case too, diagnosis was missed as smear showed only colloid material.

The investigation of choice in cases of hashitoxicosis is the estimation of auto antibodies, which includes antithyroglobulin and antimicrosomal antibodies [1, 11]. Levels of these antibodies are usually raised. The most sensitive of these is the antimicrosomal antibody. In our case, the estimation was done postoperatively and found to be raised. Hence, it will be reasonable to assume that estimation of thyroid antibodies should form integral part of any investigative protocol for thyroid disorders.

Patients with hashitoxicosis are first managed with β blockers to control toxicity and then started with thyroxine supplementation 50 mg once a day in order to slow the disease process and to reduce the level of auto antibodies [11]. However, patients can still undergo surgical resection when there is a dominant mass or an unresponsive mass despite thyroxine therapy or intermediate findings of malignancy on a cutting needle biopsy [12]. Our patient underwent near total thyroidectomy considering it to be a dominant mass of multi-nodular toxic goitre which was not expected to regress.

The patient has been followed-up every six weeks with estimation of thyroid autoantibodies and TSH level. The TSH levels are to be maintained between 1–10 μ /L. If After six months following surgery and thyroxine therapy, autoantibody levels have remained raised, such patients will have to continue life-long thyroxine.

CONCLUSION

In patients with toxic nodular goiter, possibility of Hashimoto's thyroiditis should be considered and excluded by obtaining representative samples on ultrasonography guided fine-needle aspiration biopsy. Routine estimation of thyroid antibodies should form part of protocol while investigating such patients. There is definite role of surgery under specific clinical settings. Thyroid supplementation will be required for life-long even in those patients who did not undergo any form of surgery.

Author Contributions

Somnath Gooptu – Substantial contributions to conception and design, Analysis and interpretation of

data, Drafting the article, Final approval of the version to be published

Gurjit Singh – Substantial contributions to conception and design, Acquisition of data, Analysis and interpretation of data, Critical revision of the article, Final approval of the version to be published

Iqbal Ali – Acquisition of data, Analysis and interpretation of data, Drafting the article. Final approval of the version to be published

Siddharth Mishra – Acquisition of data, Analysis and interpretation of data, Drafting the article, Final approval of the version to be published

Guarantor

The corresponding author is the guarantor of submission.

Conflict of Interest

Authors declare no conflict of interest.

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Rhinoscleroma: A case report

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ABSTRACT

Introduction: Rhinoscleroma is a chronic, granulomatous infection caused by Gram negative facultative intracellular encapsulated, non-motile bacillus *Klebsiella rhinoscleromatis* that most frequently affects the respiratory mucosa, especially the nasal cavity and eventually extending through the lower respiratory tract. **Case Report:** We report a case of rhinoscleroma in a 32-year-old female presenting with extensive swelling of mid-face encroaching onto the lower two-thirds of the nose, upper lip and oral cavity and nasal blockage with a duration of more than five years. A biopsy was taken for histological diagnosis which confirmed the diagnosis of rhinoscleroma followed by excision of the lesion and reconstruction of the nose and upper lip. For five years, there was no evidence

of recurrence. **Conclusion:** When the disease progresses with proliferation, it may simulate a tumor, as in our patient with typical Hebra nose.

Keywords: Chronic granulomatous infection, Hebra nose, *Klebsiella rhinoscleromatis*, Mikulicz cells, Rhinoscleroma

How to cite this article

Bhagyalakshmi A, Rao CV, Krishna KT, Kartheek BVS. Rhinoscleroma: A case report. Int J Case Rep Images 2014;5(12):868–872.

doi:10.5348/ijcri-201457-CI-10014

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Received: 08 July 2014
Accepted: 31 July 2014
Published: 01 December 2014

INTRODUCTION

Rhinoscleroma or scleroma is a chronic granulomatous disease caused by Gram negative bacillus called *Klebsiella rhinoscleromatis* or Frisch bacillus. Nasal cavity is the most common predilection site and may extends to nasopharynx, oropharynx, larynx, trachea and bronchi. Females are more frequently affected between 10–30 years of age. Rhinoscleroma is mostly endemic in tropical countries [1]. The disease was first described by the dermatologist Ferdinando Von Hebra in 1870 [2]. We present a case of Rhinoscleroma with typical Hebra nose.

