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Contents

Cover Image

Figure 4: Four sections of the specimen using a band saw showing the tumor.

Review Article

323 Aversion for case reporting by major medical journals: A trend towards perfect dichotomy
Segun Adeoye

Case Series

329 Proliferative vitreoretinopathy secondary to degenerative retinoschisis: A case series
Mansukhli Amer, Safia Ahmed, Carpi Akikio Olali, Sohail Ahmed, Gupta M

Case Report

334 Bilateral traumatic distal femoral physeal slips: A very rare case presentation
Anil Mehtani, Jatin Prakash, Suresh Chand, Abhinav Sinha, Ajeet Singh, Harvinder Dev

339 Syphilitic paroxysmal cold hemoglobinuria associated with peripheral gangrene: A diagnostic dilemma
Segun P Adeoye, Seema Tayal, Apar Bains, Prabhjot Manes

345 A rare case of myositis ossificans progressiva presenting as multiple progressive contracture
Anil Mehtani, Jatin Prakash, Suresh Chand, Abhinav Sinha, Ajeet Singh, Harvinder Dev

351 Imaging findings in a case of cystic neoplasm of pancreas: A case report
Pooja Gupta, Devika Gupta, Kamlesh Kumar Singh, Satish Mendonca

355 Urinary bladder perforation due to encrusted cystitis: A rare entity
Faouzi Mallat, Wissem Hmida, Khaled Ben Ahmed, Sarra Mestiri, Faouzi Mosbah

360 Retroperitoneal desmoid tumor with unusual appearance as a spontaneous ureteral rupture and rare association with desmoid tumor of the breast
Faouzi Mallat, Wissem Hmida, Sarra Mestiri, Badreddine Srha, Moncef Mokni, Faouzi Mosbah

Vol. 5, No. 5 (May 2014)

365 Self resolving non-parasitic splenic cyst: A case report
Meetal Shah, Abdul Quyyum Khan, Maria Vittoria Cavalletti, Luciano Perrone, Chenji Ratnavel, Giovanni Domenico Tebala

370 Familial myelinated nerve fibres with proliferative vitreoretinopathy
Mehra A, Safia H Ahmed, Carpi Akikio Olali, Ahmed S, Gupta M

373 Longest and left-sided gallbladder
Atul Kumar Mittal, Sourabhi Sharma, Selva Kumar Balakrishnan, Jeevan Kankaria, Rajkamal Jenaw

377 Primary aggressive non-Hodgkin lymphoma of the parotid gland in a young individual: A case report
Devika Gupta, GPS Gahlot, Vandana Rana, Rajat Jagani, Davendra Swarup

382 Gastric outlet obstruction due to a giant antral polyp which have malignant transformation
Erkan Oymaci, Ali Coskun, Deniz Ucar, Erdem Sari, Nazif Erkan, Mehmet Yildirim, Hale Kizanoğlu

387 A rare cause of upper gastrointestinal bleed: Posttraumatic pseudoaneurysm
Negi RC, Brij Sharma, Bhupender, Gaurav Kapoor, Bal Beer Verma, Ashok Sharma

Case in Images

391 Neglected telangiectatic osteosarcoma of the femur presenting as surgical emergency
Youssef Mahdi, Lamiaa Rouas, Abdellouahed Amrani, Abderrahmane Malihy, Najat Lamalmi, Zaitouna Alhamany

Clinical Images

398 Hemodialysis catheter malposition: How to prevent this fault?
Fateme Shamekhi Amiri
Aversion for case reporting by major medical journals: A trend towards perfect dichotomy

Segun Adeoye

ABSTRACT

Introduction: This case report introduces the traditional form of medical literature. It chronicles the contributions made by landmark case reports in the times of old as well as in the present. Review: The author details the current case report publishing-aversion by major medical journals, and attempts to explain the rationale. The article describes the impact of evidence-based medicine and journal impact factor protection as facilitators of the “discrimination” against case reporting. The paper analyzes the case report acceptance policy of 20 medical journals across the whole spectrum of impact factors and identifies interesting patterns. The author chronicles the proliferation of case reporting journals and identifies the trend towards a perfect dichotomy in medical publishing. Conclusion: In concluding, the author gives his take on the many issues raised while proposing potential solutions.

