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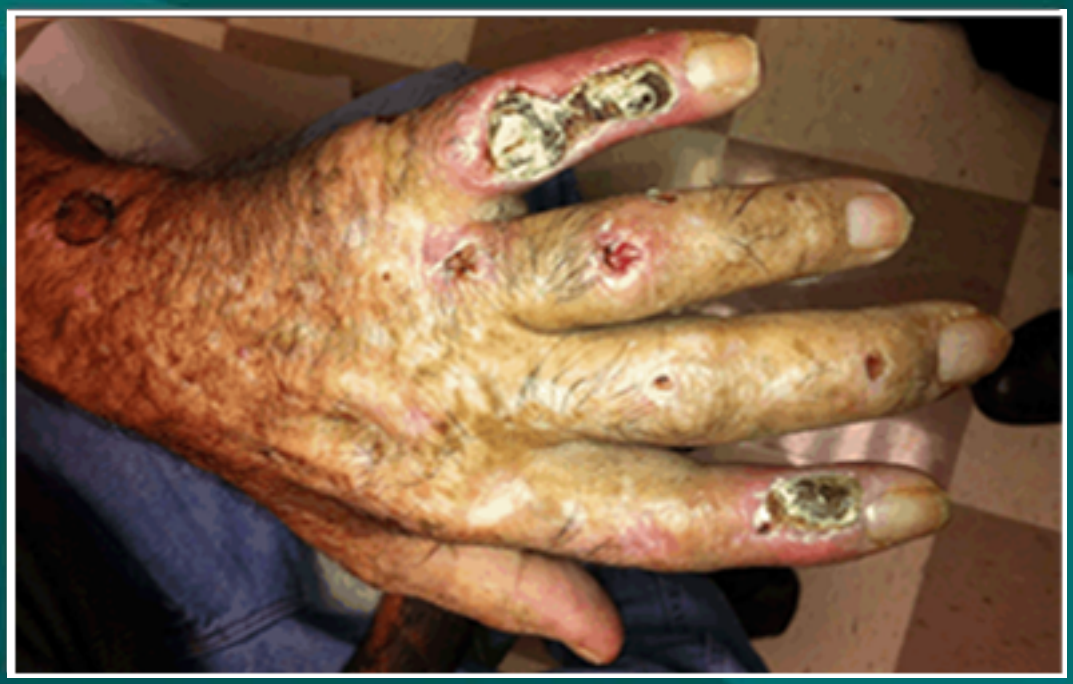

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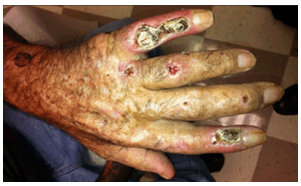

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CASE REPORT PEER REVIEWED | OPEN ACCESS

Cutaneous sarcoidosis presenting with diffuse panniculitis: A case report

Assane Diop, Diallo Moussa, Maodo Ndiaye, Abbaspour Valiollah, Pauline Dioussé, Boubacar Diatta, Fatimata Ly, Suzanne Oumou Niang, Mame Thierno Dieng, Assane Kane

ABSTRACT

Introduction: Cutaneous lesions in sarcoidosis are polymorphic. They can simulate most dermatological conditions. We report a case of cutaneous sarcoidosis presented as diffuse Panniculitis. Case Report: A 62-year-old male with no significant past medical history consulted for infiltrated lesion on abdominal skin infiltration lasting for two years. On examination, there were multiple indurated plaques, topped with keratotic papules giving an orange peel view, extending on abdominal genitalia and thighs skin associated with lymph nodes enlargement in different sizes and elastic consistence in axillary and inguinal regions. The histopathological examination of the skin lesion and the lymph nodes revealed typical sarcoidosis granulomas. Thoracic abdominal CT scan was normal. After six months of treatment with methotrexate and prednisone, evolution was remarkable by a rapidly skin lesions des infiltration and disappearance of lymphadenopathy. Conclusion: Cutaneous sarcoidosis as disseminated panniculitis is rarely reported in literature. In our patient, methotrexate combined with prednisone was effective. Other studies have confirmed the efficacy of methotrexate, especially as a steroid sparing treatment. This efficiency would be the result of adenosine production and a decrease in TNF α secretion in granulomatous lesions.

Keywords: Diffuse panniculitis, Methotrexate erythematous, Panniculitis, Sarcoidosis

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INTRODUCTION

Sarcoidosis is highly polymorphic in its clinical manifestations [1]. Clinically, sarcoidosis can mimic many skin diseases. However, Panniculitis as presenting feature in sarcoidosis is rarely reported [1–3].

We report a case of generalized granulomatous panniculitis in a patient with sarcoidosis.

CASE REPORT

A 62-year-old male, without remarkable past medical history, was admitted for a large erythematous indurated plaque on abdominopelvic region lasted for two years. Physical examination showed an erythematous indurated plaque with an orange peel appearance in abdominal

lower quadrant, pubic, external genitalia and thighs. The plaque was studded with hyperkeratotic papule especially on pubic region. A generalized ichthyosis sparing scalp and palmar-plantar region was noted (Figure 1). There were also consistent, painless, fixed lymphadenopathies of 2 to 7 cm in diameter, in auxiliary and inguinal regions. The skin and lymph node histopathologic studies revealed typical sarcoidosis granulomas. In skin, these granulomas were deeply situated in subcutaneous tissues and occupying fat lobules (Figure 2). The abdomen and thoracic CT scan was normal. Tuberculin test was anergic and HIV serological test was negative. CBC and blood smear were normal. After six months of treatment, combining methotrexate 25 mg intramuscularly per week and prednisone 0.5 mg / kg /day, the outcome was favorable, marked by fast skin lesions desinfiltration, disappearance of ichthyosis and lymphadenopathy regression (Figure 3).

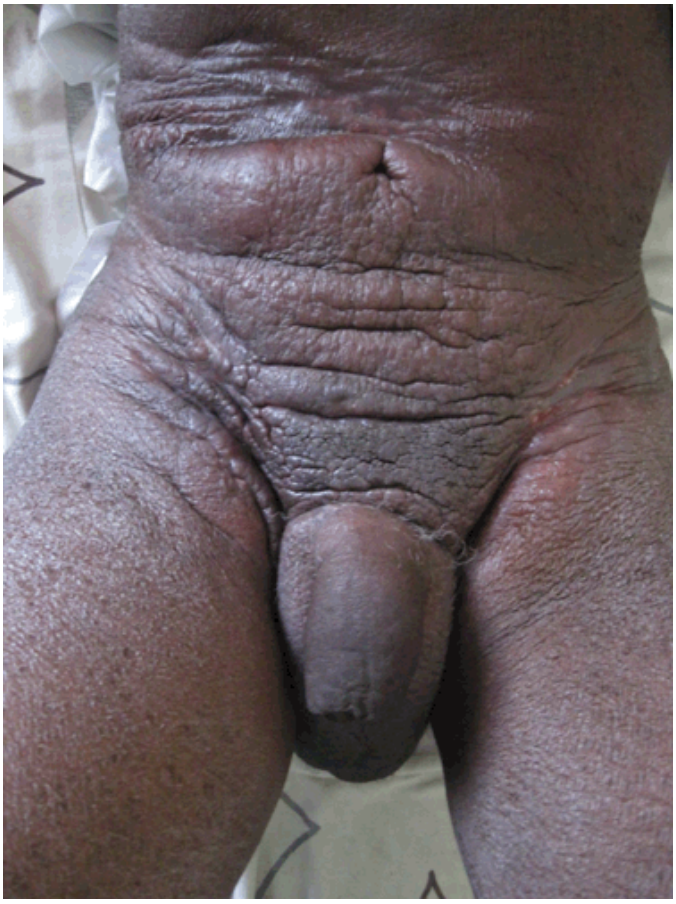

Figure 1: Very large indurated plaque.

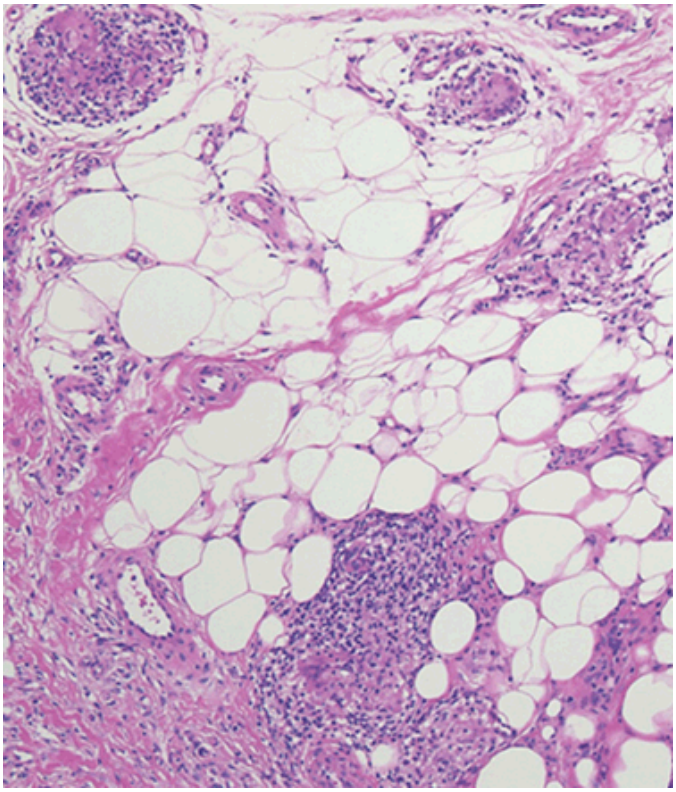

Figure 2: Subcutaneous tissue invaded by epithelioid granulomas and giganto-cellulaire with non caseating necrosis.

DISCUSSION

We report a cutaneous and lymph node sarcoidosis in which skin involvement was confirmed by histological examination as panniculitis. Specific manifestations of cutaneous sarcoidosis are related with the location of granulomas in the dermis. In literature, the frequency varies between 9% and 30% in different studies [4, 5].

They are characterized by small and large nodules called sarcoid, infiltrating sarcoid and sarcoid on scars. Hypodermic sarcoid of Darier-Roussy represents 4–12% of the specific lesions of sarcoidosis [5, 6] and appear as cold nodules developing in a normal-looking skin. They are located more frequently on the limbs and rarely on the trunk.

Cutaneous sarcoidosis with panniculitis as clinical presentation, it has rarely been reported [1, 7, 8]. To our knowledge, only 2 cases of sarcoidosis with extensive panniculitis have been reported. In one case, it was only a localized lesion on one limb [8]. In another case, the lesions were on supraclavicular, shoulder and upper back [7].

In our patient, the lesions were more generalized, on abdomen, pelvis, genitals and thighs. This very unusual clinical presentation can rise a problem of differential diagnosis with cutaneous lymphoma. However, the histopathologic features of sarcoidosis can make the difference with the skins as well as the lymph node lymphoma.

The treatment of sarcoidosis remains is poorly codified [9]. Corticosteroids are the standard treatment, but with a purely suspensive effect [10, 11]. It is indicated in severe eye, neurological, cardiac, renal, laryngeal and lung involvement or progressive disabling lesions and lupus pernio with severe hypercalcemia. However, glucocorticoids cause many side effects at more than 50% of patients.

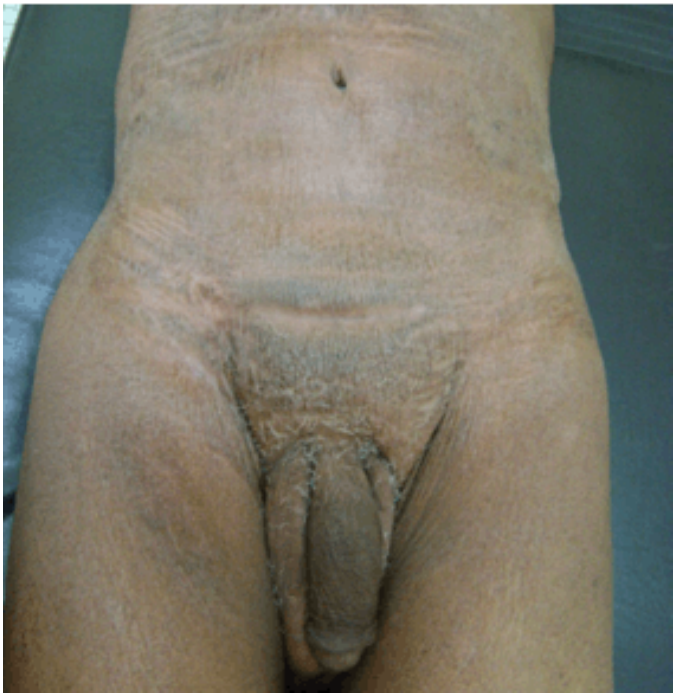

Figure 3: Skin desinfiltration after six months of treatment.

Side effects are even more frequent in height doses and long duration treatment [12] thus, methotrexate is often proposed as an alternative, especially as a reserve when long-term treatment with prednisone is obligatory [12]. In our case, treatment with methotrexate and prednisone had achieved a complete remission of lesions after six months. Other studies [13–15] have confirmed the effectiveness of methotrexate, especially as steroid sparing [16] treatment. This effectiveness would be the result of an increase in adenosine production and a decrease in the secretion and TNF α at granulomatous lesions [12].

CONCLUSION

Diffuse panniculitis as a presenting feature of sarcoidosis is rare. In such cases, only histopathology can confirm the diagnosis. Although treatment is not well classified, Methotrexate associated with prednisone seems to work well.

****

Author Contributions

Assane Diop – 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
Diallo Moussa – Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published
Maodo Ndiaye – Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published
Abbaspour Valiollah – Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published
Pauline Dioussé – Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published
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Suzanne Oumou Niang – Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published
Mame Thierno Dieng – Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published
Assane Kane – Analysis and interpretation of data, 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 PEER REVIEWED | OPEN ACCESS

Osteogenesis imperfecta complicated with psychosis secondary to complex partial seizures

Roya Samadi, Ali Akhoundpour Manteghi, Mehri Baghban Haghighi, Shervin Assari

ABSTRACT

Introduction: Osteogenesis imperfecta (OI) is an uncommon hereditary connective tissue disorder affecting collagen type I. The most common manifestations are frequent bone fractures and deformities, blue sclera, dental abnormalities and hearing loss. Seizures and mental retardation are not so common. Case Report: A 25-year-old male with usual symptoms of OI, mild mental retardation and a psychotic feature due to complex partial seizures, after an experience of head trauma. He was treated with risperidone, aripiprazole, oxcarbamazepine, alendronate and vitamin D3. Conclusion: It seems that seizure in OI is more common than general population. This may happen probably as a result of the complications of the head trauma in background of osteogenesis imperfecta. Moreover, clinical manifestations of complex partial seizure, can be mistaken with primary psychosis. Mental retardation and hearing loss might complicate this manifestation. So, psychiatric counseling and neurological evaluations should be carried out in OI patients.

Keywords: Complex partial seizures, Lobstein syndrome, Osteogenesis imperfect, Psychosis

How to cite this article

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INTRODUCTION

Osteogenesis imperfecta (OI) or Lobstein syndrome is a hereditary connective tissue disorder affecting collagen type I quantity or quality. It has 8 main types. Type 1 is the mildest and most common one. The pattern of inheritance is often autosomal dominant, although autosomal recessive and new mutation cases have also been reported [1]. Diagnosis is often made based on clinical manifestations and confirmed by DNA analysis. A range of clinical features can be seen in the skeletal system, such as osteopenia, frequent fractures, bone deformities, triangular face, short stature, disproportional body, hypotonia and laxity of skin and joints. Also, blue sclera, dental abnormalities and hearing loss are usually observed. The intellect is often normal [1]. Prevalent neurological complications include macrocephaly, communicating hydrocephalus, cerebral atrophy, basilar invagination, brainstem compression and skull fractures [2]. However, seizures are rather uncommon [2, 3].

CASE REPORT

The patient was a 25-year-old male, from a remote village in North Khorasan, Iran. When he was 21, he experienced a motorcycle accident, resulting in humerus fracture and head trauma, but without any seizures, amnesia, or coma. Brain CT-scan did not show any complication at the time. However, within 1–3 months after that, he developed a mental dysfunction, including intermittent psychotic periods, presenting with disorganized behavior and thought, disturbed form of thought, visual hallucinations, self-talking and laughing, aggressiveness and reduced need to sleep. He would have no recollection of these episodes afterwards. He also had states of left hand clawing, headache, vertigo and blurred vision just before the beginning of these episodes. He had been an easygoing as a child and had reached milestones normally. Family psychiatric history was negative.

He had been admitted in psychiatric centers twice previously and diagnosed with schizophrenia. The treatment following discharge from hospital had not been regular and effective. Duration of each episode was variable from several days to one or several months-based on information on past admissions or history taking from family. In his most recent admission in our hospital in Mashhad, his abnormal appearance, history of frequent bone fractures and skeletal deformities as a result of mild to moderate traumas were noticed. One of his five siblings had similar facies, skeletal deformities and frequent atypical fractures which led us to consider OI. Nevertheless, parental history of frequent fractures and deformities was negative. Therefore, more precise examinations, laboratory tests and genetic counseling were performed.

In general appearance, short stature, kyphoscoliosis (Figure 1), triangular face and macrocephaly were evident. Easy bruising and laxity of skin and ligaments, decreased muscular mass and strength, flat foot (Figure 2) and genu varum were observed. Deep tendon reflexes were normal.

In X-ray studies of extremities and vertebra, beside the osteoporosis (Figure 3), previous fractures were seen in atypical locations, such as compressed fracture of thoracic vertebra (T_7) (Figure 3A–B), shaft of the left femur (Figure 3C), left wrist and ankle and right elbow (Figure 4). Subsequent deformities included decreased height of the vertebra (Figure 3A–B), external rotation of left ankle and foot and flexion deformity of the elbow joint (Figure 4). Also, bone healing following the fractures seemed impaired as evidenced by malunion of the femoral shaft and elbow joint (Figure 3C–D). In skull X-rays, there were wormian bones (Figure 3E).

Audiometry indicated a unilateral conductive hearing loss. IQ test, he was not cooperative. However, mild mental retardation was suggested clinically based on his educational background: He had barely finished elementary school, with poor grades. Electroencephalogram (EEG) showed a generalized epileptiform spike and waves.

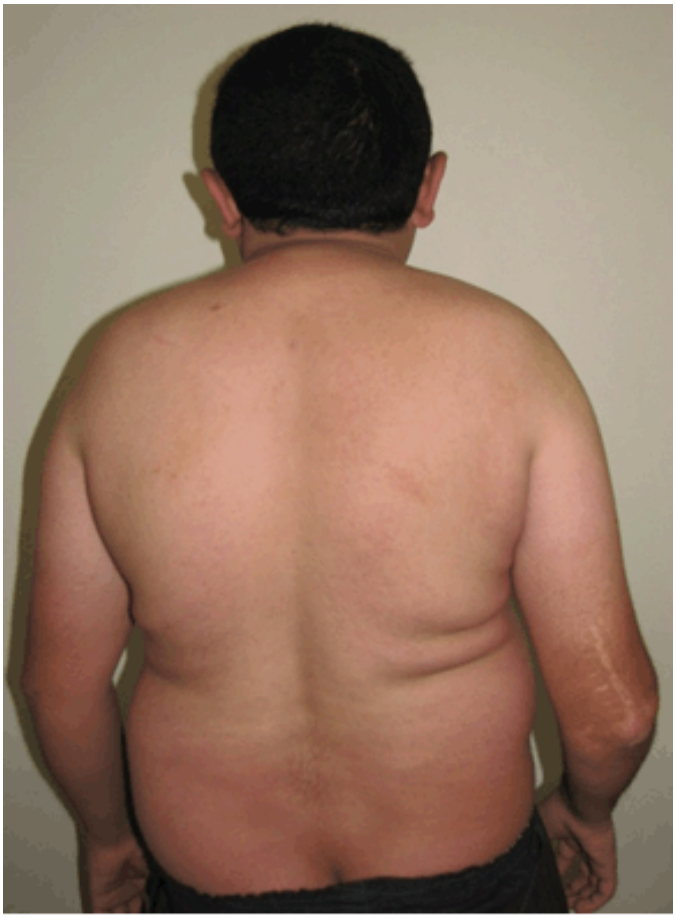

Figure 1: Kyphoscoliosis.

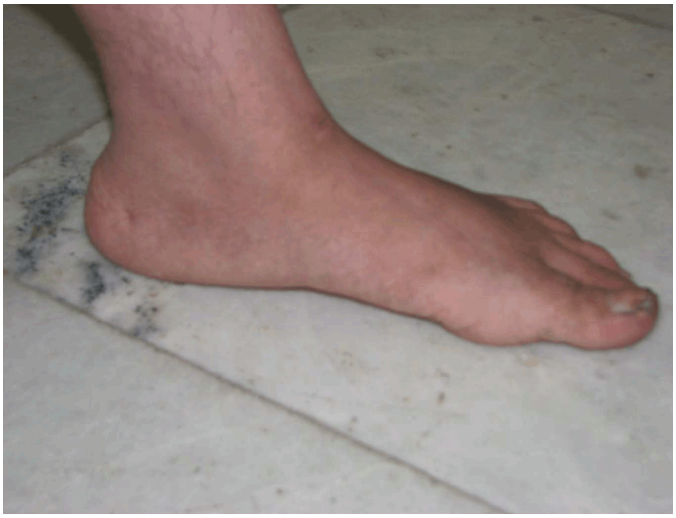

Figure 2: Flat foot.