CASE REPORT

A 32-year-old female from tribal area of Visakhapatnam district, Andhra Pradesh (India) presented with extensive swelling of five years duration on mid-face occupying the lower two-thirds of the nose, upper lip and oral cavity (Figure 1). The swelling started as a small nodule over the tip of the nose and gradually increased in size. The patient had applied some native treatment on the swelling. The swelling occupied the entire area described. The surface was red and irregular and was bleeding on touch. There

was crust formation all over producing offensive smell mimicking a malignant lesion. She was able to open the mouth and take food as the tongue; lower lip were not involved. The patient was breathing through the oral cavity. It was painless swelling with no regional lymph nodal involvement. The lesion was biopsied twice and the initial histological diagnosis was an inflammatory lesion. As the lesion was large, distorting the facial configuration, radical excision of the swelling and reconstruction of the nose and upper lip using forehead skin was planned.

Accordingly, the patient was operated upon under general anesthesia and the lesion was excised and reconstruction of the nose and upper lip was done using oblique classical Indian forehead flap in stages. On Gross examination the excised lesion was measured 7x7 cm, grayish white on cut section and firm in consistency (Figures 2 and 3). The histopathological examination with hematoxylin and eosin staining (H&E) revealed hyperplasia of surface squamous epithelium with subepithelial sheets of mixed inflammatory cell infiltrates composed of neutrophils, lymphocytes, plasma cells, Mikulicz cells and homogenous eosinophilic round to ovoid bodies (Russel bodies) and areas of fibrosis. (Figures 4–6). Special staining with Gram's stain and periodic acid (PAS) stain revealed small bacilli in Mikulicz cells (Figures 7 and 8). The diagnosis was confirmed to be rhinoscleroma. Postoperative period is uneventful and the patient was discharged (Figure 9). We could follow the case for five years, there was no evidence of recurrence.

DISCUSSION

Rhinoscleroma is a chronic infectious disease caused by *Klebsiella rhinoscleromatis*, a Gram-negative, facultative intracellular, non-motile encapsulated bacillus identified by Von Frisch in 1882 [3]. The nose, pharynx, larynx, trachea, and occasionally also the skin of the upper lip are distorted and infiltrated with hard, granulomatous masses. The disorder always begins in the nose [4]. The nose is affected in 95–100% of cases, the pharynx in 18–43%, larynx in 15–80%, trachea in 12%, and bronchi in 2–7% [5].

The disease evolves through three stages:

- (1) Catarrhal stage: It is characterized by foul smelling purulent nasal discharge and crusting resembling atrophic rhinitis.
- (2) Granulomatous stage: In this painless and nonulcerative granulomatous, nodules form in the nasal mucosa. There is also subdermal infiltration of external nose and upper lip giving a 'woody' feel.
- (3) Cicatricial stage: This causes stenosis of nares, distortion of upper lip, adhesions in the nose, nasopharynx and oropharynx.

There may be subglottic stenosis with respiratory distress [6]. Rhinoscleroma spreads from person-to-person by air-borne transmission. The pathogenesis



Figure 1: Clinical photograph showing nasal lesion leading to external expansion of nose known as Hebra nose.



Figure 2: Gross photograph of the excised lesion measuring 7x7 cm.



Figure 3: Gross photograph of the cut section of the lesion showing grayish white, firm in consistency with focal yellowish areas.

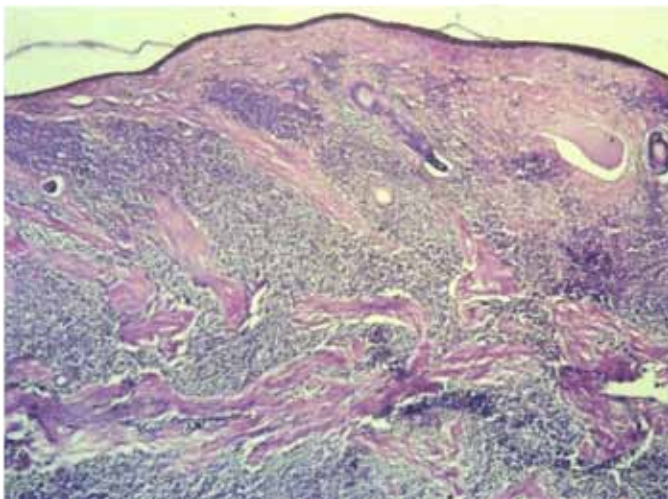


Figure 4: Photomicrograph showing sheets of inflammatory cell infiltrates and bands of fibrosis (H&E stain, x400).