Keywords: Case report aversion, Case studies, Impact factor, Medical journal publishing

INTRODUCTION

The cornerstone of the incremental medical knowledge in the times of old was the fervor of early physicians to publish case reports. Case reporting is the traditional way of publishing novel diagnosis and treatments, report adverse effects or unique clinical successes or medical breakthrough. They also serve the backbone of medical education [1]. Many landmark medical advances were documented as case reports: Broca described aphasia complicating left hemispherical lesion, Tillet and Sherry described the use of fibrinolytic agents to complement antibiotic therapy treatment of empyema, Thomas Addison, and Harvey Cushing’s reported Addison’s disease and Cushing syndrome, respectively. Marcintyre authored the first description of multiple myeloma and Barnard; the world’s first heart transplant. All these and many more case reports revolutionized medicine in one way or another.

The testament to the fact that the goodness of old time case reporting remains undiluted in the present time is evident in some relatively recent landmark case reports and series: identification and characterization of the West Nile virus (1999) and SARS-CoV virus (severe acute respiratory syndrome-Corona virus) (2002) in New York and Guandong Province (Southern China), respectively after case series reported epidemic pattern of unknown etiology, as well as steroidogenic acute regulatory (STAR) protein mutations in adrenal steroid hormone production from a case series on six patients [2]. These landmark case report opened new scientific avenues and pushed the
frontiers of medicine to levels unimaginable.

The times have changed however, with developments in research methodology and biostatistics ushering in an era where there appears to be contempt for this traditional way of medical reporting. The major medical journals accept <5% and 15–25% [3], respectively of case report and original research article submissions for publication (even higher for review articles). One motivation for publishing preference for original research and review article is the need to maintain or improve journal impact factor. It is also fair to infer that perhaps a much larger number of case reports relative to original research and review article submissions may partly contribute to the publication rate disparity. While case reporting provides a complete descriptive of a case-specific encounter, emphasizing uniqueness and interesting clinical caveats, research articles provide answers to well-focused diagnostics or therapeutic questions, by exploiting large samples to reduce bias and robust statistical tests to confirm or refute significance of findings. The two reporting forms are at opposite ends of the spectrum of medical literature, each with unique applicability and appealing to different audience. The intent of this article is not to victimize research or review articles, rather to vindicate case reports.

Advent of evidence-based medicine (EBM) and evidence ranking scales

Most appear to have joined the bandwagon of EBM faithfults. There has been for some time now, a call to adopt only diagnostics and interventions with proven effectiveness. Widely accepted scales for ranking evidence of levels for diagnostics and interventions includes: the United States Preventive Task Force (USPTF), the United Kingdom National Health Service (UK-NHS) and GRADE Working Group. Physicians preferential choose diagnostics and therapies with superior evidence ranking unless there exist preclusive practice-, patient- or cost-related factors.

Medicine, as it is today is increasingly protocol-driven, guidelines are reflective of evidence rating of competing diagnostics or interventions. The rationalization is that more effective and safer interventions receive higher evidence ranking. It is the expectation that diagnostic considerations and therapeutic options are vetted, and assigned rankings based on accrued objective evidence obtained from research and clinical experience. The type and source of evidence also impacts ranking, for example evidence from randomized clinical trials (RCTs) and systematic reviews receive higher evidence rankings than that from a consensus of expert panels or from case reports: with case report accorded anecdotal evidence level status (a fore-runner evidence needful of confirmatory testing), well favored for documenting idiosyncrasies, ambiguities, novelties, and provocateurs.