Considering the intermittent pattern of his episodes, clear inter-episodic phases, the EEG findings and the non-deteriorating nature of his disease (he had had a constant level of function during these 4 years, in spite of not receiving sufficient treatment), and a favorable response to anticonvulsants; the symptoms of hand clawing, headache, vertigo and blurred vision prior to attacks were altogether considered as sensory auras

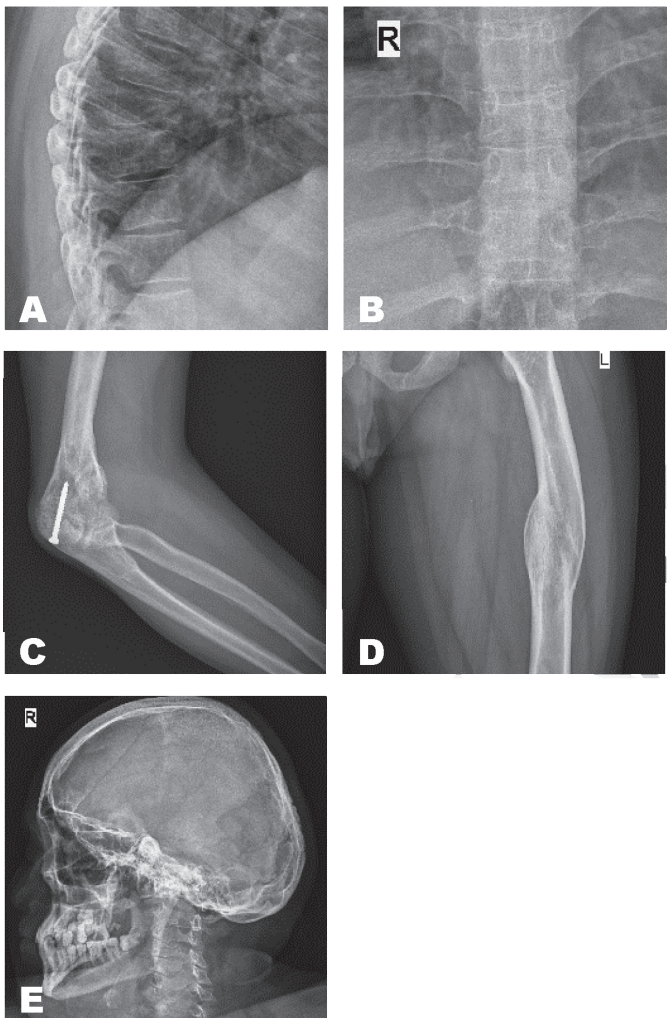

Figure 3: (A) Anteroposterior radiograph from dorsal spine shows anterior Wedge fracture in T7 and spinal mal alignment in the form of Kyphoscoliosis. Diffused reduction in spinal density due to Osteoporosis is seen, (B) Lateral radiograph from dorsal spine shows anterior Wedge fracture in T7 and spinal mal alignment in the form of Kyphoscoliosis. Diffused reduction in spinal density due to Osteoporosis is seen, (C) Elbow joint radiograph shows severe decrease in joint space (fusion article), flexion deformity and malunion due to fracture following previous trauma suggestive of severe secondary Osteoarthritis. Bowing deformity of Radius is seen. Relative reduction in bones' cortical thickness is suggestive of Osteoporosis. Metal fixator is observed in the distal of Humorous, (D) Anteroposterior radiographs from left Femur bone shows malunion of Femoral shaft following previous fracture along with Bowing deformity of Femur. Relative reduction in bone's cortical thickness is suggestive of Osteoporosis, and (E) Lateral radiograph from skull shows small bones with irregular border among Parietal and Occipital bones (Lambdoid suture) suggestive of Wormian bones.

(simple partial seizures), subsequently complicating with psychotic symptoms (complex partial seizures).

Magnetic resonance imaging scan of brain showed wide fissures, dilated ventricles and a sort of cortical atrophy, disproportionate with age. Serum levels of calcium and phosphorus were normal (9.7 and 3.7 mg/dL

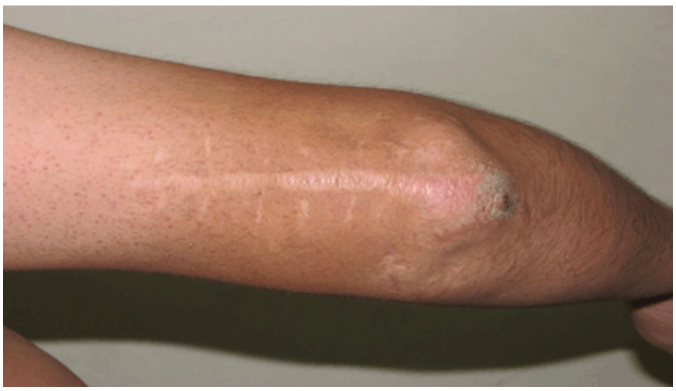

Figure 4: Flexion deformity of elbow joint.

respectively). However, serum alkaline phosphatase rose to 382 IU/L, probably due to frequent bone fractures. Thyroid function tests were normal. Genetic counseling indicated autosomal recessive OI.

Therapeutic plan was based on an antipsychotic (aripiprazole, 10 mg/day) an antiepileptic (carbamazepine, 200 mg/day initially and increased to 400 mg/day, 2 days later) and bone enhancing agents (alendronate, 70 mg weekly, and vitamin D_3 injection, 300000 IU, monthly). After admission aripiprazole was started and next day carbamazepine was added on. As the patient experienced the side effects of carbamazepine (dizziness and diplopia), it was changed to oxcarbazepine 150 mg/bid. This treatment satisfactorily prevented the episodes and thus proving that the symptoms were secondary to CPS. Loosening of associations and other psychotic symptoms improved after 10 days. Table 1 gives the differences between primary psychosis and CPS with psychosis.

Eventually, he was diagnosed with mild mental retardation and psychosis due to CPS. History of head trauma is thought to be involved in development of CPS in this patient, as OI may predispose patients to severe complications of minor traumas.

He was referred to an endocrinologist after discharge to better treat his OI. He has been followed-up in three visits in the last 2.5 years and is under treatment with oxcarbamazepine 150 mg, twice a day. No neuro-psychiatric symptoms have been reported or observed in this period.

DISCUSSION

Seizures are not frequently comorbid with connective tissue disorders such as osteogenesis imperfecta [3, 4]. However, in a study of the neurologic profile of OI patients by Charnas et al. [5], 5 cases out of 76 patients (6.5%) suffered from seizures, while the prevalence of seizures in the general population of diverse countries has been reported 1.5 to 57 per 1000 [6]. Therefore, it seems that seizures in OI are not absolutely rare and at least are more common than the general population.

Table 1: Differentiation between primary psychosis and CPS with psychosis.

	CPS with psychosis	primary psychosis
Pattern of episodes	The intermittent pattern	Not specific
Inter-episodic	clear inter-episodic phases	Not specific
EEG findings	Epileptiform spike/ sharp and waves	Not specific
Deterioration	Usually not	Maybe depends on psychotic disorder type
Streothypic auras	+	-
Hallucination type	Visual hallucinations are more common	Auditory hallucinations are more common
Response to treatment	Rapid response to anticonvulsant	Response to Antipsychotics within days to months- depends on psychotic disorder

It seems that OI patients (as well as our patient) have an increased tendency for bleeding during surgeries and even minor traumas, as a result of impaired collagen, capillary fragility, impaired platelet retention and aggregation and factor VIII deficiency [7–9]. These factors may lead to brain hematomas and, thereby, seizures and psychomotor retardation in OI patients [10].

This case report is an example of clinical manifestations of patients with CPS, which can be mistaken with primary psychosis. This was probably a complication of the head trauma in a patient with underlying osteogenesis imperfecta. Mental retardation, hearing loss, poor socioeconomic situation and low level of education, might have worsened the capability of the patient to cope with these problems and affected the outcomes as well. Therefore, besides orthopedic surgeries, additional interventions such as lifestyle modifications, physical rehabilitations and psychological counseling should be done in OI patients [1].

CONCLUSION

Clinical manifestations of complex partial seizure, can be mistaken with primary psychosis. As it seems that seizures in Osteogenesis imperfecta (OI) are more common than the general population, neuropsychiatric evaluation can also be beneficial in achieving the best therapeutic results and quality of life in these patients.

****

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Author Contributions

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

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

Mehri Baghban Haghighi – Acquisition of data, Drafting the article, Final approval of the version to be published

Shervin Assari – 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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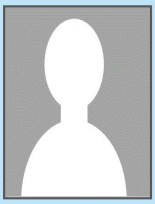

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CASE REPORT PEER REVIEWED | OPEN ACCESS

A rare cause of quadriplegia: Bilateral medial medullary syndrome presenting with "heart appearance sign"

Suryanarayana Sharma P. M., Mahendra J. V., Rohan R. Mahale, Acharya P. T., Madhusudhan B. K., Srinivasa R.

ABSTRACT

Introduction: Bilateral medial medullary infarct (MMI) is a very rare form of cerebrovascular disease presenting with quadriplegia, tongue weakness and posterior column sensory deficit. Initial reports of bilateral MMI were on autopsy. Only 38 magnetic resonance imaging (MRI) proven cases of bilateral MMI have been published in English literature till March 2011. Case Report: In the present case, patient presented with progressive quadriplegia of three days duration with respiratory involvement, MRI scan of brain revealed diffusion restriction in bilateral paramedian medulla appearing as characteristic heart appearance sign diagnostic of bilateral MMI. Conclusion: High index of suspicion is required to make early diagnosis in this rare stroke subtype. Optimal respiratory management may significantly improve the clinical outcome.

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INTRODUCTION

Medial medullary infarct (MMI) was initially described in late 19th century by Spiller [1]. Subsequent description has been credited to Dejerine [2]. Medial medullary infarction is uncommon; accounting for less than 1% cases of all brain infarctions [3]. Bilateral MMI is even rarer. Previously, the diagnosis of bilateral MMI was made at autopsy. Presently, with increasing use of the magnetic resonance imaging (MRI) in acute ischemic strokes, increasing number of bilateral MMI cases have been identified. Clinical presentation, stroke mechanism and outcome in patients with bilateral MMI is variable. Only 38 MRI proven cases of bilateral MMI have been published in English literature till March 2011 [4]. Herein, we report a patient with bilateral medial medullary infarction with a typical heart shaped sign on brain MRI.

CASE REPORT

A 74-year-old male with diabetes was referred to emergency department of our hospital with giddiness, dyspnea and left hemiparesis of three days duration.

He was intubated in local hospital and was referred to us for further management. Computed tomography (CT) scan of brain done outside did not reveal any acute infarct/bleed. On examination, the patient was drowsy, but obeying simple commands. His blood pressure was 190/100 mmHg and heart rate was 96/min. He had bilateral horizontal nystagmus with torsional component in all directions. His pupils were equal, reactive and eye movement was full. Gag reflex was impaired. Tongue movement could not be assessed. He had left hemiparesis (3/5 on MRC scale). He had generalized hyperreflexia with bilateral Babinski sign. Sensory system could not be evaluated. He had an ulcer over dorsum of right foot with amputated 4th and 5th toes. Other systems are unremarkable.

Hematological investigations did not show any significant abnormality. He had deranged renal parameters are creatinine 1.8, random blood sugar 283, HbA1C 8.7, total cholesterol 198, triglyceride 264.3, low-density lipoprotein 115.3 and his urine analysis revealed 15-20 pus cells with 1+ proteinuria. MRI scan of brain diffusion weighted (DW) imaging (1.5 T) revealed heart shaped hyperintensity areas in the bilateral ventral medulla with apparent diffusion coefficient (ADC) reversal as shown in (Figure 1). Similar findings were observed in the same region on T2 and fluid attenuated inversion recovery sequence (FLAIR) (Figures 2 and 3). Old infarcts in right occipital and parietal region were noted. Based on these findings, patient was diagnosed as having an acute bilateral medial medullary infarction (Figure 4) and he was treated with dual anti-platelets, enoxaparin, atorvastatin, ventilatory support, antibiotics and chest physiotherapy and DVT prophylaxis. Echo was normal. However, weakness progressed to quadriplegia on day-5 of hospitalization. He developed aspiration pneumonitis. He remained quadriplegic during his subsequent stay for 34 days in the hospital. Endotracheal culture revealed *Pseudomonas* and *Acinetobacter*, antibiotics were escalated to meropenem and colistin. He died on day-34 of hospitalization due to pneumonia and sepsis.

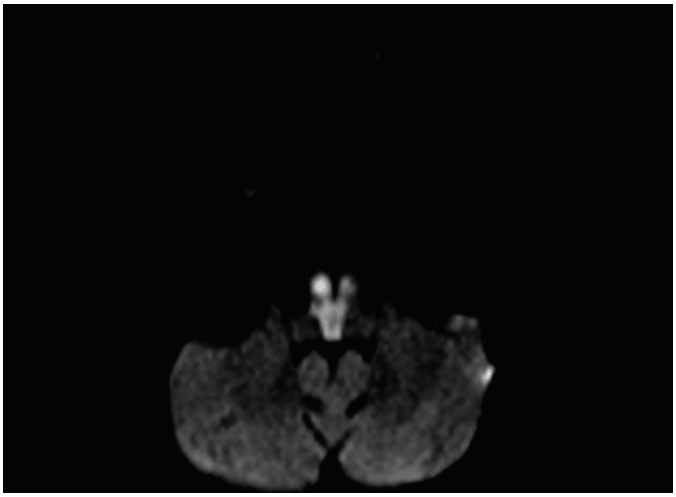

Figure 1: Diffusion restriction seen in bilateral medial medulla in diffusion weighted imaging sequence.

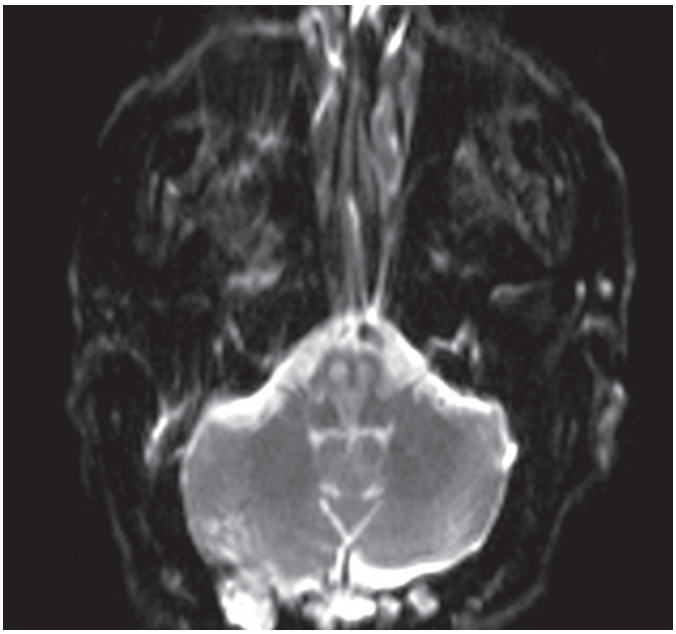

Figure 2: Hyperintensity noted in T2 sequence in the same region.

DISCUSSION

Medial medullary syndrome is a rare stroke subtype characterized by ipsilateral hypoglossal nerve palsy, contralateral hemiparesis sparing face and impairment of contralateral deep sensation [5]. It results from infarction of paramedian region of medulla oblongata due to occlusion of vertebral/anterior spinal artery or their small branches. Davison described a case of bilateral MMI in 1937 [6]. Bilateral MMI is even rarer stroke subtype and only 12 anatomically proven cases have been reported in English literature till date [7]. Subsequently, with advances in imaging technology with widespread use of MRI in diagnosis of acute ischemic strokes worldwide, additional 38 cases have been reported in the last 20 years. Bilateral medial medullary syndrome is characterized by classical triad of quadriplegia, tongue weakness and posterior column sensory deficits [8]. Katoh and Kawamoto classified bilateral MMI into type I, with an infarction area from medullary pyramid to pontine medial longitudinal fasciculus and type II with infarction confined to bilateral medullary pyramids [9]. Our patient was classified as having type I disease which has a worse prognosis.The vascular events likely to be associated with bilateral MMI are occlusion of vertebral artery or anterior spinal artery and its intrinsic penetrating branches. The infarcted area usually includes the pyramidal tracts, medial lemniscus, medial longitudinal fasciculus, hypoglossal nucleus or hypoglossal nerve fibres and medullary reticular formation bilaterally [10]. Before the advent of MRI scan, it was often confused with Guillain–

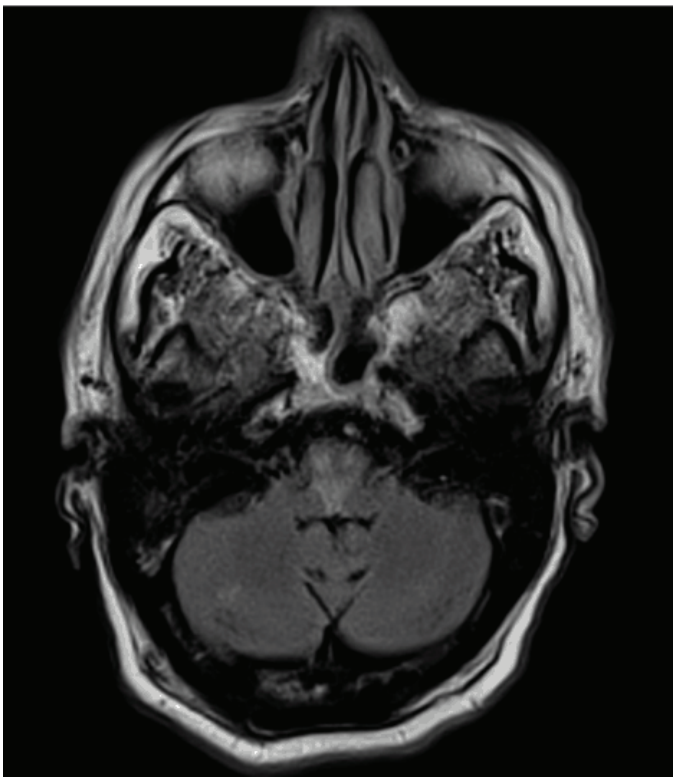

Figure 3: Fluid attenuated inversion recovery sequence (FLAIR) hyper intensity noted in the same region.

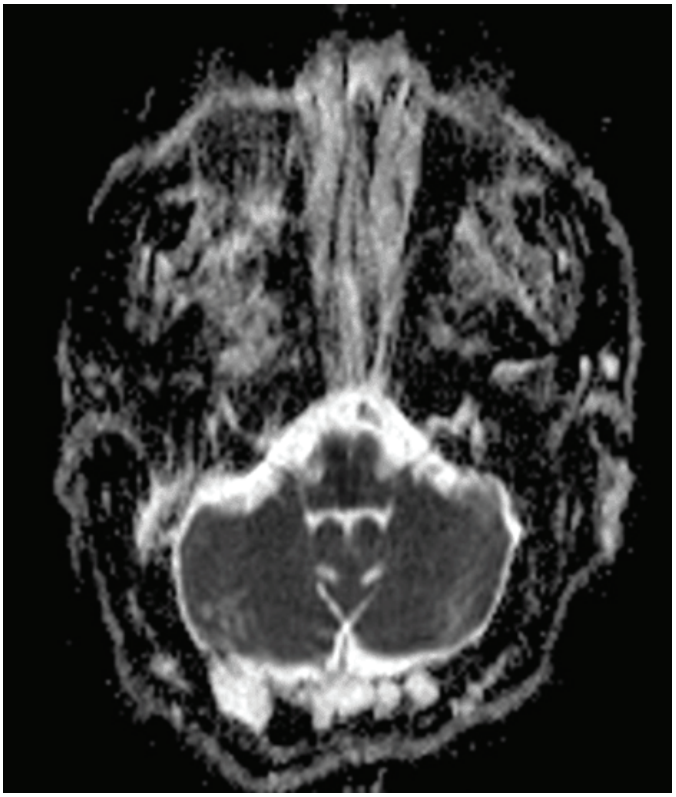

Figure 4: Diffusion restriction seen in bilateral medial medulla with apparent diffusion coefficient reversal.

Barre syndrome as it presented predominantly with quadriplegia. Limb weakness is a constant finding in this syndrome (85.3%) as in our case where patient initially presented with hemiparesis, subsequently progressing to quadriplegia with dysarthria and dysphagia [11]. Tongue weakness was not evident clinically as he was intubated elsewhere. Respiratory disturbance occurs in significant number of cases (29.4%) requiring ventilator support [10]. Computed tomography scan of brain is not a sensitive tool for posterior fossa, especially for medullary infarcts as in our case, CT scan of brain was normal even after three days of onset of symptoms. DWI sequence of MRI shows the characteristic heart appearance sign due to infarction of anteromedial and anterolateral territory of medulla [12]. Acute to subacute MMI can be differentiated accurately only by MRI. In acute MMI, there will be diffusion restriction with ADC reversal and no abnormality on T2/FLAIR sequences. Subacute MMI shows T2 shine through phenomena- no ADC reversal with hyperintense signal changes on T2/FLAIR. According to vascular supply, medulla is divided into anteromedial, anterolateral, lateral and posterior territory. Blood supply to these areas is predominantly by vertebral and anterior spinal arteries [13]. It is often difficult to identify to occluded vessel on MRA due to vastly complex network often formed by these vessels. Misdiagnosis/delay in diagnosis in this syndrome is common as patient presenting with areflexic quadriplegia sparing face may be misdiagnosed as Guillain–Barre syndrome [14], myasthenia gravis, brain stem encephalitis, inflammatory myopathy, periodic paralysis and paraneoplastic syndrome as other possible differential diagnosis. Bilateral MMI should be suspected in patients presenting with acute onset quadriparesis, tongue weakness and facial sparing. Pongmoragot et al. [4] have done a systematic review of bilateral MMI and have identified 38 MRI proven cases and concluded rostral medullary infarct (V-shaped) as the most common MRI finding in these cases. Aspiration pneumonia is the major cause of death in up to 66% of cases in different series [4]. Urosepsis and pulmonary thromboembolism contribute to other causes of mortality. In our case, patient succumbed to respiratory infection. Recognizing the severity of the respiratory symptoms in these patients is critical to avoid recurrent aspiration pneumonia and to improve the clinical outcome. Intravenous/intra-arterial thrombolysis using recombinant tissue plasminogen activator (r-TPA) may be beneficial in cases with bilateral MMI. Pfefferkorn et al. have demonstrated that a combination of intravenous thrombolysis with consecutive in endovascular mechanical thrombectomy may be a option in this difficult clinical situation [15].