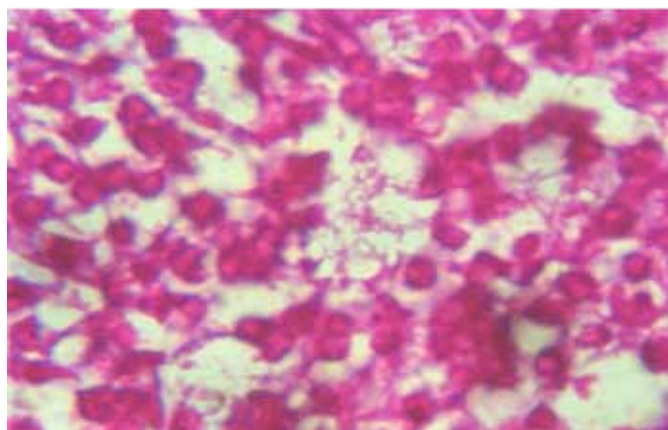


Figure 7: Photomicrograph showing Small rods in Mikulicz cells (Gram's stain, x1000).

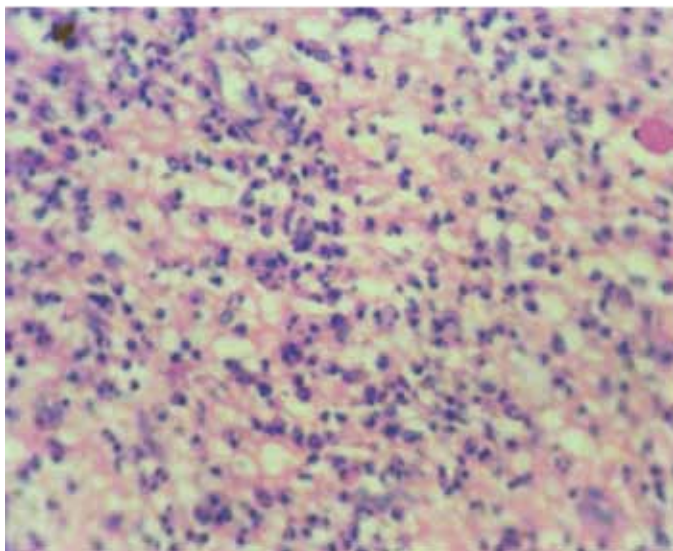


Figure 5: Photomicrograph showing mixed inflammatory cell infiltrates composed of neutrophils, lymphocytes, plasma cells and homogenous, eosinophilic round to ovoid bodies (Russell bodies) (H&E stain, x100).

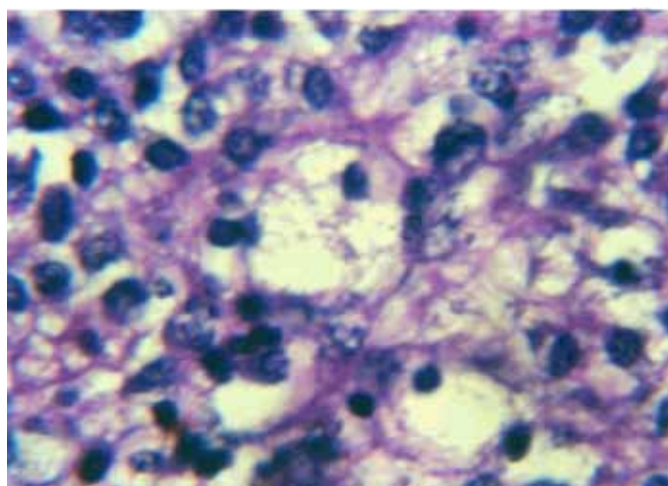


Figure 8: Photomicrograph showing Mikulicz cell with bacilli (Periodic acid schiff stain, x400).