The reliability on the superior ranking of evidence obtained from RCTs has been called to question too many a time in the recent past. Notably, the rather shocking finding that medical management of coronary artery disease is not inferior to percutaneous coronary interventions, except in cases of myocardial infarction. That there are many more reversals of evidence obtained from RCTs is evident in the writings of Prasad et al. in three landmark publications: “A Decade of Reversals: An Analysis of 146 Contracted Medical Practices” [4], “The Frequency of Medical Reversals” [5] and “Reversals of Established Medical Practices: Evidence to Abandon Ship” [6]. Many such reversals and retractions have origins in case reports and series. This is not to say that case report is infallible, for it is, especially as it is reliant solely on accurate description, impression and reasoning provided by the author, absent the check and balance inherent in large study sample sizes, significance levels and p-values.

Discrimination Against case reporting by Major Medical Journals

The publication of case reports by major medical journals is dwindling by the day. There appears a concerted effort by these journals to stifle the already faltering case reports market. Many of these journals report 0–5% [3] publication rates for case reports, the Annals of Family Medicine clearly state on their website that they do not usually publish case reports [7]. If the current trend continues, we run the risk of attaining perfect dichotomy in medical publishing. The main reason offered by major medical journals for not publishing more case reports is the fact that case reports have a lower evidence ranking and are less impactful when compared to original research and review articles. The impetus for this is the euphoria for EBM as well as the wielded power of statistical analysis (our addiction to p-values).

In rejection correspondence to authors of case reports, editors commonly cite space limitations in print journals relative to volume of submissions as a reason for manuscript rejection. Others yet state that the manuscript is not novel enough or does not expand medical knowledge well beyond what is already known. An interesting comment in one rejection letter read—“a manuscript’s intent to raise the level of suspicion of clinicians for certain rare or re-emerging conditions does unfortunately not provide sufficient priority to consider publication” [3]. Many such letters conclude advising the author to consider publishing elsewhere; some even go further by suggesting submission to clinical journals dedicated to the publication of case reports [3].

In an analysis of 20 medical journals of the whole spectrum of impact factors, three case report acceptance policy patterns were identified (Table 1): an open policy to consider case reports and series (with one—Journal of American Board of Family Medicine, declaring preference for case series over case reports); a declared policy not to consider case reports or series; and an unclear policy of not listing case reports and series in the list of acceptable article types. The top-tier journals
Table 1: Summary of declared and demonstrated case report acceptance policy of 20 medical journals.