CONCLUSION

In summary, patient presenting with rapid onset quadriplegia with sensory loss and bulbar weakness should raise the suspicion of bilateral medial medullary infarct (MMI). Advances in imaging technology like diffusion weighted MRI has greatly improved the yield of early diagnosis which is a key factor in predicting the

outcome. Bilateral MMI which was previously considered fatal can be effectively treated if respiratory management is optimally performed during acute period. We report this case because of rarity of its occurrence.

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Author Contributions
Suryanarayana Sharma P. M. – 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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Guarantor
The corresponding author is the guarantor of submission.

Conflict of Interest
Authors declare no conflict of interest.

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CASE REPORT PEER REVIEWED | OPEN ACCESS

Macroglossia and periorbital ecchymoses in a patient with systemic amyloidosis: A case report

Jamille Hemétrio Salles Martins Costa, Aloísio Benvindo de Paula, Leonardo de Oliveira Campos, Rafaela Brito de Paula, Daniel Riani Gotardelo

ABSTRACT

Introduction: Amyloidoses comprise a group of rare diseases associated with the extracellular deposition of misfolded proteins, which can compromise the function of target organs and give rise to clinical disease with a broad range of manifestations. The aim of this study was to report a case of systemic amyloidosis with macroglossia and periorbital ecchymoses - two uncommon semiological findings. Case Report: A 59-year-old female presented with dyspnea, vomiting, abdominal pain and distension. The patient was admitted for diagnostic workup, during which malnutrition, infiltrative thickening of the suprapubic abdominal wall, anasarca, macroglossia, and tongue petechiae were identified. The clinical picture was compounded by hematochezia and periorbital ecchymoses during hospitalization. Biopsy of the dermis and subcutaneous tissue of the hypogastrium revealed amorphous eosinophilic extracellular depositions on Congo red staining which had green birefringence under polarized light microscopy, consistent with amyloidosis. Conclusion: Patients with amyloidosis are usually extensively investigated before a diagnosis is made because in addition to being a rare disease with multifaceted presentation features, the signs and symptoms of amyloidosis are nonspecific. In the present report, cutaneous thickening with formation of periorbital ecchymoses accompanied by macroglossia were suggestive of amyloidosis, whose treatment and prognosis are influenced by timely diagnosis.

Keywords: Amyloidosis, Macroglossia, Periorbital ecchymoses

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INTRODUCTION

Amyloidoses are a subgroup of diseases caused by the aggregation of misfolded proteins with extracellular deposition which compromises the function of target organs and gives rise to clinical disease. Amyloidosis is a rare disease and a diagnostic challenge because of its nonspecific presenting features [1, 2].

Being a rare disease, the exact incidence of amyloidosis is unknown. In the United States, incidence rates seem stable at around 6–10 cases/million/year. Older adults

and males account 65–70% cases of the disease. The mean age at diagnosis is 64 years; Less than 5% patients are under 40 years of age at the time of diagnosis [3].

The term "amyloid" was attributed by Rudolph Virchow in 1854, when he noted a reaction of metachromasia to iodine in necropsied tissue samples, similarly to what occurs with starch, and assumed the material was of glycidic origin. Although Friedreich and Kekule demonstrated in 1859, that the material was in fact protein, the denomination was already incorporated into the medical vocabulary and was thus maintained. The "amyloid" deposit is necessarily composed of a fibrillar protein, glycosaminoglycans and serum amyloid P-component. Amyloid fibrils have a secondary structure in common—a beta-pleated sheet configuration—and a single ultrastructure which determines the 30 different precursor proteins known to date [4].

Amyloid diseases can be categorized as systemic or localized; hereditary or acquired. The current classification is based on the different types of protein of the amyloid fibrils, most often related to the distinct clinical presentations. The prognosis of localized disease is generally good with surgical treatment. If there is systemic involvement, the disease can be severe; with cardiomyopathy, nephrotic syndrome/ renal failure, hepatosplenomegaly, diarrhea, intestinal pseudo-obstruction, peripheral neuropathy, autonomic neuropathy, arthropathy, carpal tunnel syndrome, bleeding, adrenal dysfunction, gout, weight loss, pulmonary problems, fatigue, and malaise [1].

A tissue biopsy and histopathological examination are done to establish the diagnosis. Amyloid deposition is identified using Congo red histological staining and subsequent observation of green birefringence under polarized light—the established gold standard. The precursor fibril is then characterized using histochemical and biochemical testing, and genetic analysis [5]. The correct and specific diagnosis of the amyloidosis type is essential to guide treatment.

CASE REPORT

A previously healthy 59-year-old female was admitted with mild dyspnea, vomiting, abdominal pain and distension for diagnostic workup. The patient complained of lower abdominal heaviness in addition to postprandial bloating and decreased appetite of approximately one year duration, resulting in progressive weight loss that warranted extensive medical investigation at the time. She denied fever, inflammatory signs and changes in bowel habits. On physical examination, malnutrition, anasarca, macroglossia (Figure 1), tongue petechiae, and infiltrative thickening of the suprapubic abdominal wall were identified. During hospital stay, the disease progressed with hemorrhagic phenomenon consisting of massive hematochezia and bilateral periorbital ecchymoses in addition to extensive left-eye conjunctival hemorrhage (Figure 2). Pericardial effusion, bilateral pleural effusion and ascitis were noted on computed tomography scan. Abdominal magnetic resonance imaging showed edematous infiltration of the mesenterium and subcutaneous tissue in the hypogastrium. A barium meal revealed reduced small bowel motility. Hyperemia and ulcers were found in the colon and rectum on colonoscopy, the etiology to be determined by histopathology. Laboratory tests revealed antinuclear antibody (ANA), double-stranded deoxyribonucleic acid (ds DNA), rheumatoid factor (RF), hepatitis B surface antigen (HBsAg), anti-hepatitis C virus (Anti-HCV) antibody and human immunodeficiency virus (HIV) antigen within normal limit. Mantoux (with 10 U tuberculin) and alcohol acid-fast bacilli (sputum): were negatives. Blood glucose, urea, creatinine, sodium, potassium and bicarbonate levels were normal. Routine peripheral smear examination showed hypochromic microcytic anemia. C-reactive protein (CRP) 48 mg/L. TSH 6.06 ng/dL (0.27–4.2) ng/dL. free T4 1.19 ng/dL (0.93–1.7 ng/dL). Urinalysis was within normal limits except for a erythrocyturia (20/HPF). Fundoscopy showed abundant diffuse opacities in the vitreous. Electrophoresis of urine and blood proteins showed a monoclonal peak in the region of alpha-2-globulins. Immunohistochemistry of the bone marrow biopsy specimen was positive for CD138 with numerous plasma cells (90% of cell count), in addition to the presence of light-chain kappa and lambda globulins. Echocardiography showed grade III left ventricular diastolic dysfunction, pericardial effusion in addition to parietal and visceral thickening. Biopsy of a fragment of the dermis and subcutaneous tissue of the hypogastrium revealed amorphous eosinophilic extracellular deposits on Congo red staining and green birefringence under polarized light, both consistent with amyloidosis. The possibility of the disease being associated with multiple myeloma was excluded given the clinical manifestations and evidence from ancillary tests (absence of hypercalcemia, renal failure, or osteolytic lesions consistent with myeloma).

Cardiac decompensation followed, as the patient was in a condition of severe advanced disease, while the medical team was expecting the results of the biopsy with Congo red staining. The patient died before the specific therapy was instituted.

DISCUSSION

Macroglossia is the most frequent oral manifestation of amyloidosis and can be found as the only presenting symptom or as only one of the symptoms of the disease. Before considering the presence of amyloid protein, other more likely causes of tongue enlargement, such as malignant tumors of the tongue, vascular abnormalities, hypothyroidism and deficiency of vitamin B12 and folic acid, should be considered. Xavier et al. described a case of an older adult with macroglossia, weight loss,

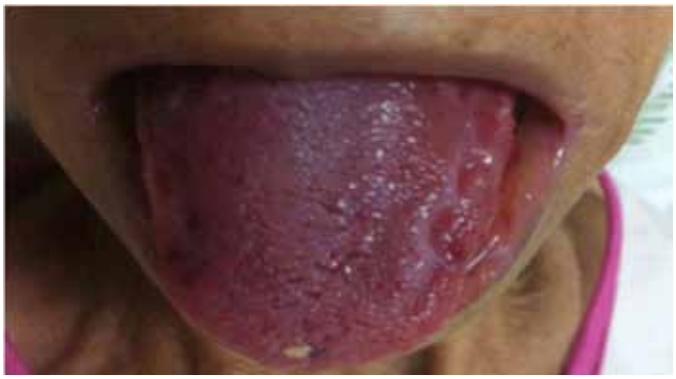

Figure 1: Macroglossia demonstrated by the impression of the teeth on the surface of the tongue. Associated petechiae suggesting coagulopathy.

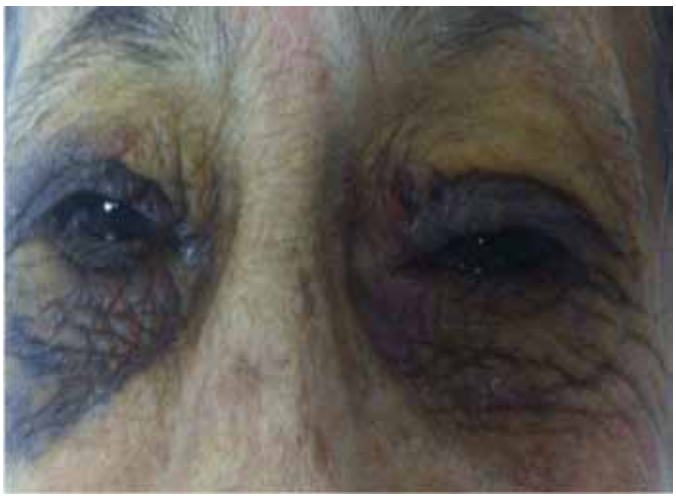

Figure 2: Bilateral periorbital ecchymosis due to the involvement of blood vessels by the infiltration of amyloid proteins.

and dysphagia that seemed at first to be a malignant tumor of the tongue and that, after proper workup, was defined as amyloidosis [6]. Tsourdi et al. reported a case of macroglossia as the sole manifestation of amyloidosis secondary to a monoclonal gammopathy of undetermined significance [7].

Purpuric eruptions such as ecchymoses and hematomas can also be found in individuals with amyloidosis. They are due to coagulation factor X deficiency, likely the result of its absorption by the amyloid fibrils, in addition to amyloid infiltration of capillaries causing microvascular fragility. Few cases have been described in literature in which the two findings—macroglossia and periorbital ecchymoses—were concurrent in patients with systemic amyloidosis; in most such cases, amyloidosis was associated with multiple myeloma [8–9].

In the case reported herein, no underlying diseases were found to account for the amyloid deposition, hence the diagnosis of primary systemic amyloidosis was considered. This type of amyloidosis is known as AL, with the first letter ("A") corresponding to "amyloidosis" and the second representing the biochemical makeup of the constituent fibril—in this case, amyloidosis involving the deposition of light-chain immunoglobulins ("L" for "light-chain").

Three of the four diagnostic criteria to confirm AL-type systemic amyloidosis were verified in our study: (1) presence of a syndrome related to the amyloid deposits (heart failure and macroglossia, among other signs and symptoms); (2) evidence of amyloid deposition on Congo red staining in a tissue biopsy sample; and (3) presence of monoclonal plasma cell proliferation. The fourth diagnostic criterion would be the confirmation of light-chain proteins in the amyloid material through immunohistochemistry or other molecular biology techniques. These tests were not performed because of the rapidly fatal outcome [10].

The prognosis of AL amyloidoses is typically poor. Heart failure and renal failure are the main causes of death. When amyloidosis is secondary to multiple myeloma, the mean survival is five months, while the primary form of the disease is associated with a survival of 2.1 years. The treatment for AL amyloidosis is intended to reduce the amount of circulating precursor proteins produced by B-lymphocytes and plasma cells, which can be achieved with cytotoxic agents such as prednisone and melphalan [1].

CONCLUSION

Macroglossia, periorbital ecchymoses, and other hemorrhagic manifestations are among the multiple presenting features to be found in systemic amyloidosis, which is a severe disease with complex symptomatology requiring thorough clinical examination and early recognition by the medical team to ensure timely treatment.

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Author Contributions

Jamille Hemétrio Salles Martins Costa – 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
Aloísio Benvindo de Paula – 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
Leonardo de Oliveira Campos – 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
Rafaela Brito de Paula – 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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Authors declare no conflict of interest.

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CASE REPORT PEER REVIEWED | OPEN ACCESS

Intraoperative transesophageal echocardiographic detection of intracardiac thrombus and pulmonary embolism during orthotopic liver transplant

Amie Hoefnagel

ABSTRACT

Introduction: Transesophageal echocardiography (TEE) utilization during liver transplantation is beginning to gain favor in many medical centers. The intraoperative course during liver transplant includes periods of increased and decreased peripheral vascular resistance, large amounts of third spacing, high volume replacement needs, and the possibility of acute right heart failure and circulatory collapse at reperfusion. Additionally, these patients may have underlying systolic dysfunction and coronary artery disease. Intraoperative TEE provides the anesthesiologist with the only single monitoring modality that can be used to diagnose all of these. Fear of bleeding complications due to esophageal varices, and the lack of provider competency with TEE are often sited as reasons to avoid TEE in this patient population. Case Report: This is a case of an intracardiac thrombus and pulmonary embolism in a 44-year-old male undergoing orthotopic liver transplantation for Laennec's cirrhosis. Conclusion: In this case, the routine use of intraoperative TEE provided for diagnosis of a massive intracardiac thrombus and pulmonary embolism during the dissection phase of liver transplantation, adding to the growing body of case reports supporting TEE as a diagnostic tool during orthotopic liver transplantation.

Keywords: Intracardiac thrombus, Orthotopic liver transplantation, Pulmonary embolism, Transesophageal echocardiography

How to cite this article

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INTRODUCTION

There are several reports of pulmonary embolism (PE) and/or intracardiac thrombus (ICT) during the intraoperative period of liver transplantation [1]. Patients with end-stage liver disease (ESLD) have defective coagulation due to impaired synthesis of clotting factors. They also have increased rates of fibrinolysis, increased concentrations of tissue plasminogen activator (tPA) and decreased concentrations of tPA-specific inhibitor [2]. Given the complexity of the balance between coagulation, anticoagulation, and fibrinolysis combined with the stresses of a major abdominal surgery with large volume loss, multiple vascular anastomosis, transfusion of blood product, exposure to citrate toxicity, and the presence of intracardiac monitors, it is quite amazing that these events are as rare as they are. TEE is the only diagnostic modality available to the anesthesiologist for intraoperative evaluation of PE and ICT, providing the ability to directly visualize thrombus and to garner information about the physiologic cardiac affects [3].

CASE REPORT

A 44-year-old male presented for orthotopic liver transplantation (OLT) due to acute decompensation of Laennec's cirrhosis. He had been hospitalized for approximately three weeks with worsening mental status and acute renal failure requiring continuous veno-venous hemofiltration (CVVH). His model for end-stage liver disease score (MELD) was >40 at the time of transplantation (INR 2.2, total bilirubin 24.1 mg/dL, and creatinine 2.52).

The day prior to transplantation the patient required transfusion of several units of red blood cells (pRBCs), and fresh-frozen plasma (FFP) due to genitourinary hemorrhage after a traumatic Foley catheter placement. He was also placed on an aminocaproic acid infusion at 1 g/hr after a 5 g loading dose, which was stopped prior to surgical incision. In the operating room, bilateral radial arterial lines, a rapid infusion catheter, an introducer, and pulmonary artery catheter were placed after induction of general anesthesia. Post-induction hemodynamics were consistent with end-stage liver disease and showed an increased cardiac output and systemic vasodilation. A transesophageal echocardiogram (TEE) was performed. Initial evaluation revealed a patent foramen ovale best imaged in the midesophageal RV inflow-outflow view (Video 1, http://www.ijcasereportsandimages.com/archive/2015/006-2015-ijcri/CR-10520-06-2015-hoefnagel/ijcri-1052006201520-hoefnagel-full-text.php). The imaging was also positive for a hyperdynamic and under filled left ventricle.

During the dissection phase of surgery, there was a sudden drop in blood pressure with near immediate equalization of the systemic and pulmonary pressures. The end-tidal carbon dioxide fell from 34 mmHg to 10 mmHg, and mixed venous saturation fell from 78% to 58%. A midesophageal four-chamber view showed severe enlargement of the right atrium and ventricle and an intracardiac thrombus attached to the pulmonary artery catheter. There were additional TEE signs consistent with further embolization of the thrombus into the pulmonary arteries, such as a slit like, under filled left ventricle, and a continued left-ward bowing of the intra-atrial septum during the entire cardiac cycle. A slight leftward rotation of the TEE probe was used to focus on the right atrium and ventricle for better visualization of the thrombus (Figure 1). The patient developed pulseless electrical activity (PEA) that was treated with chest compressions and one dose of epinephrine. Visualization of the main pulmonary artery after chest compressions did not show thrombus. Attempts at aspiration of thrombus via the introducer and pulmonary artery catheter were unsuccessful. High doses of epinephrine were required to maintain an adequate blood pressure and the decision was made to abort the transplant. The patient's abdomen was closed, and he was transported to the surgical intensive care unit. Attempted catheter thrombectomy was aborted after pulmonary angiography revealed patent main pulmonary arteries. The patient developed a clinical picture consistent with disseminated intravascular coagulopathy, worsening acidosis, and pupils became fixed and dilated. He died twelve hours after the initial thrombotic event.

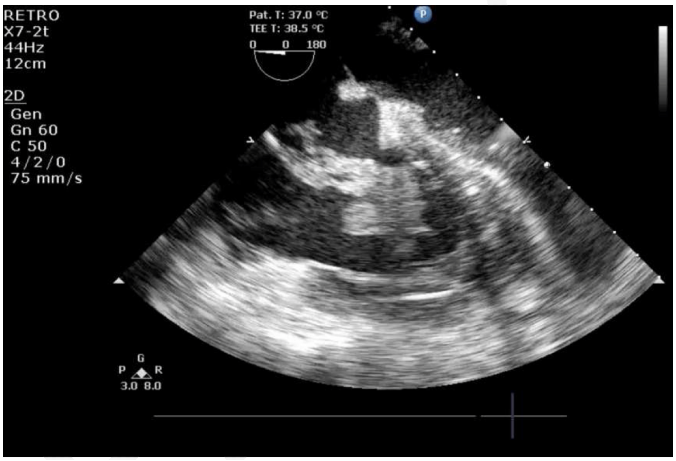

Figure 1: The intracardiac thrombus crossing the tricuspid valve. Severe right atrial enlargement and leftward curvature of the intra-atrial septum are also seen.

DISCUSSION

TEE is the only diagnostic modality available to the anesthesiologist for intraoperative evaluation of PE and ICT. An intracardiac thrombus is defined as an echo dense, discrete, mass that is seen during both systole and diastole. The mass must be discrete from the endocardium. Treatment of intracardiac thrombi involves anticoagulation and serial monitoring with echocardiography to follow resolution. Occasionally with large thrombi, or ones that further embolize to the pulmonary circulation, surgical removal may be considered [4]. Unlike intracardiac thrombus, direct visualization of pulmonary thrombus on TEE is seen in roughly one-quarter of patients with known pulmonary emboli [5]. Therefore, indirect markers are utilized for diagnosis. Right ventricular dysfunction, leftward bowing of the intra-atrial septum, and moderate to severe tricuspid regurgitation all have high sensitivity for PE [5, 6]. McConnell's sign--akinesia of the RV free wall with sparing of the apex-- (Video 2, http://www.ijcasereportsandimages.com/archive/2015/006-2015-ijcri/CR-10520-06-2015-hoefnagel/ijcri-1052006201520-hoefnagel-full-text.php) has a sensitivity of 77% and specificity of 94% for PE [7]. Additional criteria consistent with PE include RV dilation with an RV/LV end-diastolic diameter >1, or an RV end-diastolic diameter >30 mm [8]. To obtain these values the transgastric mid short axis-view is used. From this view the transgastric right ventricular apical short-axis view can be found by advancing the transducer slightly, rotating the probe to the right and antiflexing it. This

view will allow for better measurement of the right ventricle [9]. Additional TEE signs consistent with PE are pulmonary artery systolic pressure >30 mmHg and a tricuspid regurgitant velocity >2.8 m/s [8].