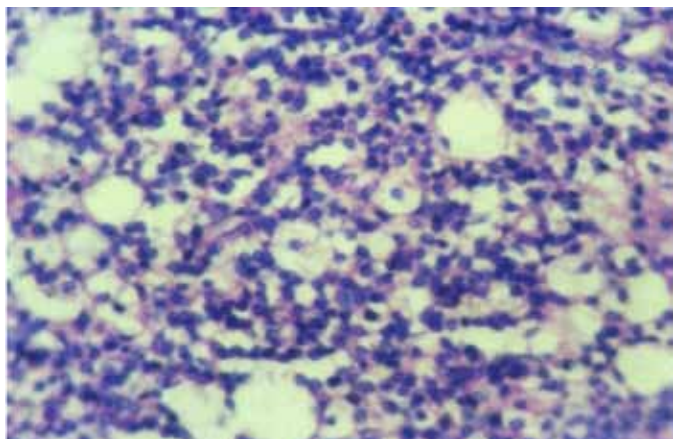


Figure 6: Photomicrograph showing lymphocytes, plasma cells and Mikulicz cells (H&E stain, x100).



Figure 9: Postoperative photograph of the patient after excision of the lesion and reconstruction of the nose and upper lip using oblique classical Indian forehead flap.

of rhinoscleroma is still unclear. Host susceptibility is thought to be important in development of the disease. Altered immune response with impaired cellular

immunity also plays an important role in the development of the disease apart from the infection. However, an alteration in CD4:CD8 population in blood has been postulated as a cause of chronicity of this disease. The altered proportion of CD4+ and CD8+ lymphocytes in the lesion may produce disabled macrophages, allowing bacterial multiplication inside them and an ineffectual delayed type hypersensitivity response [7].

The patient in our case had presented typically with a hypertrophic stage lesion involving the nose along with hypertrophy of nose ring areas as she used to put nose rings in both nostril areas as seen in the clinical and gross photograph.

Sood et al. [7] studied the cytohistological features of rhinoscleroma and correlated the cytological findings with the histopathological findings along with immunostain CD68. We could not do cytological examination and immunohistochemical study in our case.

Histopathological features are characterized by a chronic granulation tissue with abundant plasma cells and Mikulicz cells. Russell bodies (plasma cells with retained globules of immunoglobulins) are frequently observed. The characteristic cell is the Mikulicz cell, a large histiocyte with a pale, vacuolated cytoplasm containing causative bacilli which are diagnostic of rhinoscleroma. They can be seen faintly in sections stained with hematoxylin and eosin but are better visualized with the Giemsa stain or a Warthin-Starry silver stain. They are also stained magenta color by the PAS technique. They are Gram-negative rods that measure 2–3 µm in length and appear round or ovoid in cross section. Humans are the only identified host of *Klebsiella Rhinoscleromatis* [8].

A positive culture in MacConkey agar is diagnostic of rhinoscleroma, but it is positive in only 50–60% of patients. The diagnosis is confirmed by histology. Differential diagnosis includes other granulomatous conditions, e.g., tuberculosis, leprosy and fungal infections [9].

The mucosal epithelium overlying the cellular infiltrate often exhibits hyperplasia, which may be so pronounced as to give rise to a mistaken diagnosis of squamous cell carcinoma. Special stains and associated clinicopathologic features usually enable to distinguish rhinoscleroma from other conditions^[4]. In our case, marked hyperplasia of surface squamous epithelium but there is no doubt of malignancy.

In initial catarrhal stage, treatment with antibiotics streptomycin and tetracycline eradicates the infection. Combined surgery and antibiotic treatment are required in granulomatous and cicatricial stage. In our case, combined antibiotic treatment and surgery was curative and the case had been followed-up for five years.

CONCLUSION

Scleroma (rhinoscleroma) is a chronic granulomatous condition caused by *Klebsiella rhinoscleromatis* which is

endemic in northern parts of India. This report presents a case of nasal rhinoscleroma from South India presenting in granulomatous stage. When the disease progresses with proliferation, it may simulate a tumor, as in our patient with typical Hebra nose.

Author Contributions

Bhagyalakshmi A. – Substantial contributions to conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published

Rao C.V. – Acquisition of data, Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published

Krishna Kishore T. – Acquisition of data, Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published

Kartheek B.V.S. – Acquisition of data, Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published

Guarantor

The corresponding author is the guarantor of submission.

Conflict of Interest

Authors declare no conflict of interest.

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Article citation: Bhagyalakshmi A, Rao CV, Krishna KT, Kartheek BVS. Rhinoscleroma: A case report. Int J Case Rep Images 2014;5(12):868–872.



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