<table>
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<tr>
<th>Journal Name</th>
<th>Location</th>
<th>Impact Factor</th>
<th>Open policy “Will consider statement”</th>
<th>Closed policy “Will not consider statement”</th>
<th>Case reports not listed as acceptable literature forms</th>
<th>Research article(s) per issue</th>
<th>Review article(s) per issue</th>
<th>Case report(s) article(s) per issue</th>
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<td>NEJM</td>
<td>America/Europe (1) vs. Other (2)</td>
<td>51.67</td>
<td>x</td>
<td></td>
<td>4</td>
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<td>1</td>
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<td>Lancet</td>
<td>1</td>
<td>39.06</td>
<td>x</td>
<td></td>
<td>3</td>
<td>1</td>
<td>1</td>
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<tr>
<td>JAMA</td>
<td>1</td>
<td>30.03</td>
<td>x</td>
<td></td>
<td>5</td>
<td>1</td>
<td>0/1*</td>
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<tr>
<td>British Med. Journal (BMJ)</td>
<td>1</td>
<td>17.22</td>
<td>x</td>
<td></td>
<td>4</td>
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<td>13.98</td>
<td>x</td>
<td></td>
<td>4</td>
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<td>BMC Medicine</td>
<td>1</td>
<td>6.68</td>
<td>x</td>
<td></td>
<td>10</td>
<td>2</td>
<td>0</td>
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<td>6.45</td>
<td>x</td>
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<td>Mayo Clin. Proceed</td>
<td>1</td>
<td>5.70</td>
<td>x</td>
<td></td>
<td>9</td>
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<td>Annal of Family Medicine</td>
<td>1</td>
<td>5.36</td>
<td>x</td>
<td></td>
<td>4</td>
<td>3</td>
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<tr>
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<td>4.25</td>
<td>x</td>
<td>x</td>
<td>4</td>
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<td>6</td>
<td>3</td>
<td>4</td>
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<tr>
<td>Mount Sinai Journ. Of Medicine</td>
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<td>2.00</td>
<td>x</td>
<td></td>
<td>8</td>
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<td>J. Am. Board of Family Medicine</td>
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<td>1.76</td>
<td>x ***</td>
<td></td>
<td>14</td>
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<td>1.72</td>
<td>x</td>
<td></td>
<td>1</td>
<td>1</td>
<td>0**</td>
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<tr>
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<td>2</td>
<td>1.59</td>
<td>x</td>
<td></td>
<td>6</td>
<td>2</td>
<td>6</td>
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<tr>
<td>Israel Medical Association Journal</td>
<td>2</td>
<td>1.02</td>
<td>x</td>
<td></td>
<td>7</td>
<td>2</td>
<td>4</td>
<td></td>
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<tr>
<td>The National Med. Journal of India</td>
<td>2</td>
<td>0.59</td>
<td>x</td>
<td></td>
<td>2</td>
<td>1</td>
<td>1</td>
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<tr>
<td>Journal of Pakistan Med. Ass.</td>
<td>2</td>
<td>0.49</td>
<td>x</td>
<td></td>
<td>15</td>
<td>1</td>
<td>5</td>
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<tr>
<td>Annals of African Medicine</td>
<td>2</td>
<td>NA</td>
<td>x</td>
<td></td>
<td>7</td>
<td>1</td>
<td>3</td>
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<tr>
<td>Annals of Nigerian Medicine</td>
<td>2</td>
<td>NA</td>
<td>x</td>
<td></td>
<td>4</td>
<td>1</td>
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* only one case report/series in 2013
** few issues with 1 case reports.
*** indicate preference for case series over case reports
NA: not available
(with the highest impact factor) have an open declared case report acceptance policy but appear to have less open demonstrated policy. This is evident in a 1:3 published case report: research article ratio. Surprisingly, the next-tier of journals have an even much less balanced case report: research article ratio: BMJ, Annals of Internal Medicine, BMC Medicine, Journal of Internal Medicine, and the Annals of Family Medicine and Medicine being absolutely case report publishing averse, with most of these having declared an open policy to case reporting on their websites. The BMC Medicine and Mount Sinai Journal of Medicine are research and translational research-specific journals, by virtue of their declared niches, are absolutely case report averse. It is not surprising the Journal of Postgraduate Medicine has the most demonstrated open policy to case reporting with case report: research article ratio of 1:1. This is reflective of the popularity of case reporting amongst physicians in residency training. Medical journals located outside the Americas and Europe appear to be more receptive to case reporting.

I do not intend to play jury or advocate for these editorial decisions, but will state unequivocally that case reporting does not necessarily have to report new disease entities, rather it may present interesting caveats on already existing entities, especially atypical presentations, unique pathogenesis, unusual associations and unconventional interventions.

The Advent and Proliferation of Dedicated Medical Case Report Journals

The past two decades has witnessed the proliferation of medical case report journals, notable mentions are the Journal of Medical Case Reports (JMCR), Journal of Medical Cases (JMC), International Medical Case Reports Journal (IMCRJ), and the International Journal of Case Reports and Images (IJCRI). The impetus for this unprecedented proliferation was the need to cater for “disenfranchised” case reporters who felt shut out by the major medical journals. That these case report journals saw the need and cashed in is evident in the mission statements of some of these journals detailed on their websites. No better description is provided than the acknowledgements on IJCRI website.

“The IJCRI was born of necessity to provide authors with a forum where they are welcome to submit reports of unusual cases and atypical presentation of common cases. In recent years, many international journals with a print version have stopped accepting case reports for publication. There is always a tough competition between review articles and case reports for very limited page-space in the print version of the journals. In this competition “case report” invariably lose. As a consequence, it has become very difficult to publish interesting and unusual cases in international, peer-reviewed journals. This lack of publishing opportunities for authors was the motivation for us to start IJCRI.”