Another indirect echocardiographic sign of acute PE is the 60/60 sign—pulmonary artery acceleration time of <60 milliseconds with a maximal tricuspid regurgitant pressure gradient of <60 mmHg. The pulmonary artery acceleration time is the interval between the onset of systolic flow in the pulmonary artery and its peak velocity. This is measured with pulsed-wave Doppler (PW) interrogation of the pulmonary artery. Several TEE views can be utilized for this measurement including the midesophageal ascending aorta SAX, upper esophageal aortic arch LAX, or the transgastric RV inflow-outflow view [10].

CONCLUSION

In this case intraoperative Transesophageal echocardio-graphy (TEE) allowed for diagnosis of massive intracardiac thrombus (ICT) and pulmonary embolism (PE) within moments of its occurrence. Unfortunately, this patient did not survive the event, however, the rapid diagnosis allowed for re-allocation of the donor organ and a successful transplant for a different patient.

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Author Contributions

Amie Hoefnagel – 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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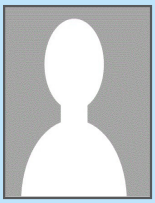

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CASE REPORT PEER REVIEWED | OPEN ACCESS

Primary cavernous hemangioma of the thyroid

Meryem Ilkay Eren Karanis, Arif Atay, Ilknur Kucukosmanoglu, Cevdet Duran, Alpaslan Sahin

ABSTRACT

Introduction: Hemangiomas are common benign vascular tumors. Primary thyroid cavernous hemangiomas are extremely rare and have been reported only as case reports in literature. In this report, a case with thyroid cavernous hemangioma was reported. Case Report: A 45-year-old female with history of enlarging anterior neck mass referred to our clinic. Ultrasonography showed a single hypoechogenic nodule in the right lobe of the thyroid. Right hemi-thyroidectomy was performed and cavernous hemangioma was diagnosed. Conclusion: Preoperative differential diagnosis thyroid hemangioma is very difficult. Surgery should be indicated when malignancy or cavernous hemangioma is suspected or compressive symptoms developed and it provides a good prognosis.

Keywords: Cavernous hemangioma, Nodular goiter, Thyroid, Vascular tumor

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INTRODUCTION

Hemangiomas are common benign vascular tumors composed of blood vessels of various size lined by plump to flattened endothelial cells with no atypia [1]. Cavernous hemangiomas are common in children and adults and tend to invade the upper half of the body and mostly seen in cutaneous tissue, and it can be located in the deep soft tissue and in organs. Primary thyroid cavernous hemangiomas are very rare and have been reported in limited case reports [2]. Secondary hemangioma may occur as a result of repeated fine-needle aspiration biopsy (FNAB) [3].

Herein, we report a case with primary thyroid cavernous hemangioma.

CASE REPORT

A 45-year-old female was referred to our clinic with enlarging anterior neck mass for six months. She had no compressive symptoms, history of trauma, previous fine-needle aspiration biopsy or other invasive neck procedures. There was no family history of thyroid diseases. On physical examination, hard, painless, mobile mass was detected in thyroid region. Serum thyroid stimulating hormone and free T4 levels were normal, and no antithyroid antibodies were detected. Ultrasonography (USG) showed a single hypoechogenic mass in the right lobe of the thyroid with a size of 41x60x65 mm in diameter. There were neither abnormal findings in the left lobe nor

in cervical lymph nodes. Right hemithyroidectomy was performed and macroscopically; 5.5 cm in diameter, well-circumscribed encapsulated nodular lesion was extracted. Cross-sectional surface of nodular lesion was hemorrhagic in most areas, with patchy areas of fibrosis and myxoid change (Figure 1). Touch imprint method was applied to the nodular lesion and microscopically, intensive blood cells, and some small number of swollen endothelial cells were seen (Figure 2). On histopathology; the nodular lesion was composed of large, cystically dilated, anastomosing blood vessels that were filled with blood cells and lined by flat endothelium without atypia (Figure 3) and degenerative areas were also seen. Endothelial cells stained with immunohistochemical CD34 stain (Figure 4). As a result, primary cavernous hemangioma of the thyroid was diagnosed.

The patient was discharged from the hospital two days after surgery with no signs of complications. The patient currently remains asymptomatic 10 months after the operation.

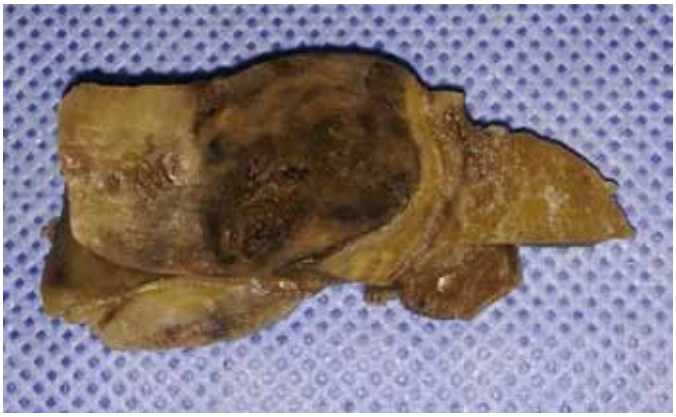

Figure 1: Macroscopic appearance of the thyroid cavernous hemangiomas: Well-circumscribed nodular lesion with hemorrhagic cut surface in most areas, irregular areas of fibrosis and myxoid change.

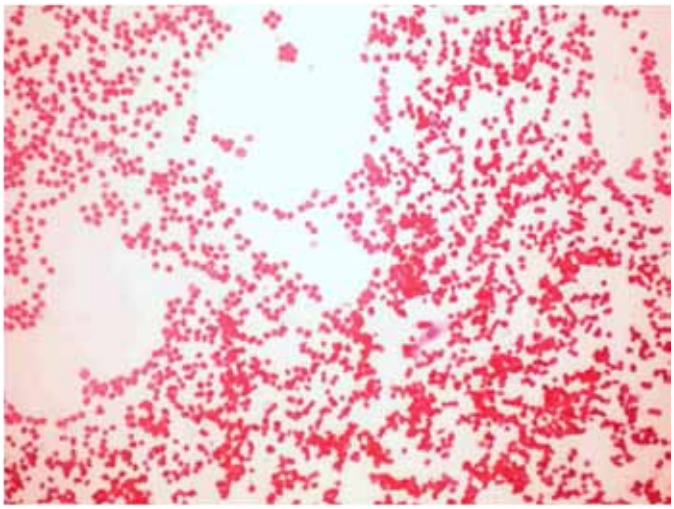

Figure 2: Intensive blood cells and only very small number of swollen endothelial cells in touch imprint preparations (H&E stain, x50)

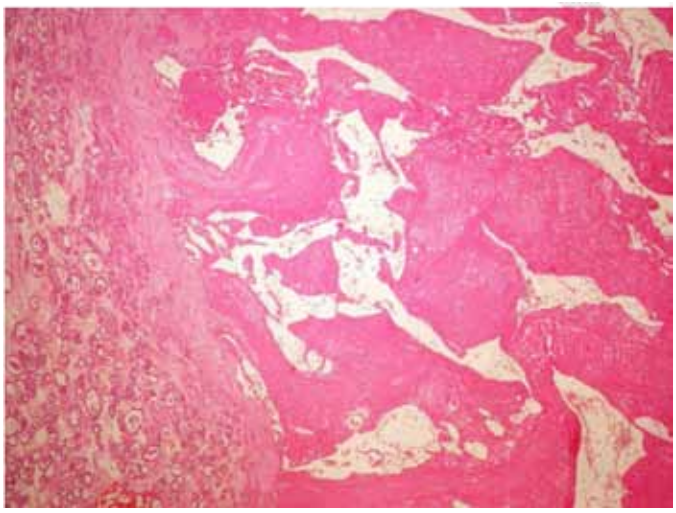

Figure 3: Microscopic appearance of the thyroid cavernous hemangiomas: Large, cystically dilated, anastomosing blood vessels in the right part of the picture and thyroid follicules in the left part of the picture (H&E stain, x50)

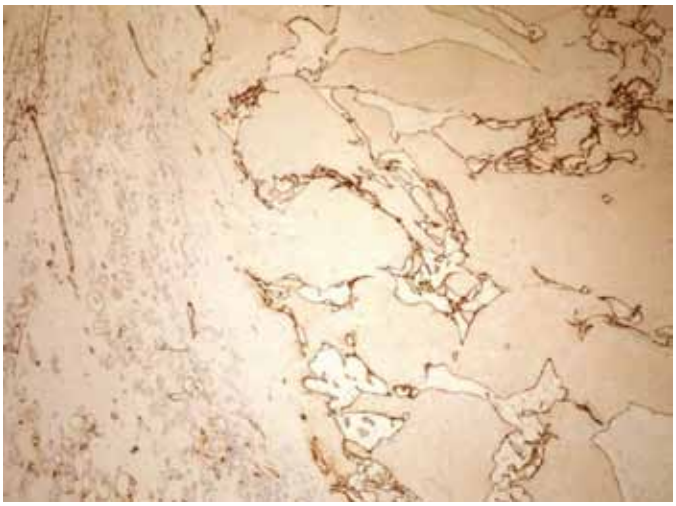

Figure 4:Immunohistochemical CD34 positivity in endothelial cells (CD34, x50).

DISCUSSION

Hemangiomas are one of the most common soft tissue tumors. They are benign and composed of various sized blood vessels, lined by plump to flattened endothelial cells with no atypia. Cavernous hemangiomas are common in children and adults. Cavernous hemangiomas tend to hold the upper half of the body and mostly seen in cutaneous tissue, else it can be located in the deep soft tissue and in organs [1].

Primary cavernous hemangiomas of the thyroid are very rare. In most cases, thyroid gland hemangiomas may be considered as a consequence of vascular proliferation that follows the organization of a hematoma after a trauma or FNAB and such hemangiomas of the thyroid are named secondary thyroid hemangioma. [3]. Primary thyroid hemangiomas are considered to be

a developmental anomaly [4]. A few cases of primary cavernous hemangioma of the thyroid gland have been reported [5]. We evaluated the case as primary thyroid cavernous hemangiomas because there are no history of trauma, previous FNAB or other invasive neck procedures.

Primary thyroid hemangiomas mostly seen as asymptomatic cervical mass. In the presence of intra-tumoral bleeding, fast growing masses could also be seen [6]. Most of the reported thyroid hemangiomas are located in left lobe, and are slightly higher in males [2]. Our case is women and the tumor was located in the right lobbe of the thyroid.

Due to no specific pathognomonic findings on USG, and FNAB or computed tomography, diagnosis of cavernous hemangioma of the thyroid mostly cannot be made preoperatively. It is suggested by Shpitzer et al., that magnetic resonance imaging, single photon emission computed tomography, digital subtraction angiography and red blood cell scans can be useful for the preoperative diagnosis of hemangiomas [7]. Though, these methods are not used routinely because of their high cost and inaccessibility [8]. Most of the published cases, were able to get diagnosed by histopathologic examination after surgery [9].

In thyroid hemangiomas, FNAB provides intensive blood cells and are insufficient for diagnosis. Although not useful in the diagnosis of thyroid hemangiomas, FNAB is recommended for excluding of the other thyroid tumors [2].

If a thyroid nodule is suspicious for malignancy or there is presence of signs of compression, surgical treatment is proposed. Total thyroidectomy or hemithyroidectomy could be carried out for the treatment [10]. Surgical treatment provides a good prognosis at thyroid hemangiomas.

CONCLUSION

In conclusion, primary cavernous hemangioma of thyroid is quite rare and benign. Preoperative differential diagnosis is very difficult. Surgery should be indicated when malignancy or cavernous hemangioma is suspected or compressive symptoms developed and it provides a good prognosis. A definitive diagnosis can only be achieved by postoperative histopathological evaluation.

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Author Contributions

Meryem Ilkay Eren Karanis – 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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Ilknur Kucukosmanoglu – Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published
Cevdet Duran – Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published
Alpaslan Sahin – 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 PEER REVIEWED | OPEN ACCESS

Late-onset thoracic aortic graft infection: A case report

Liran Shani, Yuval Geffen, Gil Bolotin, Ayelet Raz-Pasteur

ABSTRACT

Introduction: Thoracic aortic graft infection is a rare and devastating complication of aorta replacement surgery with an incidence range of 0.9–1.9%. Prosthetic aortic graft infections represent a major diagnostic and therapeutic challenge. Despite latest advancements in imaging and microbiological investigations there are still no agreed criteria to confirm the diagnosis. Case Report: We present a case of late onset culture negative thoracic aortic graft infection in a Caucasian 65-year-old male, nine years after aortic replacement due to acute aortic dissection. Conclusion: Culture-negative vascular graft infection has not been described as a clinical entity so far. Review of current literature and Issues of diagnosis and management will be discussed.

Keywords: Thoracic aortic graft infection, Culture negative vascular graft infection

How to cite this article

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INTRODUCTION

Thoracic aortic graft infection is a rare and devastating complication of aorta replacement surgery with an incidence range of 0.9–1.9% and carries mortality rates ranging from 25–75%. Although most cases appear shortly after the surgery there have been descriptions of cases manifesting up to 15 years after the initial procedure [1–3].

Since sepsis is a common complication in these patients, early diagnosis and treatment are important. It is often difficult to identify the primary infection site, and there are no consensus criteria for diagnosing the condition.

We present a case of late onset culture negative thoracic aortic graft infection in a male patient. Issues of diagnosis and management will be discussed.

CASE REPORT

A 65-year old male with 10 days of fever and malaise was admitted for evaluation to the department of internal medicine. Nine years earlier the patient suffered from acute aortic dissection without aortic valve involvement. He had undergone emergency replacement of the ascending aorta with a GORE-TEX aortic graft.

Fever started two days after an invasive dental procedure for which he was treated prophylactically with two doses of post-procedure amoxicillin. He continued to take his antibiotics under his dentist advice for 10 more days up until two days prior to his admission. His fever did not improve under the treatment. The patient had no history of contact with farm animals; he has not been traveling and did not consume out-of-routine food products including unpasteurized milk products. He had no respiratory, gastrointestinal or urinary symptoms. His other medical history included medically treated hypertension, prostatectomy following malignancy 15 years earlier treated with fesoterodine, smoking and venous insufficiency of the lower limbs.

On admission the patient was in good condition. Vital signs showed a blood pressure of 118/81 mmHg, irregular heart rhythm of 88 bpm, oxygen saturation level of 98% at room air, and an oral temperature of 37.7°C. His stature was obese with a body mass index of 35.6 kg/m^2.

Physical examination of the heart and lungs was normal. His abdomen was soft with no tenderness. He had bilateral chronic lower legs edema due to venous insufficiency with no signs of cellulitis. Laboratory findings of the blood on admission disclosed a slightly elevated white blood cell count (10.96x10^3/µL), normal hemoglobin (14.6 g/dl) and platelets level (162x10^3/µl), elevated CRP (103 mg/l) and erythrocyte sedimentation rate (70 mm/hr). BNP level was 130 Pico/ml.

ECG of the patient revealed a normal sinus rhythm with atrial premature beats, his chest x-ray showed no signs of consolidation.

Dental examination of the patient showed no abnormality, panoramic dental X-ray ruled out an infectious process.

Further workup included computed tomography scan of the chest, abdomen and pelvis. While his abdomen and pelvis CT scan showed no specific finding, on chest CT, a small amount of fluid was demonstrated around his ascending aorta which did not exist five years earlier during workup for chest pain.

Transesophageal echocardiography (TEE) demonstrated fluid surrounding the aortic graft, from the Sino tubular junction to the distal ascending segment of the graft. Suspected dehiscence of the proximal graft from its connection to ST junction was observed (Figure 1).

Positron emission tomography- computed tomography (PET-CT) scan demonstrated pathological absorption around the ascending aorta and the nearby fluid which corresponded to an infectious process (Figure 2).

Serologic panel taken included *Bartonella*, *Brucella*, *Syphilis* (RPR, TPHA) and *Coxiella burnetii*. *Bartonella henselae* IgG test was the only positive result. The patient had no contact with cats and did not meet the clinical criteria for *Bartonella* infection. PCR of blood and later aortic graft tissue excluded the diagnosis. Blood cultures, including long incubation periods and different culture mediums, were sterile.

Ciprofloxacin treatment was started four days after admission when low grade fever did not resolve and a positive urine culture yielded a susceptible bacterium. Two days later ciprofloxacin was replaced with ceftriaxone when fever did not resolve.

On the seventh day of his admission, under antibiotic treatment, the patient developed signs of sepsis with fever up to 39°C , elevated white blood cell count (30x10^3 cells/µl) and acute renal failure manifested with anuria and creatinine level of up 7.5 mg/dl. The patient was treated with careful fluid administration, diuretics for volume control and went through several dialysis sessions until signs of renal recovery were shown. Broad spectrum antibiotic regimen that included piperacillin/tazobactam and vancomycin (for the possibility of graft infection with Methicillin-resistant *Staphylococcus aureus*) was given to the patient. With signs of improvement from sepsis and renal failure treatment was replaced to a combination of ciprofloxacin, metronidazole and vancomycin (treatment of culture negative vascular graft infection). This regimen was maintained until his discharge.

Upon recovery, the patient was taken to the operating room. Surgical findings included an abscess between the superior vena cava and the proximal posterior part of the aortic graft. Structural integrity of the graft was maintained despite the infection. Under total circulatory arrest and hypothermia a new Dacron aortic graft was implanted after thorough debridement and rinsing. Samples were taken from the old graft and the abscess fluid.

Postoperative recovery was slow. Remarkable complications included deep venous thrombosis of the lower extremity. Renal function did not return to normal and the patient was discharged with a stable creatinine level of 2 mg/dl.

Pathologic examination of aortic graft segments extracted during surgery together with the fluid revealed

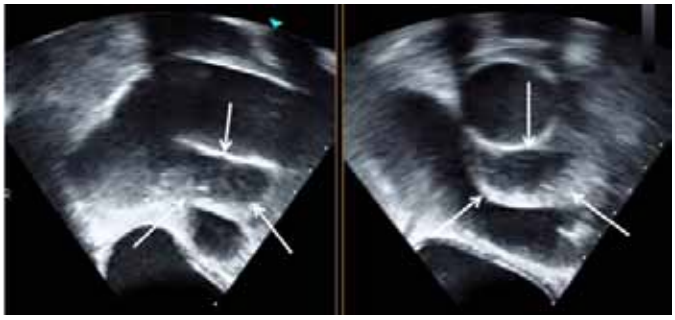

Figure 1: Transesophageal echocardiography (TEE) demonstrating accumaltion of fluid (arrows) behind the aortic graft, from the Sino tubular junction to the distal ascending segment of the graft.

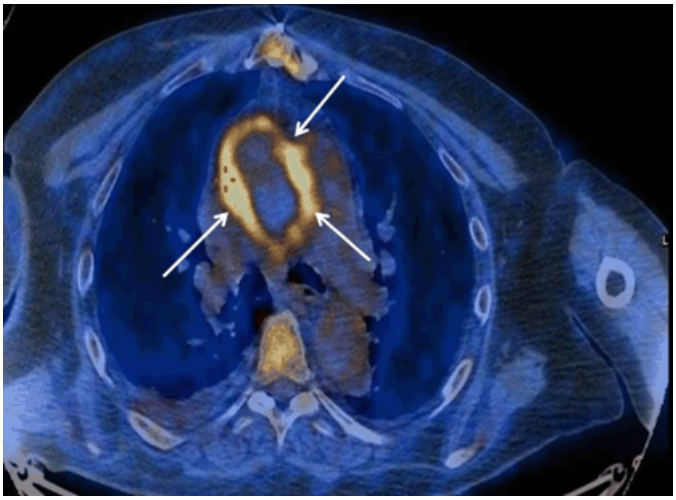

Figure 2: Positron emission tomography-computed tomography (PET-CT) scan demonstrating pathological enhanced absorption (arrows) around the ascending aorta and the nearby fluid which corresponded to an infectious process.

inflammation, necrosis and calcifications. Cultures of the tissue samples including the abscess fluid were sterile. PCR analysis was negative for bacteria (including specific testing for *Bartonella*) and fungi.

DISCUSSION

Four hundred and fifty thousand vascular grafts are being implanted annually in the United States with all graft types infection rate of 4% [4]. Diagnosis and management of thoracic aortic graft infections poses a major clinical challenge. Typical presentation is of a patient with no specific symptoms other than fever and malaise [5]. There is no consensus on diagnostic criteria or on the best management of aortic graft infection.

Several imaging modalities can be used to establish a diagnosis of aortic graft infection.

Computed tomography has a sensitivity of 94% and a specificity of 85% for diagnosing prosthetic graft infections in general [6]. MRI scan has been used on a limited scale to evaluate patients with suspected vascular graft infection. Shahidi et al. have demonstrated a sensitivity of 68% (95% CI 0.50-0.86) and specificity of 97% (95% CI 0.91–1.02) [7, 8].

Positron emission tomography-computed tomography (PET-CT) scan has been increasingly used to help in the diagnosis of vascular graft infection.

Table 1 demonstrates the sensitivity, specificity, positive and negative predictive value for several clinical series.

In general, the specificity of imaging for graft infection rises as time elapses form the surgical procedure [6].

Serological testing

Serology testing of patients' blood can aid in the diagnosis and proper treatment of the infection. Our patient's blood serology, taken almost two weeks after fever started, was positive for *Bartonella henselae* IgG. IgM result was negative. While IgM serology carries a high specificity rate for cat scratch disease, ranging from 89% to 96%, IgG serology demonstrates a lower 60–80% specificity rate for acute disease [9]. The rate of positive *Bartonella* IgG serology in the general population is 4–6% and there is evidence that the immune system's response to *Bartonella* infection varies considerably between patients [10]. Similar rules apply for other serological tests of pathogens like *Coxiella burnetii* and *Brucella*.

Tissue and blood PCR ruled out the presence of *Bartonella* infection in our patient.

Microbiology of aortic graft infections

Microbiology profile of infected aortic grafts is of somewhat similar to that of infected prosthetic cardiac valves with *Staphylococcus* species as the most commonly causative organisms. *Staphylococcus aureus* is usually more prevalent in early infection and coagulase-negative staphylococci such in late infections [11]. Gram-negative bacilli and *Enterococcus* species are regularly recovered from cultures as are anaerobes and fungi, but these often represent colonization when isolated from superficial wound swabs. In addition, sizable minorities (14%) of infections are polymicrobial [11]. However, many suspected aortic graft infections are treated without knowing the identity or antimicrobial susceptibilities of the causative organism, because suitable specimens were not obtained or because antibiotic treatment was instituted before the collection of appropriate samples for culture.

Culture negative vascular graft infection

Culture negative vascular graft infection has not been described yet as a clinical entity. The importance of discussing the clinical evidence in this field is emphasized in the era of endovascular procedures which considerably raised the numbers of prosthetic grafts introduced into the vascular system. The reason for failure to isolate and identify a causative agent in vascular graft infection is similar to that of culture negative endocarditis and includes prior antibiotic treatment and fastidious bacteria [12]. Technical reasons include unknown PCR inhibitors in the sampled tissue or inadequate sensitivity of PCR to detect the involved organisms. Great effort should be made to achieve the identity of the pathogen and to offer the patient the optimal antibiotic treatment.

Late-onset vascular graft infection

Late-onset vascular graft infection is defined as infection of the graft later than four months after the surgical procedure. Blood cultures are often negative. Special techniques such as broth culture or mechanical

Table 1: Sensitivity, specificity, positive and negative predictive value of PET-CT scan for the diagnosis of vascular graft infection

Study	Graft	No.	Sensitivity	Specificity	PPV	NPV
Fukuchi et al.	Multiple aortic grafts	33	91%	64%	56%	93%
Bruggink et al.	Multiple aortic grafts	25	93%	70%	82%	88%
Keidar et al.	Multiple aortic grafts	39	93%	91%	88%	96%
Tokuda et al.	Thoracic aortic graft	9	100%	80%	-	-

surface biofilm disruption by sonication or scraping, of the graft may be used to enhance the recovery of biofilm-forming organisms [11].

Late-onset vascular graft infection is a rare event. In a series of 41 patients, jones et al. presented 50% of cases in the first 500 days after the surgical procedure. Only four patients suffered an infection more than seven years after the surgery, only one of them had culture negative infection [11]. Coselli et al. published in their clinical series a single case of very late-onset (15 years post-surgery) thoracic aortic graft infection [13].

The etiology of late vascular graft infection is said to be implantation of bacteria at the time of initial surgery, but in a few cases infection may result from seeding onto the graft during a bacteremia. Factors influencing late graft infection include type of graft material and the identity of the offending pathogen. Dacron grafts are more likely to develop a partial 'pseudointima', making it less susceptible to late bacteremic seeding compared to polytetrafluoroethylene. Infections with high-virulence organisms are unlikely to remain dormant for years after the initial procedure [13]. Thus, late onset infection usually represents infection of originally implanted (during the first surgery) bacteria of low virulence nature or bacteremic seeding of high virulence bacteria.

Prevention of aortic graft infection

The American heart association and the European society of cardiology have issued updated guidelines regarding antibiotic prophylaxis before dental procedures for infective endocarditis prevention. According to the guidelines antibiotics should be offered only to high risk populations (Patients with a prosthetic valve or a prosthetic material used for cardiac valve repair, patients with previous infective endocarditis events, patients with congenital heart disease, in particular those with complex cyanotic heart disease and those who have postoperative palliative shunts, conduits, or other prostheses) [8].

There is no reference to antibiotic prophylaxis in people with vascular graft implantation.

Much like endocarditis prevention strategies, secondary prevention of vascular graft infection has not been investigated in a viable research model. The low incidence and multifactorial pathophysiology makes it difficult generating such a model. Jones at el. Have reported a series of 41 patients with vascular graft infection over a course of 26 years, out of those only four experienced the complication over 10 years after the initial procedure and the data collected was limited. These four patients had isolates of high virulence bacteria indicating a mechanism of recent bacteremia [13].

In 2002, Lockhart conveyed a Survey of infectious disease experts regarding antibiotic prophylaxis for medical conditions. Sixty percent (477) of the members of the Infectious Diseases Society of America Emerging Infections Network responded. 35% recommend prophylaxis for patients with prosthetic vascular grafts [14].

Our patient, a 65-year-old male, described going through an invasive dental procedure two weeks prior to his admission. Although not indicated by the ESC or AHA guidelines his dentist prescribed him amoxicillin but only after the procedure itself. While the patient is not classified as high risk population (due to the presence of thoracic aortic graft) the nature of the dental procedure does make him a possible candidate for antibiotic prophylaxis (which he got but not in the appropriate manner).

Surgical treatment

A combination of medical and surgical treatment is used for most infected vascular grafts. Infections of aortic grafts are treated with either axillofemoral bypass grafting, followed by excision of the infected graft, or graft excision plus in situ replacement with cryopreserved homografts, autologous vascular conduits, or if the infecting organism has low virulence (such as coagulase-negative *Staphylococcus*) prosthetic grafts. Ascending aortic grafts infections are commonly treated with in situ replacement of the infected graft [4].

CONCLUSION

Aortic graft infection remains to this day a catastrophic complication of aortic surgery. High suspicion and fast workup should be implemented for patients with an aortic graft presenting with signs of infectious disease at any period of time after the initial procedure. The term culture-negative vascular graft infection has not been coined yet and should be addressed as a clinical entity. Antibiotic prophylaxis that is given in the setting invasive medical procedures in patients with vascular grafts is largely unstudied. In the era of endovascular medicine and growing numbers of vascular grafts implanted, the current approach of withholding antibiotic prophylaxis for invasive medical procedure in this population may warrant reconsideration.

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Liran Shani – 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

Yuval Geffen – 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

Gil Bolotin – 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

Ayelet Raz-Pasteur – 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

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Authors declare no conflict of interest.

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CASE REPORT PEER REVIEWED | OPEN ACCESS

Recurrence of gastric cancer invading the main pancreatic duct: A case report

Hiroshi Maekawa, Hajime Orita, Mutsumi Sakurada, Tomoyuki Kushida, Tomoaki Ito, Koichi Sato

ABSTRACT

Introduction: Locoregional recurrence of gastric cancer is sometimes seen in clinical practice, but the finding of intraductal spread to the main pancreatic duct is unique. Here, we report a case of recurrent gastric cancer involving the main pancreatic duct caused by lymphatic spread into the pancreas. Case Report: A 76-years-old female was admitted to our hospital because of abnormality of the pancreas without symptoms. She had been treated with distal gastrectomy due to locally advanced gastric cancer 24 months before admission. Computed tomography scan showed a swelling in the pancreatic body containing a low-density area. MRI scan revealed that the low-density area of the central pancreas was the main pancreatic duct dilated and filled with a tumor. Resection of the gastric remnant with distal pancreatectomy and splenectomy were performed under a diagnosis of a primary pancreatic tumor or gastric cancer recurrence. Pathological examination revealed that a tubular adenocarcinoma packed the main pancreatic duct, and the same neoplasm infiltrating the pancreatic parenchyma was also found. Finally, we diagnosed the lesion as the lymphatic recurrence of gastric cancer. The patient had survived for 12 months since metastasectomy without signs of recurrence. Conclusion: Recurrence of gastric cancer sometimes invades to pancreatic parenchyma and mimics the intraductal pancreatic neoplasm. If the complete resection of locoregional recurrence of gastric cancer is performed, surgical treatment will contribute to prolonging the survival.

Keywords: Gastric cancer, Locoregional recurrence, Pancreas, Metastasis

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INTRODUCTION

Clinically, recurrences of radical gastrectomy are usually noticed as peritoneal dissemination and locoregional recurrence [1]. Pancreatic invasion with locoregional recurrence is sometimes seen in clinical practice, but the finding of intraductal spread to the main pancreatic duct is unique. Here, we report a case of recurrent gastric cancer involving the main pancreatic duct caused by lymphatic spread into the pancreas. Complete resection for locoregional recurrence will contribute to prolonging survival.

CASE REPORT

A 76-years-old woman was admitted to our hospital because of pancreatic abnormality during post-radical gastrectomy follow-up. She had received distal gastrectomy with Billroth II reconstruction for locally advanced gastric cancer of the gastric antrum 24 months before admission. Pathological findings of gastric cancer were tubular adenocarcinoma T3, N1 (only one positive node on proximal splenic artery), P0, H0, M0 stage IIIA according to UICC classification. After distal gastrectomy, she started the administration of S-1 at 80 mg/day every other day for adjuvant therapy. Twelve months after gastrectomy, the serum level of CA19-9 was 12 IU/ml. Follow-up CT scan was performed but it failed to reveal recurrence, showing only postoperative change on the pancreatic surface. Subsequently, the serum level of CA125 has been elevating up to 70 IU/ml. There have been no symptoms such as abdominal pain, appetite loss or abdominal distention. On admission, there was no abnormal mass palpable in the abdomen. Regarding laboratory findings, the serum level of CEA was 8 ng/ml, CA19-9 was 40 IU/ml, and CA125 was 90 IU/ml. The serum level of amylase was 51 IU/ml. Computed tomography scan demonstrated a three-cm low-density area in the pancreatic body, and the main pancreatic duct of the pancreatic tail was dilated (Figure 1). Neither swollen para-aortic lymph nodes nor abnormal ascites were noted. MRI scan revealed the existence of the tumor in the pancreatic body, and the tumor seemed to infiltrate the main pancreatic duct (Figure 2). ERCP was performed but failed to cannulate the main pancreatic duct. Positron emission tomography (PET) using ^{18}F-fluorodeoxyglucose demonstrated a hot spot superimposing on the pancreatic tumor. Although we could not confirm that the lesion was a primary pancreatic tumor or recurrence of gastric cancer, it was considered to be a potentially malignant lesion based on PET findings. We performed resection of the gastric remnant and distal pancreatectomy and splenectomy. Regarding the operative findings, the pancreatic tumor adhered to the remnant stomach and the splenic artery was involved. There was no peritoneal dissemination or swollen para-aortic nodes. The pancreas was cut on the left side of the portal vein. The remnant stomach and distal pancreas and spleen were resected en bloc. Concerning the pathological findings of the resected specimen, the pancreatic body was thickened and the main pancreatic duct was dilated with the tumor (Figure 3). The splenic artery was involved with the tumor. Histopathologically, the intraductal tumor was composed of tubular adenocarcinoma (Figure 4), and the parenchyma of the pancreatic body also contained tubular adenocarcinoma (Figure 5). The cancer showed infiltrative growth and invaded the remnant gastric wall. We compared the immunohistochemical characters of the resected tumor with those of the gastric cancer previously resected. Both tumors were positive for MUC6 and CK7, and negative for MUC2. With these findings, we finally diagnosed the patient with the recurrence of gastric cancer that had infiltrated the pancreas and spread into the main pancreatic duct. The surgical margins were all negative for cancer. The patient suffered from ileus after resection because of intra-abdominal infection due to postoperative pancreatic fistula (ISGPF grade B), and was discharged 30th postoperative day. She has shown no signs of recurrence for 12 months.

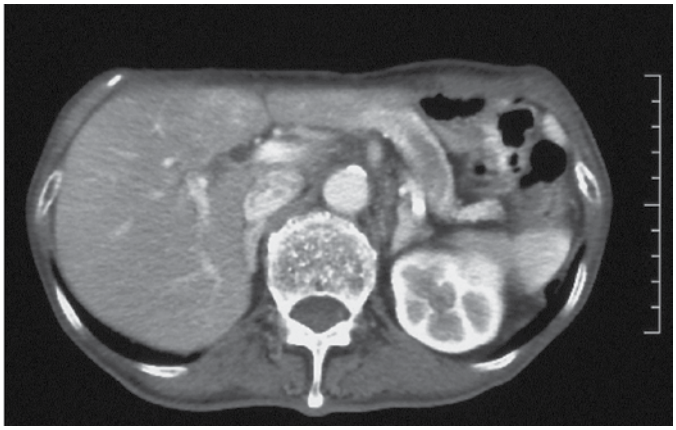

Figure 1: Computed tomography scan two years after distal gastrectomy. Abdominal CT scan demonstrating a low-density area in the pancreatic body, and the main pancreatic duct of the pancreatic tail is dilated.

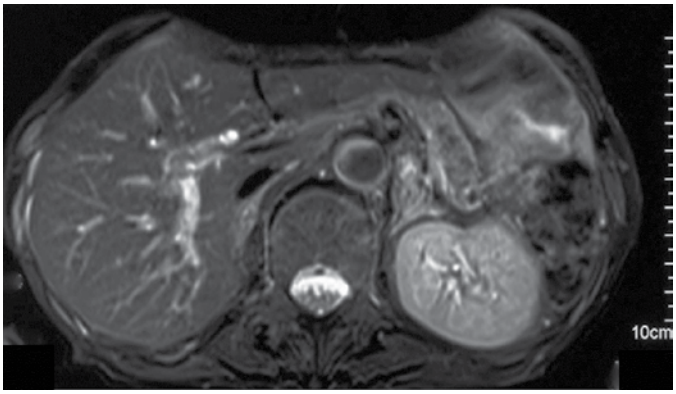

Figure 2: Magnetic resonance imaging scan two years after distal gastrectomy. T2-weighted MRI scan showed the existence of the tumor in the pancreatic body, and the tumor seemed to infiltrate into the main pancreatic duct.

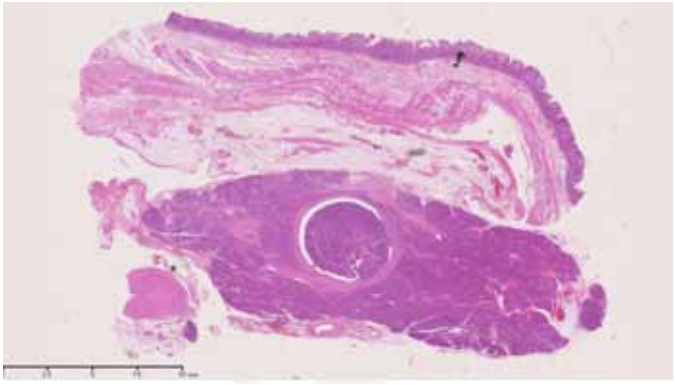

Figure 3: Pathological findings of the resected specimen on gross inspection (H&E stain, x40). The tumor cells infiltrated and packed the main pancreatic duct.

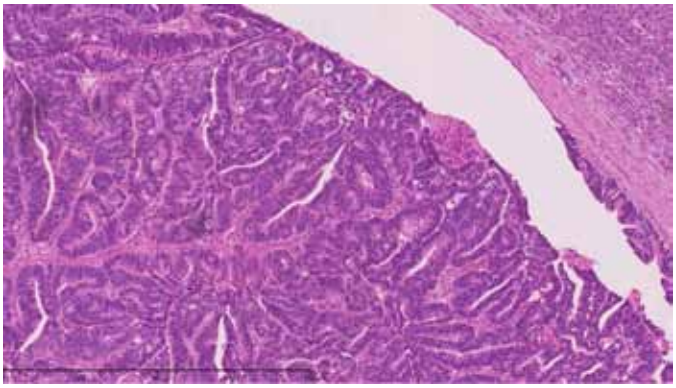

Figure 4: Histological findings of the tumor infiltrating the main pancreatic duct (H&E stain, x100). Histopathologically, the intraductal tumor was composed of tubular adenocarcinoma.

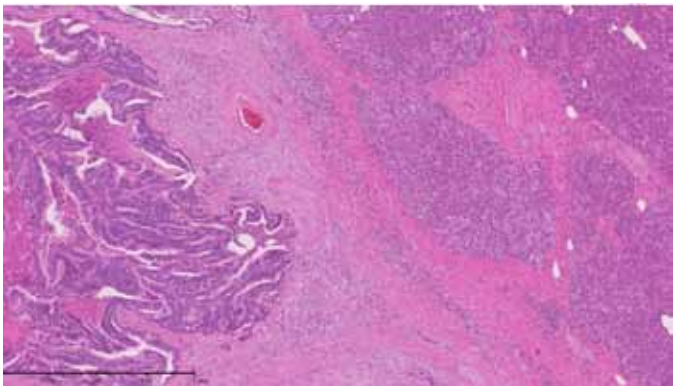

Figure 5: Histological findings of the tumor infiltrating the main pancreatic duct (H&E stain, x100). The parenchyma of the pancreatic body also contained tubular adenocarcinoma.

DISCUSSION

Recurrences often occurred in clinical practice after the curative resection of gastric cancer. Recurrence patterns following radical gastrectomy are classified as locoregional, peritoneal, and hematogenous. Wu et al. [2] reported that half of recurrence cases showed an initial single recurrence and 45% were locoregional recurrence. The incidence of recurrence patterns differs regionally throughout the world. Peritoneal recurrence is the most common pattern of recurrence in Asian countries [1], but locoregional recurrence is the most common in Western countries [3]. Locoregional recurrence is defined as recurrence at the resected margin, lymph nodes including para-aortic nodes, or in the surgical bed within the resection. The most common treatment for the recurrence of gastric cancer including locoregional recurrence is chemotherapy because recurrent gastric cancer is considered unresectable and early recurrence may occur after metastasectomy. However, in some cases of locoregional recurrence, if complete resection of the recurrent lesion is performed, the patients' survival may be prolonged [4]. Metastatic pancreatic tumors are relatively rare, with an incidence of 2–4.5% in all pancreatic tumors [5]. Most of them are hematogenous metastases of renal cell carcinoma [6]. Other than a hematogenous origin, local recurrence of malignancy from an organ adjacent to the pancreas is another origin of secondary malignancy of the pancreas. Advanced gastric cancer sometimes invades the pancreas, because the pancreas is adjacent to the stomach. In our case, the cancer cells remained in the surgical bed of the pancreatic surface or adventitia of the splenic artery where the positive lymph node existed. Also, the residual cancer cells infiltrated the pancreas and invaded the main pancreatic duct. The intraductal invaded cancer cells grew and finally packed the pancreatic duct. Maehara et al. [7] classified pancreatic invasion of gastric cancer into three types: invasion only to the pancreatic capsule, invasion to the capsule and interlobular tissues, and invasion to the capsule and intralobular tissues. It has been reported that the degree of serosal invasion and lymphovascular invasion may be associated with locoregional recurrence [8]. Our case showed the invasion to the intralobular tissues. It mimicked an intraductal tubulo-papillary neoplasm of the pancreas. We could not find the incidence of such intraductal spread appearance of pancreatic metastatic tumor in previous reports. The appearance of our case is considered to be a unique. Computed tomography (CT) scan is considered a useful examination for the detection of recurrence. However, CT-based diagnosis of recurrence can be difficult due to treatment-induced morphologic changes. In our case, CT scan demonstrated the thickness of the pancreatic surface, but we missed the fibrous scar on the surface of the pancreas showing recurrence. PET is another examination for detecting gastric cancer recurrence. The positivity and specificity of PET for gastric cancer recurrence are reportedly 75–85 and 77–90%, respectively [9]. In our case, the findings of PET/CT were helpful to decide on surgical resection. The prognoses of the patients with recurrent gastric cancer are poor even though chemotherapy regimens have been developed, but some recurrent gastric cancer patients with locoregional recurrence may show a prolonged survival time with surgical treatment. However, the surgical indication for locoregional recurrence should be limited to R0 resection. An approximately 20% five-year survival rate was expected with complete resection [10]. Intensive follow-up for advanced gastric cancer is necessary. We should not miss the opportunity to perform R0 resection for locoregional recurrence.

CONCLUSION

Recurrence of gastric cancer, if it invades to pancreatic parenchyma, sometimes mimics an intraductal tubulo-papillary neoplasm of the pancreas. If the complete resection of locoregional recurrence of gastric cancer is performed, surgical treatment will contribute to prolonging the survival.

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Author Contributions

Hiroshi Maekawa – 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
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Koichi Sato – 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

Guarantor

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Conflict of Interest

Authors declare no conflict of interest.

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CASE REPORT PEER REVIEWED | OPEN ACCESS

A rare presentation of calciphylaxis in normal renal function

Parin Rimtepathip, David Cohen

ABSTRACT

Introduction: Calciphylaxis is a rare and life-threatening condition in which extensive microvascular calcification in arterioles and occlusion of vessels lead to painful non-healing ulcers with high mortality rate. Calciphylaxis is mainly associated with end stage renal disease or hyperparathyroidism, with rare cases reported in cirrhosis patient. Case Report: A 55-year-old Caucasian male with significant history of porphyria cutanea tarda and Hepatitis C complicated by cirrhosis with normal renal function presents with history of non-healing ulcers on both of his hands. The diagnosis of calciphylaxis was made by X-ray. Due to rapid progression of the ulcers to tissue necrosis and gangrene with no definite underlying pathology, the patient's hands were eventually amputated due to the inability to withstand pain. Conclusion: Patients presenting with painful ulceration of their fingers with history of cirrhosis and normal renal function should be worked up for calciphylaxis as part of the differential diagnosis, especially with low serum albumin level. Site of calciphylaxis also matters as there is a great difference between mortality rates of proximal versus distal. We postulate the idea of pathophysiological mechanism with further research needed. This case report should alert physicians that calciphylaxis does occurred in patients with cirrhosis and normal renal function.

Keywords: Calciphylaxis, Cirrhosis, Porphyria Cutanea Tarda, Hepatitis C

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INTRODUCTION

Calciphylaxis is a rare and life-threatening condition in which extensive microvascular calcification due to calcium deposition in arterioles and occlusion of vessels lead to painful, violaceous, mottled skin lesions, which progress to non-healing ulcers, tissue necrosis, gangrene, sepsis, and potentially death [1]. Two years mortality rates from sepsis ranges from 50–80%. Patients with skin involvement over the trunk or proximal extremities have a poorer prognosis and higher mortality rate when compared to distal extremities. The lack of understanding the pathophysiology of the disease and numerous postulation result in unsatisfactory answer to why there is a difference in mortality rate. Biopsy of the calciphylaxis ulcer would reveals calcium deposits lining the vascular intima, while tissue calcification may also be seen on plain radiographs [2]. Risk factors for calciphylaxis are female gender, hyperphosphatemia with elevation of calcium phosphate axis, elevated parathyroid hormone, high alkaline phosphatase, and low serum albumin [3]. Many of the risk factors can be seen in patients on hemodialysis from chronic renal failure especially if the chronic renal failure is the complication of diabetes

with 61% had acral gangrene compared to 34% of the non-diabetic calciphylaxis [4]. Calciphylaxis is therefore most common in hyperparathyroidism secondary to chronic renal impairment and rarely occurs in the setting of normal renal function [2]. However, these risk factors are not set in stone or fundamental in diagnosing patients with calciphylaxis. We report an extremely rare case of calciphylaxis in a 55-year-old Caucasian male with significant history of porphyria cutanea tarda and hepatitis C complicated by cirrhosis with normal renal function that presents with non-healing ulcers on both of his hands, with rapid progression to tissue necrosis and gangrene.

CASE REPORT

A 55-year-old Caucasian male with significant history of hepatitis C, porphyria cutanea tarda (PCT), and cirrhosis presents with non-healing ulcers on both hands. The patient was referred to the dermatological center for the evaluation of both hands as a possible complication of PCT. The clinical examination showed a cachectic patient with lesions shown in Figure 1. The patient noticed the new skin lesions on his hands to be different from the lesions of PCT several years ago. Even though PCT caused pain, the new onset of the non healing ulcers were far more painful. The patient described the ulcer as they first appeared as shallow eroding of the skin. Then the ulcers would accompanied by extreme pain and the tissue in the area would started to necroses. Some of the differential diagnoses for the patient's painful ulceration of the fingers were Raynaud syndrome, scleroderma, peripheral vascular disease, and vasculitis. During the course of treatment, patient denied any skin biopsied due to the existence of severe pain. Therefore, we decided to perform X-ray of his left hand. The finding of his left hand (Figure 2) showed mildly increased soft-tissue density and stipple calcification of the vessels. Calciphylaxis was diagnosed.

Due to the unknown pathogenesis which makes good clinical treatment extremely difficult, labs were ordered in an attempt to exclude any risk factors. All labs including CBC and biochemistry were normal except the low albumin level of < 3 g/dL. The patient was given analgesic for his pain but increase dosage did not alleviate his condition. Patient's fingers were ultimately amputated due to unbearable pain and unsalvageable tissue necrosis and gangrene.

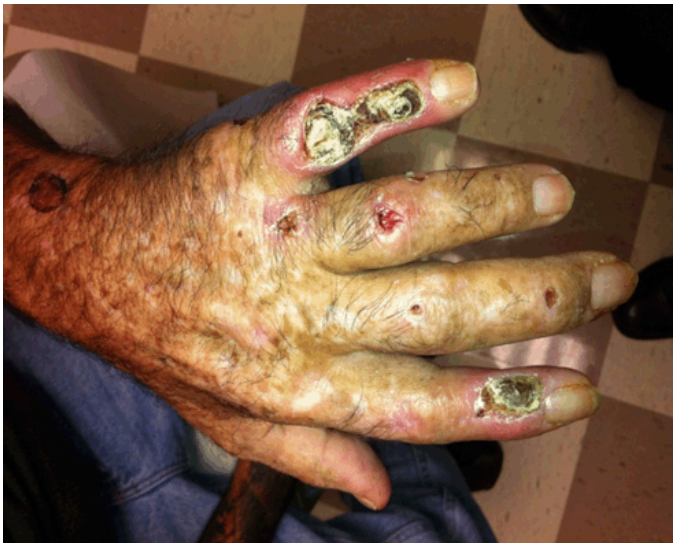

Figure 1: Presentation of calciphylaxis on patient's left hand with characteristic of ulceration, tissue necrosis, and gangrene.

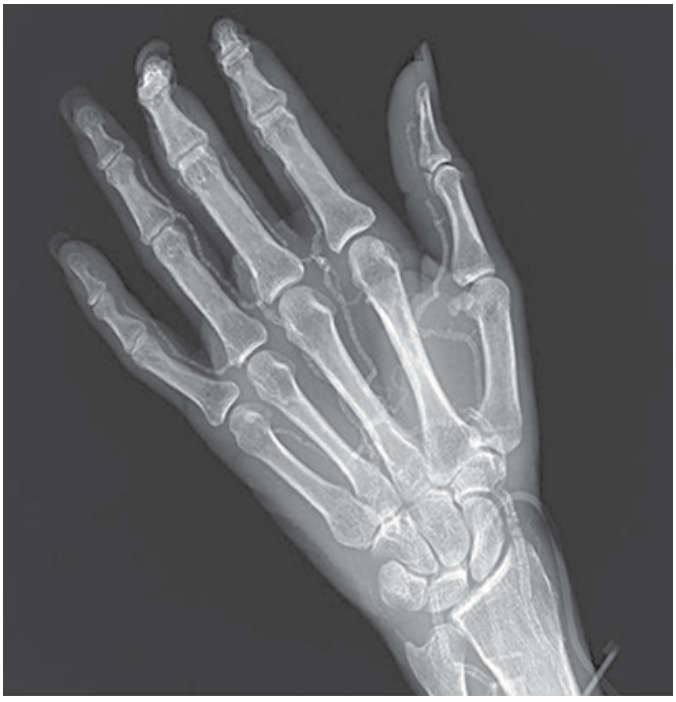

Figure 2: Presentation of calciphylaxis on X-ray showing mildly increased soft-tissue density and stipple calcification of the vessels.

DISCUSSION

Calciphylaxis (synonym calcific uremic arteriolopathy) results from calcification of the arterioles and subsequent thrombosis which lead to skin ischemia. Calciphylaxis is associated with high morbidity and mortality resulting primarily from infections. According to Mazhar et al., elevated parathyroid hormone levels, elevated serum phosphate and calcium-phosphate products especially in renal failure patients, elevated alkaline phosphatase, high serum levels of iPTH, low serum albumin, female, Caucasian origin, ESRD, medications such as warfarin, prednisone, calcitriol, and calcitriol salts are associated with risk factors at the time of diagnosis of calciphylaxis. Since most calciphylaxis typically occurs in patients with end-stage renal disease undergoing dialysis or patients who have secondary hyperparathyroidism, patients without these underlying risk factors are often misdiagnosed at presentation [3]. An important postulated

pathophysiologic mechanism is proposed by Danziger and Demer about the increase risk for calciphylaxis in patient with hepatic disease, like our presenting patient. A possible reason is that vitamin K is reduced in liver disease, which is required for post-translation gamma-carboxylation of matrix gamma-carboxyglutamic acid protein, fetuin, or growth arrest-specific gene 6. These are calcifications inhibitors produced by vascular smooth muscle cells. Warfarin, which inhibits vitamin K-dependent carboxylation of these calcification inhibitors, is thought to encourage vascular calcification in this way [5-6]. Another propose mechanism by Goli, Shah, Byrd, and Roy, is the deficiencies in protein C and S from cirrhosis as the cause of calciphylaxis as low levels of these anticoagulant factors may be an important etiologic factor [7].

Low serum albumin level is one of the main interests as a risk factor for our patient since all lab values were normal except the albumin level. Bleyer et al. reported a 17-fold increase in the risk of developing calciphylaxis with each decrease in albumin by 1 g/dL (OR= 16.9, 95% CI, 5.25 to 54.5) [8]. This finding is later supported by Coates et al., who reported a loss of 10 % body weight over six months preceding the diagnosis of calciphylaxis in 7 out of 16 patients in their series [9]. As mentioned earlier, two years mortality rates from infections and eventually sepsis for these patients might be due to underlying poor would healing as a result of low albumin. Correcting albumin level in patients with calciphylaxis with no other risk factors might enhance their survival rates by enhancing the wound healing process, which would lead to less infection and less mortality rate. However, further research concerning the possible role of albumin level should be pursued. Suyin and I.H. Coulson proposed a summary of recommended investigations in Table 1 [2].

Why do patients with skin involvement over the trunk or proximal extremities have a poorer prognosis and higher mortality rate when compared to distal extremities which occurs below the knee? Is the fact that our patient beat the two years mortality rates of 50–80% because only his distal extremity was involved? The lesions of calciphylaxis typically develop suddenly and progress rapidly. However, does this concept apply to distal calciphylaxis with better prognosis? According to Mazhar et al., eleven patients with calciphylaxis and seven controls died during the follow-up period. Seven out of ten patients who died of complications related to calciphylaxis had proximal lesions and died of infections [3]. Proximal lesions have been observed in many studies to be one of the worst prognoses, but none of the articles postulated the underlying pathophysiologic mechanisms. A raising question surrounding this area: Is it because proximal type of calciphylaxis involves major arteries while distal type involves arterioles? In humans, vascular calcification is an active process and is not sufficient to produce skin necrosis. Vascular calcification and thrombosis are both required to produce lesions of calciphylaxis [10]. According to Rayz, V. L., et al., in addition to biochemical factors, hemodynamic factors that are governed by luminal geometry and blood flow rates likely play an important role in the thrombus formation and deposition process [11]. Arteries and arterioles have different luminal geometry and blood flow rates, therefore, calcification and thrombus formation can possibly formed much faster and much more aggressive in the proximal calciphylaxis. Also, the proximal arteries increase the chance of infections developing into sepsis because they are closer to many important organs. The idea behind this theory stimulates further research concerning the difference between mortality rates in proximal versus distal calciphylaxis.

Table 1: Summary of Recommended Investigations [2]

Full blood count
Urea and Creatinine
Liver function
Corrected calcium
Phosphate
Calcium-phosphorus index (<55 mg2/dL2)
Parathyroid hormone
Coagulation profile
Thrombophilia screen (factor V Leiden, anti-cardiolipin Ab, lupus anticoagulant, protein c, protein s, homocysteine)
Skin biopsy
Plain radiographs
Technetium-99 scintigraphy to exclude visceral calcification

CONCLUSION

In conclusion, patients presenting with painful ulceration of their fingers with significant history of porphyria cutanea tarda and Hepatitis C with cirrhosis with normal renal function should be worked up for calciphylaxis as part of the differential diagnosis. Calciphylaxis is a very rare disease with approximately 160 case reports worldwide. This case report should alert physicians that calciphylaxis does occurred in patients with cirrhosis and normal renal function.

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Author Contributions

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

David Cohen – Substantial contributions to conception and design, 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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CASE REPORT PEER REVIEWED | OPEN ACCESS

A case of classic paroxysmal nocturnal hemoglobinuria

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Ramakrishnan S. R., Giridhar Muthu

ABSTRACT

Introduction: Paroxysmal nocturnal hemoglobinuria (PNH) is an acquired hemolytic anemia characterized by a triad of intravascular hemolysis, pancytopenia and tendency for venous thrombosis. Patients with PNH present with these features which occur in various combinations as described in this case report. Several episodes of intravascular hemolysis result in hemoglobinuria associated with thrombosis at unusual sites and these patients may have varying degree of bone marrow disorders. Diagnosis can be confirmed by flow cytometry of blood granulocytes and FLAER assays. Management was supportive with transfusion and treatment of thrombosis in the past. But in the recent years the evolution of treatment strategies like hemopoietic stem cell transplantation and complement inhibition with eculizumab though very costly have been shown to be very effective. Case Report: Here we report a young girl who presented with abdominal pain, distension with a history of headache and jaundice. On evaluation, we found there was bicytopenia with evidence for hemolytic anemia and venous thrombosis of cerebral venous sinuses, hepatic veins and intrahepatic portion of IVC. With these clinical features, we suspected paroxysmal nocturnal hemoglobinuria which was later confirmed by flow cytometry. Conclusion: Having diagnosed her disease, we had to decide on various treatment options like eculizumab, hemopoietic stem cell transplantation which are efficient therapies for PNH. When these modalities are not possible in our case we had to adopt conservative management.

Keywords: Budd–Chiari syndrome, Eculizumab, Multiple venous thrombosis, Paroxysmal nocturnal hemoglobinuria (PNH)

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INTRODUCTION

Paroxysmal nocturnal hemoglobinuria (PNH) is an acquired hemopoietic disorder which is a rarity in occurrence. Available reports suggest that the incidence of clinically significant disease is in the range of 1 to 10 cases per million population and it is chiefly a disease of adults and the peak age of onset is in thirties [1]. Though PNH is caused by mutation of a gene on X chromosome it affects males and females equally [2]. This disease is classified under acquired hemolytic anemia with constellation of certain clinical findings. They present with clinical features of unexplained hemolytic anemia like fatigue, jaundice and red colored urine. Thrombosis involves

venous rather than arterial system and the presentation depends upon the site of thrombosis like hepatic, portal, mesenteric and cerebral veins. The delay in the diagnosis of this disease may be either because of the disease being rare or due to nonspecific clinical features. Prompt and accurate diagnosis is important as effective therapies have become available. This has become very much possible because diagnostic testing has evolved significantly due to the better understanding of the molecular basis of the disease and eventually the pathogenesis of hemolysis in PNH. We present a 21-year-old female with combination of symptoms and signs that made us to diagnose this rare disorder and also we have discussed the difficulties in the management of this patient.

CASE REPORT

A 21-year-old female presented with complaints of jaundice for one month; fever abdominal pain and distension for 15 days. She was admitted in an outside hospital with complaints of left sided headache, blurring of vision in the right eye for one day with no history of any significant illness in the last or chronic drug intake. Her menstrual cycles have been irregular for the last two years. She had a younger brother who is healthy. There she was found to have anemia with thrombocytopenia. MRI scan of BRAIN revealed left parietal and occipital hemorrhages (Figure 1). Workup for connective tissue diseases like ANA, dS-DNA and APLA were done and found to be negative. Bone marrow biopsy was done and revealed hypercellular marrow with no other abnormality.

As they were unable to pin point the crux of the problem, she was referred to our institution for persistent fever, abdominal pain and distension with persistent headache. On examination she was afebrile, pulse rate 80/min, blood pressure 110/70 mmHg, marked pallor was present. Cardiovascular and respiratory examination were normal. Her abdomen was soft with minimal distension and diffuse tenderness. She also had hepatomegaly which was 3 cm below the right costal margin and presence of shifting dullness. Her CNS examination revealed no focal neurological deficits. Fundus Examination showed few superficial hemorrhages in the retina of right eye with visual acuity 6/6.

Initial laboratory tests (Table 1) revealed reduced hemoglobin, low platelets, raised LDH, slightly elevated bilirubin along with transaminases (AST>ALT), negative direct coombs test and inconclusive marrow. Urine had grown *Enterococcus faecalis*.

Ultrasonography (USG) of abdomen showed thrombosis involving intrahepatic segment of Inferior vena cava and hepatic confluence. There was also hepatomegaly with coarse echotexture with ascites. Since thrombosis was made out we worked her up for thrombotic states. Homocysteine level was normal. ANA, Antiphospholipid antibody, Anti cardiolipin anti body and lupus anticoagulant were negative. Then we proceeded with CECT scan of abdomen which was consistent with USG abdomen and was suggestive of Budd–Chiari syndrome (Figure 2A–B).

Table 1: Laboratory investigation of the patient.

Hemoglobin	5. 2 g/dL%
Total count	5900
Differential count	Polymorph 64, Lymphocyte 27, Monocyte 7, Basophil 0. 6, Eosinophil 0. 4
Platelets	1. 3 lakhs/mm3
Peripheral Smear	Normocytic normochromic RBCs, Adequate platelets, MP,MF: Negative
Mean corpuscular volume	91. 9 Fl
Mean corpuscular hemoglobin	28. 6pg
Mean corpuscular hemoglobin concentration	31. 1
Erythrocyte sedimentation rate	55 mm/hr
Serum Iron	94 mg/dl
Serum ferritin	67. 50 ng/dl
Total iron binding capacity	463
Prothrombin time	14. 1
Partial thromboplastin time	41. 1
International normalized ratio	1. 34
Lactate Dehydrogenase	1849 U
T. BILIRUBIN	1. 5 mg/dl
Aspartate aminotransferase	202 U/L
Alanine aminotransferase	135 U/L
Urea &creatinine	Normal
Thyroid profile	Normal
Stool Occult blood	Negative
Direct Coomb's Test	Negative
Viral markers(HBsAg, anti HCV, HIV)	Negative
Urine routine examination	Albumin 1+, hemoglobin +
Blood culture	No growth
Urine culture	Enterococcus faecalis (colony count 105)
Bone marrow	Reactive marrow, Megaloblastic maturation with binucleate forms, All stage of maturation with giant metamyelocyte

In order to know the cause of persistent headache, we did MRI scan of brain with venogram which showed sub acute hemorrhage of size 3.5x2.5 cm in left occipital lobe and absent flow was noted in left transverse, sigmoid sinuses and upper jugular vein (Figure 3) suggestive of cerebral venous thrombosis. Later ophthalmologists

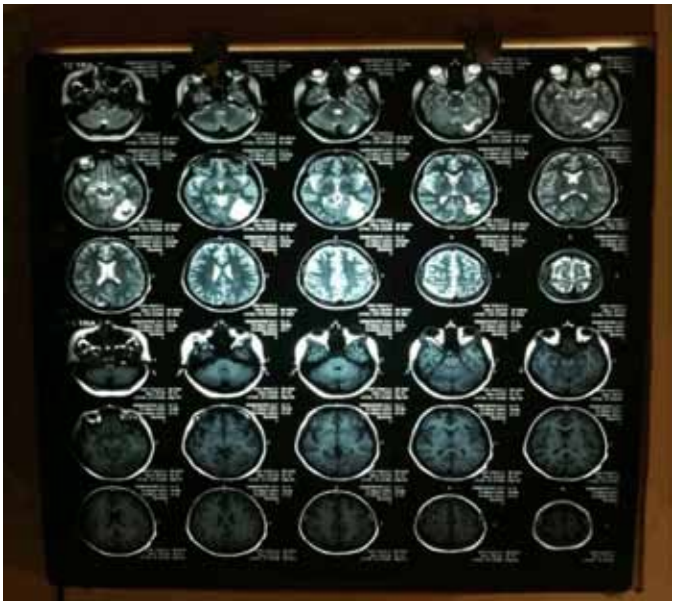

Figure 1: Magnetic resonance imaging scan of brain showing multiple areas of hemorrhages in left lobe of cerebellum, parietal and occipital lobes.

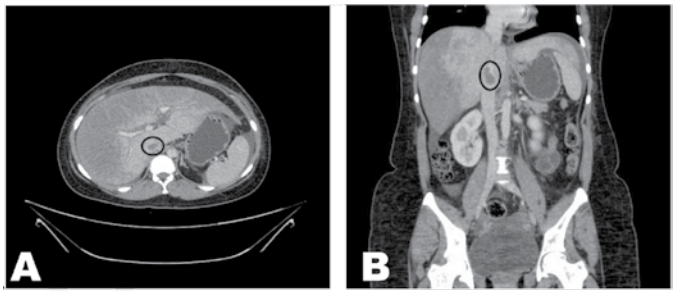

Figure 2: (A, B) Computed tomography scan of abdomen-Intra hepatic portion of inferior vena cava showing thrombosis as shown in circle.

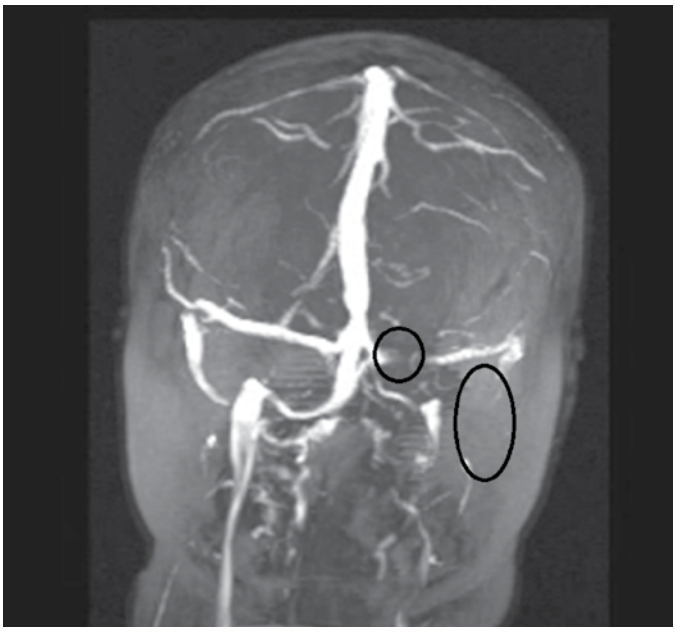

Figure 3: Thrombosis involving left transverse sinus, sigmoid sinus and upper jugular vein.

suspected branched retinal vein occlusion and advised fluorescein angiogram but patient was not willing for the procedure.

Our patient had hemolysis as evidenced by raised LDH, thrombosis at multiple sites (IVC, cerebral venous sinuses and possibly retinal vein) and bicytopenia-anemia and thrombocytopenia. This triad of features made us to suspect PNH. In order to confirm the same, we did flow cytometry which showed evidence of PNH clone upon analysis of granulocytes and monocytes [CD 59--47% NEG (>20% NEG in granulocytes) and CD 55–56.7% NEG.

With the classical triad of features and a positive flow cytometry final diagnosis of paroxysmal nocturnal hemoglobinuria was made. Hematologist opinion was sought and patient was started on LMWH, enoxaparin 60 mg subcutaneous twice daily for five days overlapped with oral anticoagulant, acenocoumarol 4 mg which was continued. She was also given three units of packed cell transfusions and appropriate antibiotics for UTI.

As a definitive therapy her brother was worked up for allogenic hemopoietic stem cell transplantation. HLA typing was done initially which didn't match with the patient. Hence we could not proceed further with bone marrow transplantation. Other option was eculizumab C5 complement antagonist which was unaffordable by the patient. Patient's hemoglobin and platelet count improved by then and she was discharged with acenocoumarol 4 mg daily, later adjusted according to PT, INR, folic acid 2 mg daily and ferrous fumerate 300 mg twice daily

On subsequent follow-up patient used to complaint of abdominal distension and pain on and off. Hemogram showed well preserved hemoglobin, leukocyte and platelet counts. Repeat USG abdomen showed the persistence of thrombosis in hepatic vein and intra hepatic portion of inferior vena cava and ascites. When patient has been followed up for almost two years we found that she had persistent hepatic vein thrombosis and developed features of cirrhosis; but she did not develop thrombosis at any other sites.

DISCUSSION

We report a young girl who presented with headache, abdominal pain and distension with past history of jaundice was found to have bicytopenia and venous thrombosis at multiple sites. Diagnosis of PNH was confirmed by typical clinical features and flow cytometry.

In PNH there is complement induced lysis of RBCs due to the abnormal sensitivity of RBC cell membrane. This is due to an acquired defect in the gene for phosphatidylinositol class A (PIG A) thereby causing deficiency of glycosylphosphatidylinositol (GPI) which is sheet anchor for cell membrane proteins [3]. CD55 and CD59, complement regulatory proteins which block intravascular and extravascular hemolysis respectively in normal human, are deficient in PNH [4]. Hemolysis

occurs in PNH because these patient's RBC's lack GPI anchor which is required to attach CD55 and CD59 to the surface of RBC [4]. This permits unregulated formation of certain complement attack complex which damages RBC membrane resulting in intravascular hemolysis. This causes reduction in hemoglobin and hemoglobinuria with resultant increase in LDH [3]. Next feature is thrombosis which is the leading cause of death in patients with PNH [4]. The pathogenesis causing thrombosis is not completely understood; but hypothesized to be due to free hemoglobin resulting from hemolysis attracts nitric oxide which induces vasoconstriction and damages the vascular endothelium forming a nidus for thrombus formation. Also platelets release procoagulant particles during complement induced hemolysis, which facilitate thrombosis. Thromboses involve the venous rather than the arterial system [4]. Venous thrombosis often occurs in locations such as hepatic, portal, mesenteric, dermal, and cerebral veins [5]. Minority of patients develop pancytopenia due to bone marrow disorders like aplastic anemia or primary myelofibrosis.

PNH is classified into classic PNH (presence of hemolysis with no marrow abnormality), PNH with marrow disorders(aplastic anemia/myelodysplastic syndrome (MDS)/primary myelofibrosis (PMF) and subclinical PNH–without clinical evidence [6]. Before making the final diagnosis of PNH, we have to rule out other hemolytic anemias like autoimmune anemias, hereditary anemias, drugs/toxin induced anemias, microangiopathic hemolytic anemias and bone marrow disorders like aplastic anemias, MDS and myelo fibrosis. Abdominal or cerebral vein thrombosis due to PNH must be differentiated from other hyper coagulable states and thrombophilias. The diagnosis of PNH can be suspected when we come across cases of coombs negative hemolytic anemia or confusing cases of pancytopenia.

The established therapies for patients with classical PNH are allogeneic hematopoietic cell transplantation (HCT) and complement inhibition with eculizumab [3]. Patients with hemolysis are better managed with eculizumab [7]. Patients with thrombosis are managed with therapeutic anticoagulation and eculizumab. Most of the patients will not be able to access this therapy due to its high cost. Allogeneic HCT is advised for patients with severe cytopenias, patients with poor response to eculizumab or when not accessible to eculizumab [3]. Supportive therapy includes red blood cell (RBC) transfusions, supplemental iron and folic acid (1 to 2 mg daily).

Our patient had features of classical PNH-bicytopenia, hemolysis and venous thrombosis at three sites intraabdominal cerebral and retinal with no marrow involvement. We learn that it is difficult to diagnose this disease unless we have a high index of suspicion. We present this case due to its rarity and the difficulties we had in diagnosing and the management when both bone marrow transplant and eculizumab were not feasible.

CONCLUSION

In this presentation, a young girl who presented to us with bicytopenia and hemorrhagic cerebral infarct with recent history of jaundice was found to have coombs negative hemolytic anemia and multiple venous thrombosis (hepatic, cerebral, retinal). It has always been said that in a case of confusing cases of hemolytic anemia and pancytopenia,we must suspect paroxysmal nocturnal hemoglobinuria (PNH); more so when it is coupled with venous thrombosis. Having diagnosed PNH, the management recommended is very costly and should be affordable for the patient.

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Krishnamoorthy Seetharaman – Substantial contributions to conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article, Critical revision of the article and Final approval of the version to be published
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Giridhar Muthu – Acquisition of data, Analysis and interpretation of data, Critical revision of the article and Final approval of the version to be published

Guarantor
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Authors declare no conflict of interest.

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SUGGESTED READING

- Medicine up-to-date, Harrison's principles of internal medicine, CMDT-2014, Davidson's principles and practice of medicine.

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CASE IN IMAGES PEER REVIEWED | OPEN ACCESS

Successful bail-out stenting of severe stenosis of the left main trunk coronary artery using guiding catheter exchange with the anchor balloon technique

Daizaburo Yanagi, Takeshi Serikawa, Masanori Okabe, Yusuke Yamamoto

ABSTRACT

Introduction: Trans radial intervention (TRI) is less invasive. However, percutaneous coronary intervention (PCI) operators may be concerned that trans femoral approach (TFI) is better than TRI according to the state of the patients, for example the patients with acute coronary syndrome (ACS) under the shock state, with severe winding subclavian artery and with the spasming radial artery. Case Report: We herein report a case of an unstable angina and acute heart failure. Coronary angiography (CAG) revealed evidence of 90% ostial stenosis of the left main trunk (LMT). But we were unable to engage a 6 Fr guiding catheter (GC) because of severe tortuosity of the left subclavian artery. Therefore, we attempted intracoronary passage of a 4 Fr JL3.5 catheter exchange the 4 Fr diagnostic catheter with a 6 Fr GC using an extension wire. However, before entering the left coronary artery, the guidewire coiled around the catheter, which prolapsed; therefore, the 6 Fr GC could not be engaged. We carefully inserted a 3.0-mm semi-compliant balloon up to the LMT lesion without GC support and were able to engage the GC by the anchor balloon technique. The process took approximately 5 s and the patient's hemodynamic state were not affected. TRI or a downsizing stenting system is essential for patients in whom the approach site is limited in size. Conclusion: The use of an extension wire after insertion of the diagnostic catheter and the anchor balloon technique has been successful in limited cases when insertion of GC is difficult.

Keywords: Trans radial intervention, Acute coronary syndrome, Left main trunk, Anchor balloon technique, Guiding catheter exchange

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INTRODUCTION

Although the transradial intervention (TRI) is increasingly used globally for coronary angiography and interventions, performing percutaneous coronary intervention (PCI) in arteries with complex anatomy remains a clinical problem. In particular tortuosity within a subclavian artery is frequently encountered

and can hamper delivery of guiding catheter to coronary artery. The management of these conditions remains controversial, with only a few reports in literature.

CASE REPORT

A female in her 60s complaining of severe chest pain and dyspnea was admitted to our hospital with a diagnosis of unstable angina and acute heart failure. Chest radiography revealed pulmonary congestion, echocardiography showed evidence of markedly decreased ventricular wall motion in all ventricular walls except the inferior wall, and electrocardiography demonstrated both marked ST-segment depression in precordial leads and ST elevation in the aVR lead (Figure 1). Risk factors for coronary artery disease, including diabetes mellitus and dyslipidemia, were noted. Access to the same region was not possible because the patient underwent right femoral artery bypass surgery five years earlier. After the insertion of an intra-aortic balloon pump (IABP) into the left femoral artery, we inserted a 4 French (Fr) sheath into the right radial artery. At this time, coronary angiography revealed evidence of 90% ostial stenosis of the left main trunk (LMT) (Figure 2). However, because of spasm in the right radial artery, we changed to the left radial artery. Using a long sheath, we attempted to continue percutaneous coronary intervention (PCI), but were unable to engage a 6 Fr guiding catheter (GC) because of severe tortuosity of the left subclavian artery. Therefore, we attempted intracoronary passage of a 4 Fr JL3.5 GC to exchange the 4 Fr diagnostic catheter with a 6 Fr GC using an extension wire (Figures 3-1, 3-2, and 3-3). However, before entering the left coronary artery, the guidewire coiled around the catheter, which subsequently prolapsed; therefore, the 6 Fr GC could not be engaged. We carefully inserted a 3.0-mm semi-compliant balloon up to the LMT lesion without GC support and were able to engage the GC by dilating the balloon and using the anchor balloon technique (Figures 4-1, 4-2, 4-3). The process took approximately 5 s, and hemodynamics were not affected. The procedure was concluded after placement of an XIENCE V® Everolimus Eluting Coronary stent (Abbott Laboratories, Abbott Park, IL, USA), and stent apposition was confirmed by intravascular ultrasound (IVUS; Figures 5–7).

Fluoroscopy time was 40 min, and the radiation exposure dose was 1.2 Gy. A total of 220 ml of radiocontrast medium was used and the total procedure time was 80 min. The patient was subsequently admitted to the coronary care unit, where heart function rapidly improved (Figure 8).

The patient was subsequently given an ambulatory discharge on hospitalization day 14. Her postoperative course was uneventful. An angiography performed at the 2-month follow-up showed no recurrence of stenosis (Figure 9).

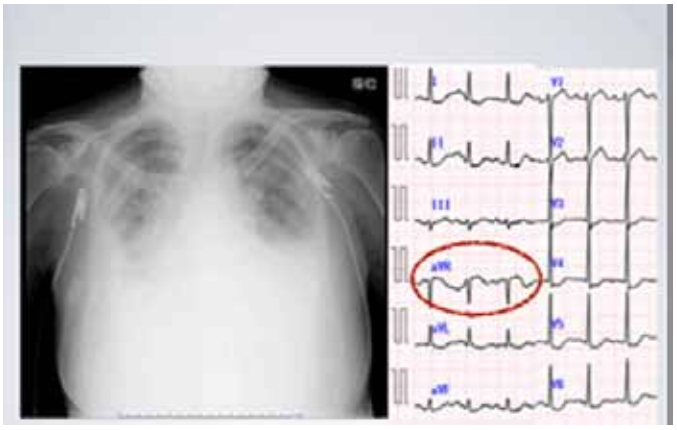

Figure 1: Chest X-ray showing the cardiomegaly and butterfly shadow. ECG showing the ST depression in V2-V6 leads and S Televation in a VR.

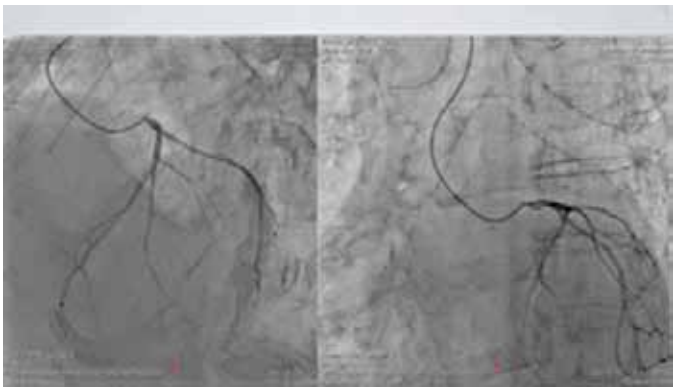

Figure 2: CAG showing severe stenosis at left main trunk ostium.

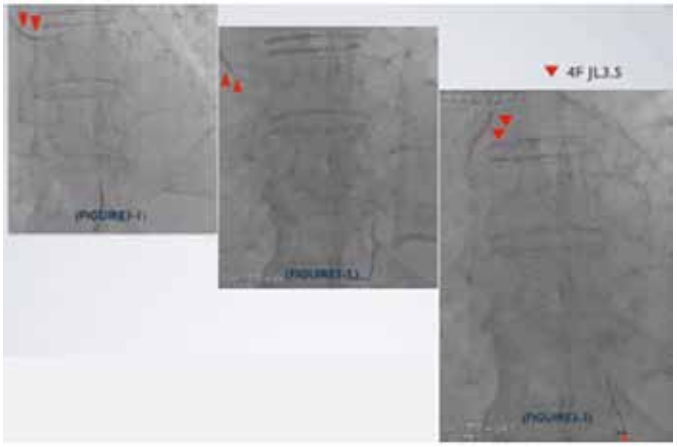

Figure 3: Guiding catheter exchange. The 4F JL diagnostic catheter could be inserted, however 6F JL catheter could not. Because Left subclavian artery and ascending aorta were very tortuous. The exchanging from 4F JL 3.5 to 6F JL3.5 using extension wire was performed.

DISCUSSION

The patient was admitted with post-acute coronary syndrome (ACS) with complicating cardiogenic shock and underwent right femoral artery bypass surgery five years earlier. Transradial intervention (TRI) was performed

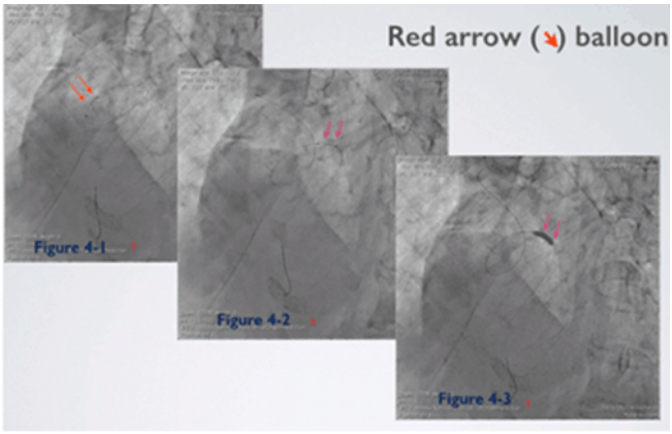

Figure 4: Anchor balloon technique. Guiding catheter position was subclavian artery because of the guide wire was prolapsed, and 3 mm balloon was advanced gently without a guiding catheter support. Finally, we inserted Guiding catheter using anchor balloon technique an left main trunk.

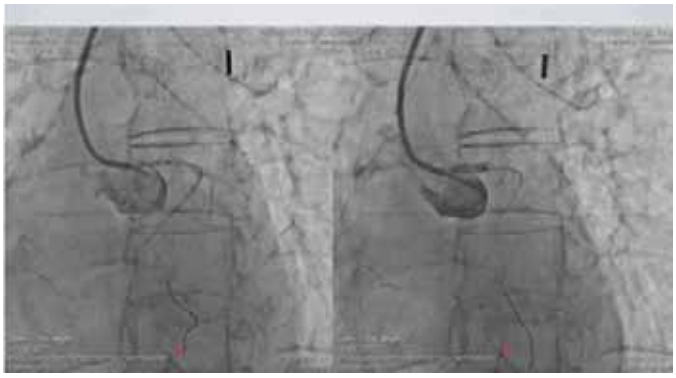

Figure 5: Successful implantation a stent in the left main trunk ostium.

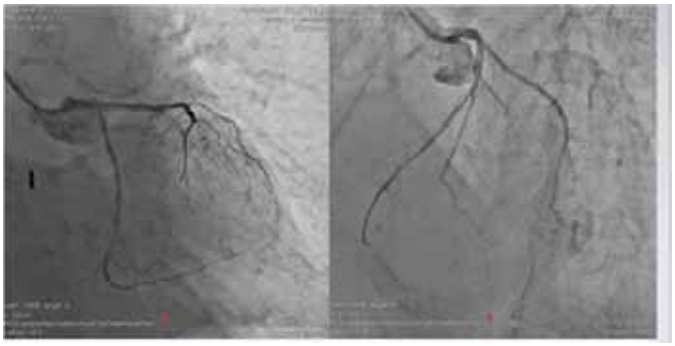

Figure 6: Final coronary angiography (CAG) showed the stent dilated sufficiently and TIMI3 flow.

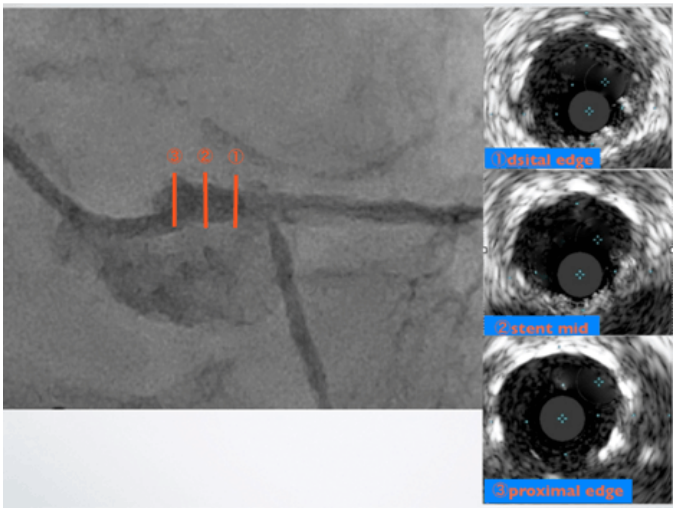

Figure 7: Final intravascular ultrasound (IVUS) showed the stent apposition.

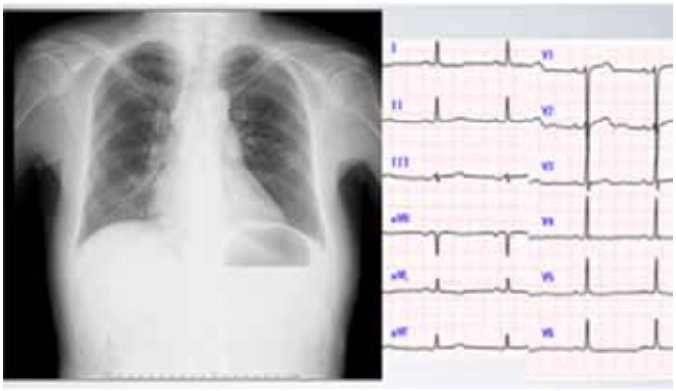

Figure 8: At discharge, heart failure and her condition recovered.

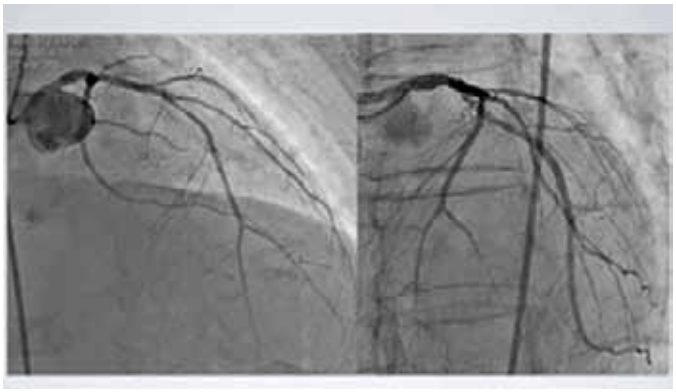

Figure 9: The coronary angiography (CAG) of 2 months after PCI showing no significant stenosis at left main trunk (LMT).

after insertion of an IABP because of limited vascular access. We attempted to insert a diagnostic catheter with a 0.014-inch guidewire before replacing this with a GC using an extension wire because insertion of the GC was initially difficult and insertion of a diagnostic catheter was possible. However, because of the severe tortuosity of the left subclavian artery, the guidewire prolapsed during the procedure, making insertion impossible. A balloon was inserted without the use of a GC, and the anchor balloon technique was implemented. Yoshimachi et al. [1] reported performing IVUS and inserting a balloon when performing PCI using the King's cloth technique without the use of a GC, although a 0.035-inch guidewire or microcatheter might have been useful in this patient. We carefully inserted a semi-compliant balloon up to the LMT, even though it was fixed to a prolapsed wire. A risk of negative effects on vital signs was present; however, inflation time was maintained within approximately 5 s. The anchor balloon technique was also simultaneously used to engage the GC. The balloon was dilated for a

short time, and the GC was inserted without any negative effects on the hemodynamic state of the patient because passage of the balloon was difficult due to severe stenosis of the LMT.

Prognoses of TRI and transfemoral coronary intervention (TFI) are not very different [2, 4]. With regard to complications in ACS [5], ST segment elevation myocardial infarction [6–10] or IABP support is associated with a favorable prognosis for TRI [11]. Transradial PCI may be considered for severely obese patients [12] and women [13] who are at a higher risk of bleeding complications.

The limitations for transradial PCI were few, and this made it particularly suitable for our patient in whom the approach site was limited in size. Maneuvering of the GC might have been easier by downsizing to a 5 Fr GC [14] or inserting a 0.035-inch guidewire during the procedure.

TRI or a downsizing stenting system is essential for patients in whom the approach site is limited. The use of an extension wire after the insertion of the diagnostic catheter and the anchor balloon technique has been successful in limited cases when insertion of a GC is difficult.

CONCLUSION

The guiding catheter (GC) insertion is difficult during transradial intervention for patients with acute coronary syndrome (ACS) and stenosis of the left main trunk and in whom the approach site is limited in size. The GC insertion and percutaneous coronary intervention can be achieved when the diagnostic catheter is exchanged with a GC using a 0.014-inch wire as well as an extension wire, after which the balloon anchor technique should be used.

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Author Contributions

Daizaburo Yanagi – 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
Takeshi Serikawa – 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
Masanori Okabe – 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
Yusuke Yamamoto – 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.

Copyright

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CLINICAL IMAGES PEER REVIEWED | OPEN ACCESS

^{18}Fluorine fluorodeoxyglucose positron emission tomography diagnosis of an aortic thoracic prosthesis infection by slow-growing bacteria

Geraldine Celine Bera, Patrick Farahmand, Françoise Cavailloles, Charlotte Lepoutre-lussey

CASE REPORT

A 66-years-old male with a history of aortic root aneurysm and bicuspid aortic valve underwent aortic root replacement with a Bentall procedure in 2009. Four years later, he presented to the emergency department with a fever and general malaise. Serial blood cultures were negative and a transthoracic echocardiogram showed no evidence of vegetations or an aortic root abscess. Although there was no evidence to support a diagnosis of infective endocarditis (IE) empirical antibiotic therapy, comprising vancomycin, gentamicin and rifampicin, was commenced. Two weeks later, a^{18}fluorine-fluorodeoxyglucose positron emission tomography (^{18}[F]-FDG PET) was performed under a diet (low-carbohydrate and high-fat during 24-hours) designed to suppress myocardial activity. One hour after the injection of 295MBq of ^{18}F-FDG radioisotope, 3 foci were highlighted around the aortic prosthesis (SUV_{max} = 6, graft-to-mediastinum ratio SUV_{max} = 3.2) (Figures 1–A, D, E) with additional uptake within mediastinal lymph adenopathies (SUV_{max} = 2.6) (Figure 1B). There was no pathological uptake noticed around the adjacent aortic valve prosthesis (Figure 1C). This result was highly suggestive of an infection of the aortic tubular prosthesis and so surgery was undertaken to replace the aortic tubular prosthesis with a mechanical valve. During surgery, macroscopic examination confirmed the presence of pus inside the envelope surrounding the prosthesis. Cultures were taken during the surgery and analysis via polymerase chain reaction revealed the growth of *Kingella kingae*, the fifth member of the HACEK (*Haemophilus*, *Aggregation bacter*, *Cardiobacterium hominis*, *Eikenella corrodens* and *Kingella species*) bacteria group.

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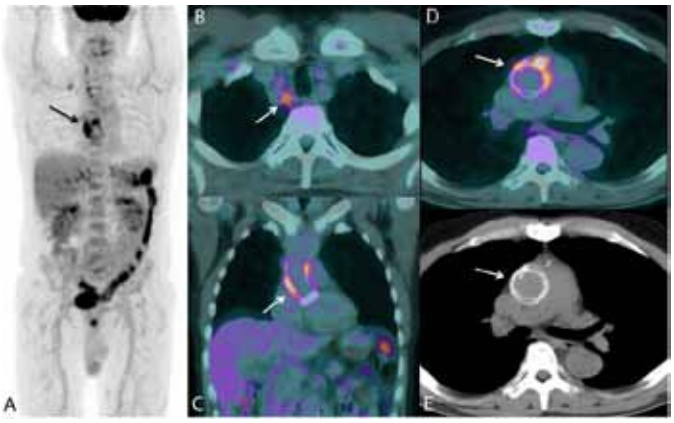

Figure 1: (A) Maximum intensity projection showing pathological mediastinal uptake and no uptake suggestive of septic emboli throughout the rest of the body, (B) Regional analysis of 18F-FDG uptakes point out on mediastinal lymphadenopathies, one on left latero-tracheal posterior area of upper mediastinum, (C, D) Typical uptake pattern around aortic thoracic prosthesis, and (E) Simultaneous computed tomography allow accurate localization.

DISCUSSION

The HACEK microorganisms, commensal residents of the oropharynx, are responsible for ~3% of cases of IE. They have an incidence of ~2–3 cases per 100,000 of population per annum and their mortality can reach up to 14%. These gram-negative bacilli are slow-growing, likely explaining the frequently negative blood cultures [1]. Diagnosis of cardiovascular prosthesis infections can be challenging with 30% having normal or inconclusive echocardiographic findings [2]. This often leads to a serious delay in instigating medical and, more importantly, surgical treatment. Saby et al. [2] have demonstrated that if used as a major criterion ^{18}[F]-FDG PET increased the sensitivity of the modified Duke classification from 80–97%, without compromising its specificity. Thoracic aortic prosthetic graft infection is particularly rare and associated with very high morbidity and mortality [3]. Its diagnosis by conventional imaging is difficult due to non-specific nature of the findings and the sensitivity of magnetic resonance imaging remains unclear [2–4]. This report shows a high added value of ^{18}F-FDG PET in the diagnosis of a sub-acute endocarditis even after fifteen days of a broad-spectrum antibiotic therapy. Focal ^{18}F-FDG uptake around the cardiovascular prosthesis has a sensitivity of 93%, specificity of 91%, positive predictive value of 88% and negative predictive value of 96% for the diagnosis of prosthetic vascular graft infection [4]. A non-homogeneous uptake pattern around the cardiovascular prosthesis is described as a poor diagnostic marker [4]. While SUV_{mean} and graft-to-mediastinum ratio represent the overall metabolic activity in the whole graft more accurately than SUV_{max}, [4] a $SUV_{max} > 8$ in the surrounding graft area has been described as a potential cut-off value for distinguishing infected from non-infected grafts with sensitivity and specificity of 100% and 80%, respectively [3].

CONCLUSION

This report illustrates that ^{18}F-FDG PET with a characteristic uptake pattern (focal uptake), a $SUV_{max} \geq 6$ and a graft-to-mediastinum ratio $SUV_{max} \geq 3.2$, allowed the detection of a cardiovascular prosthesis infection due to a slow-growing bacteria even following 2 weeks of antibiotic treatment.

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Professor Aurelie KAS MD.PhD., Department of Biophysic UPMC Paris VI, LIB, INSERM UMR 678, 91 Bd de l'hôpital 75651 Paris Cedex 13 France and Department of Nuclear Medicine, AP-HP Hospital Pitié-Salpêtrière, 47-83 Bd de l'hôpital 75651 Paris Cedex 13 France.

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Author Contributions

Geraldine Celine Bera – Conception and design, Analysis and interpretation of data, Drafting the article, Final approval of the version to be published
Patrick Farahmand – Acquisition of data, Analysis and interpretation of data, Critical revision of the article, Final approval of the version to be published
Françoise Cavailloles – Acquisition of data, Analysis and interpretation of data, Critical revision of the article, Final approval of the version to be published
Charlotte Lepoutre-lussey – Conception and design, Critical revision of 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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CLINICAL IMAGES PEER REVIEWED | OPEN ACCESS

A rare case of finger metastasis showing as the first sign of lung cancer

Yasuyuki Taooka, Gen Takezawa

CASE REPORT

An 82-year-old male was admitted to our hospital complained of dyspnea on exertion and left chest pain. three months prior to admission, he noticed swelling and pain of his left forefinger and middle finger. Before visiting to our hospital, patient consulted his primary care physician and was treated with antibiotics as trauma and infectious disease. But his finger pain continued, and then he gradually noticed dyspnea and left chest pain. When admitting to our hospital, the tip of his fingers showed erosive redness and bled easily by contact (Figure 1). X-ray showed irregular osteolytic change of distal phalanx of forefinger and middle finger (Figure 2). His serum CEA level was elevated, which was 7.6 ng/mL (normal rage was less than 5.0 ng/mL), and thoracic CT scan showed left pleural effusion and pulmonary nodules (diameter was 2.1 cm, and military nodules was also recognized in the same lobe) of left lower lobe. Since oozing from his index finger did not stop and severe pain continued, his left index finger was finally amputated. The histopathological examination of his left forefinger revealed poorly differentiated adenocarcinoma, and the cytology of pleural fluid was also showed non-small cell carcinoma. CEA level of left exudative pleural fluid was 87.3 ng/mL. We diagnosed as his having primary lung cancer (T3, N2, M1b, stage IV), and finger metastasis. Left pleural effusion was controlled by therapeutic thoracentesis. But since his performance status was poor, there was no indication for systemic chemotherapy. To relive his dyspnea and pain, oxygen therapy and administration of opioid was continued. After four months, the patient died of disseminated disease and respiratory failure after the palliative therapy.

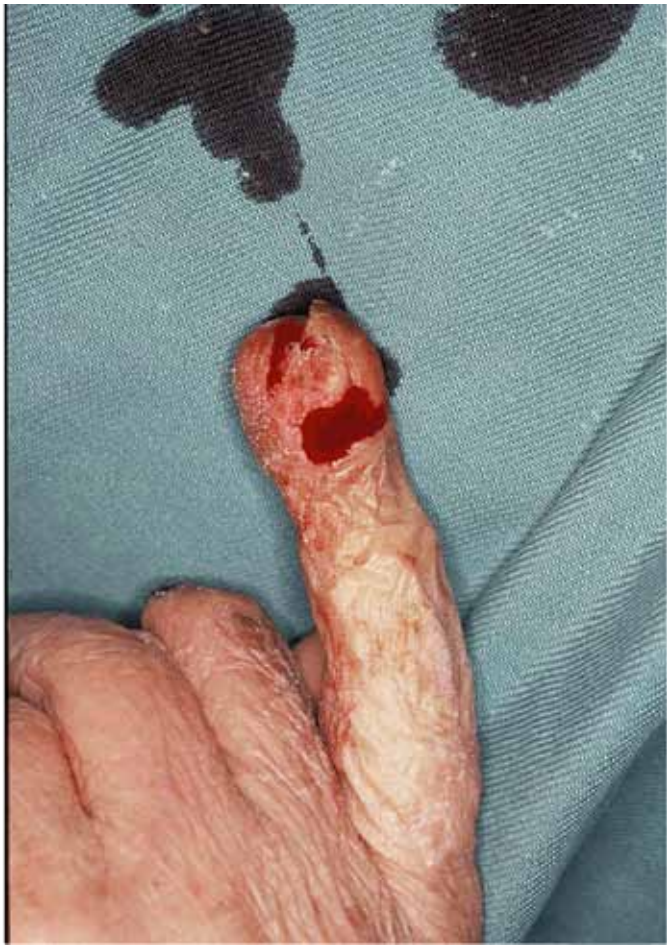

Figure 1: Left forefinger, which removed lateral-side of nail, showed erythematous swelling, and bled easily by contact.

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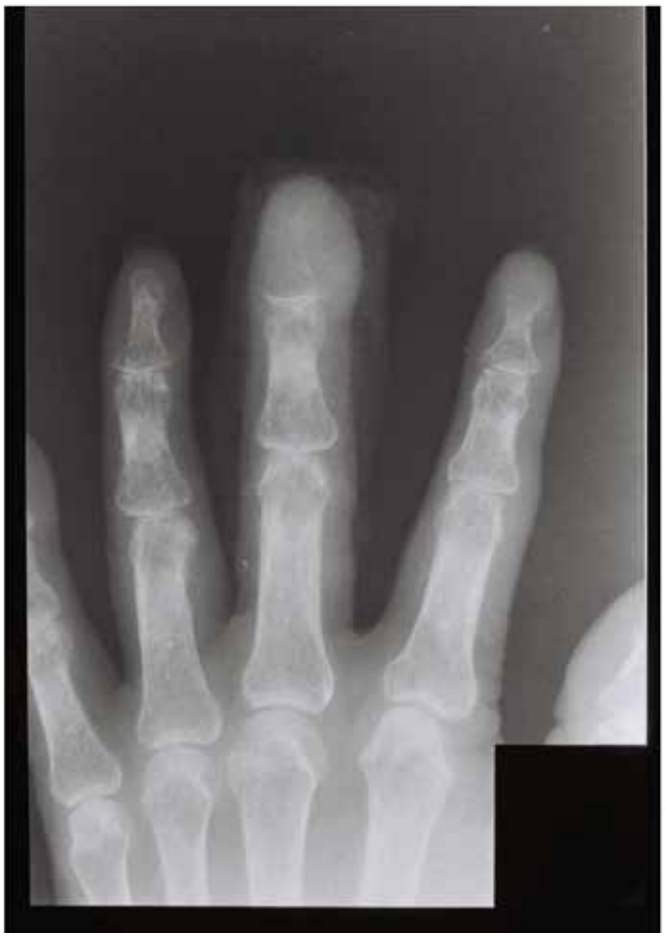

Figure 2: X-ray of left forefinger and middle finger: Destruction of cortex and irregular osteolytic lesions of distal phalanx with left forefinger and middle finger were shown.

DISCUSSION

Bone metastasis, especially axial bone metastasis is common in advanced lung cancer patients [1, 2]. But finger metastasis, as the first sign of lung cancer is extremely rare, and the incidence of finger metastasis from primary tumors is approximately 0.1% [3–5]. According to the previous reports [1–5], the most cases are mortal within six months and are correlated with poor prognosis because of advanced staging. According to the previous report [5, 6], lung, kidney, breast, and gastrointestinal cancers are known as the primary lesions of acrometastasis to the hands, and 44% of them is lung cancer. It is difficult for primary care physician to suspect the possibility of finger metastasis rather than trauma and infectious diseases because of uncommon presentation of metastasis with redness, swelling, and pain of the finger tip [4, 5]. In many cases, performance status of the patients showed already got worse, when having the diagnosis of cancer. Therefore, the treatment is usually palliative, and radiotherapy, chemotherapy, and amputation are performed. In this case, severe pain and bleeding did not discontinued and his index finger was finally amputated.

CONCLUSION

A rare case of lung cancer that metastasized to fingers is presented. Finger metastasis is generally sign for poor prognosis, and palliative treatment is important to relive the symptom.

How to cite this article

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Conflict of Interest

Authors declare no conflict of interest.

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CLINICAL IMAGES PEER REVIEWED | OPEN ACCESS

Superior semicircular canal dehiscence syndrome: A rare cause for dizziness

Han-Kuang Chen

CASE REPORT

A 35-year-old female presented with a 18-month history of intermittent dizziness triggered by hearing loud sound and symptoms resolve as soon as sound stops. She saw her general practitioner 12 months ago and was diagnosed with benign paroxysmal vertigo. However, patient's symptom gradually worsened. She described that, in recent months, her dizziness symptom could be triggered just by putting a mug on a table or talking too loudly. She also described hearing her eyeball movements at night when the surroundings were quiet. Her worsening symptoms severely affected her quality of life. She was otherwise healthy and had no medical or surgical history.

The patient was referred to an ENT specialist for further investigations. A high resolution CT scan of the left temporal bone was performed and it showed 1 mm dehiscence of the superior semicircular canal antero-superiorly; consistent with superior semicircular canal dehiscence (Figure 1). Patient subsequently underwent an elective surgery to repair the superior semicircular canal dehiscence. The surgery was done by middle cranial fossa approach and the area of dehiscence was occluded with bone pâté and covered with fascia. Postoperatively, patient was admitted to intensive care unit for overnight observation and was transferred to ENT ward on day-1. Initially, her cranial examination revealed nystagmus on lateral gazes but it resolved on day-3. Patient continued to make a good recovery and was discharged on day-4 postoperatively. At fourth week follow-up, she had complete resolution of the symptoms and she was able to perform any activities of daily living without problems.

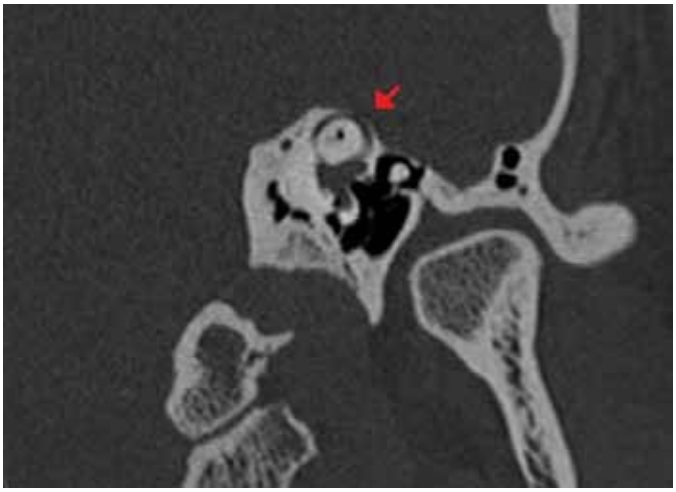

Figure 1: 1 mm dehiscence of the superior semicircular canal antero-superiorly.

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DISCUSSION

Superior semicircular canal dehiscence syndrome (SSCDS) is a rare condition which is characterized by sound or pressure induced vestibular symptoms. It is caused by dehiscence of the bone overlying the superior canal and the prevalence is estimated to be 0.5–0.6% [1]. It is postulated that the dehiscence creates a third mobile window into the inner ear. As a result, external sound and pressure can cause changes in the middle ear pressure, resulting in vertigo and oscillopsia [2]. The evoked eye movements in this syndrome align with the plane of the dehiscent superior canal. Patients may also experience hyperacusis to bone-conducted sounds.

The diagnosis is best established upon clinical findings and imaging, include the following [2]:

(i) Vertical-torsional eye movements evoked by sound or pressure stimuli noted on examination performed with Frenzel goggles.
(ii) Lowered thresholds for responses to vestibular-evoked myogenic potentials
(iii) CT scan of the temporal bones.

In terms of management, SSCDS can be treated by surgically occluding the dehiscence and it can effectively alleviate symptoms [3]. In our case, the patient had full resolution of her symptoms after receiving the surgery. On review of literature, only a small number of SSCDS cases have been reported. The majority of the patients experienced similar sound or pressure induced vestibular symptoms as observed in this case. Fortunately, all of the patients had good symptom control if not full resolution after surgical treatment.

CONCLUSION

Superior semicircular canal dehiscence syndrome (SSCDS) is characterised by vestibular symptoms induced by sound or pressure. Although uncommon, this condition can significantly affect quality of life. Awareness of this rare cause of dizziness is essential for timely diagnosis and intervention.

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EDITORIAL PEER REVIEWED | OPEN ACCESS

The value of case reports to medical science and clinical practice

Altacílio A. Nunes

We currently live in a scientific context where great discoveries, especially in medicine, are not as eloquent as for example, those of the nineteenth and early twentieth century, when almost everything we know today about knowledge in the etiology of diseases, was unveiled, leaving us in the second half of the twentieth century to the present, to clarify the molecular mechanisms and possible new associations (more subtle) between exposure and disease. Thus, the case reports in great detail, mainly in the medical field and focusing on rare, unusual, new and/or unknown diseases, as well as change the natural history of a particular disease, in addition to reports on new treatments, both medical or surgical and complications associated with them, are considered useful tools in generating hypotheses that certainly induce the scientific community in the development of clinical or basic research, seeking clarification of possible cause-effect associations, as well as other relevant aspects diagnosis treatment and prognosis.

The case reports fall within the field of descriptive epidemiology or not non-analytical epidemiology [1], not focus precisely to adhere comparisons, and therefore do not enable causal relationships and, thus has mainly focused on the description of components constituting the characterization of reported case, i.e., who got sick (personal aspects - biological, social, etc.); where the disease has occurred (spatial aspect or "geographical"), when the disease was diagnosed or reported (seasonal or temporal aspects). So all case report with good quality, the following three questions must be answered clearly: *Who? When? Where?* Obviously, the answers to such questions should be easily identified, allowing the reader to understand the report from a biological, epidemiological and socio-demographic point of view, facilitating and inciting the hypothesis formulation, triggering conducting observational studies such as cohort, case-control and cross as well as experimental studies and clinical trials, according to the nature of the case presented in the report. Clearly, for physicians who do not belong and do not work in academic institutions, but who perform activities in clinical practice, case reports represent a very useful tool of information for your profession, providing them elements that undoubtedly assist in the care their patients because by reading the details of the reports certainly update their knowledge as well as, identify the clinical details described much in common to that observed in patients [2].

Another important application area and use of case reports is that of medical education. This mode of study has important role in the training of medical students and residents of all clinical areas of medicine, and often conducting clinical sessions where intriguing case reports or even common cases are selected to be discussed comprehensively between staffs and those in training with unquestionable gains in theoretical and practical learning with more concrete and efficient results compared to only lectures [3].

Before the era of clinical trials and large observational studies, particularly cohort, case reports were the only advancement sources in all clinical areas of medicine, however, after the advent of so-called evidence-based medicine [4], in the final 1980s and early 1990s, the descriptive studies, including the case reports, have been considered by the academic community, a kind of evidence of lower hierarchy in relation to analytics studies [5]. However, even within the evidence-based medicine, currently the clinical reports again play an important role when considering the professional experience as a featured element in medical decision making, especially when considering the safety [6] of the patient and the proposed treatment, as well as the natural

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history of the disease [7]. After all, it is important for the scientific community to emphasize that considering the citation metric, the case reports are among the most cited publication types [8], so, must also for such reason, be regarded with due attention.

For all these reasons and it is no coincidence that all the major and important medical journals from all over the world, keeps in its scope the publication of case reports and case series [9]. On the other hand, has increasingly emerged specialized scientific journals in publication of cases and images related reports, which undoubtedly is a gain for all, including medical students, medical teachers, the scientific community, medical professionals, healthcare managers and patients [10].

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