The journal caters for the needs of residents and fellows as they take baby steps in scholarly publishing (often case reporting) [8]. Absent PubMed indexing, many case report journals use open access publishing to ensure the journal reach a wider audience, they increase reader access through email notifications, maintain an active presence on social networking sites, and use press releases to general and scientific press [9]. Furthermore, they achieve high volume publishing rates through frequent and high speed publishing. Some even permit “early view articles” and offer availability for free full text articles immediately upon publication [7]. That these strategies are yielding dividends is evident in the greater reader access and improved author-reported satisfaction.

MY TAKE

I appreciate all forms of medical reporting and believe in the true spirit of inclusiveness in medical literature. I am encouraged by recent proliferation of case reporting journals. These journals continually strive for PubMed indexing, receive fewer journal citing and by extrapolation, lower impact factors (IF). Defenders of this index purport that the impact factor is a reflection of journal significance, affected solely by journal article citing, but I see a more multifactorial influence: the confluence of journal citing as well as editorial policies and strategies to increase its impact factor like skimming for review articles (with higher potential for citation) and aversion for case reports.

It is not news that physicians and researchers in the third world, as well as residents and fellows in all-worlds, are lacking in sponsorship for original research. Little wonder they have to rely on case reporting as an avenue to share experiences through authorship. It is my opinion that shutting out case reports from the major medical journals not only denies subscribers to these journals the benefits of appreciating the case uniqueness, individuality and the completeness available in case reports, but is in itself, true discrimination against third-world physicians, as well as up-coming residents and fellows in all-worlds, who now have limited options to showcase their scholarly work. These physicians have to resort to collaborations with more privileged institutions and colleagues abroad to access research funding and a path to publishing with the “big-boys” medical journals.

I make the following propositions. Firstly, the major medical journals may consider increasing its allocation for and publication of case reports to meet the needs of readers and as well as to be more inclusive in its authorship base. They can achieve this by increasing journal issue size or maintain current issue size while increasing relative space allocation for case reports. Another approach currently being used by some journals is to run a case report-sympathetic version in parallel with the main journal. For example, the Journal of the Royal Society of Medicine (JRSM) uses the JRSM-Short Reports [10] version to increase case reporting, well amongst other intentions. Perhaps a perfect dichotomy (absolute case
report-averse versus absolute case reporting journals) is the Utopian state of medical publishing, however, this is acceptable only when there exist enough case report journals to accommodate the volume of case reports seeking publication.

Secondly, PubMed should increase its indexing of case report journals to accord them equal access to readership and potential for citing as it does for research and review article-leaning journals. Thirdly, present the controversy surrounding the validity of impact factors [11], a revised computation method that attempts to correct the inequality in the grading of the impact of research/review article- learning journals and case report journals should be implemented. Let us not compare apples with oranges. Perhaps the way to go may be the creation of a modified impact factor scale for case report journals. I see no reason why an IF of 51.7 is reported for the NJEM [12] while the reputable, BioMedCentral published, JMCR reports a mere 0.36 [9]. Authors and readers should understand the caveats in the computation of IF and appreciate why case report journals fair badly. Fourthly, governments and institutions of third world countries should invest in medical journals of their own, one that meet the needs of its physicians, researchers and scientists. Graduate medical programs should collectively consider managing medical journal like the McGill Journal of Medicine, a medical student-managed journal. Oversight organizations like the specialty boards and the Committee of Interns and Residents (CIR) may help in this regard. Fifthly, beyond scapegoating major medical journals for their aversion for case reports, the general medical academic community and accreditation agencies should right the decade-long injustice in medical academia, of denying academic credits to authors for case reporting. The academic rigors involved in putting together a case report (many include rigorous literature review sections) is worth some scholarly recognition even if such award is less than is accorded research papers and review articles. Perhaps, referencing case reports as case studies may be the first step in the redemption process. I suggest 0.5 credit equivalence recognition for case reporting.

In concluding, I call on the spirit of legendary Sir Richard Hutchinson to reaffirm his petition of 1958: “From too much zeal for the new (randomized clinical trials) and contempt for what is old (case reports), from putting knowledge (statistical analysis and significance) before wisdom (clinical experience and significance), science (statistical significance) before art (clinical uniqueness and significance) and cleverness (p-values) before common sense (personal experience detailed in case reports); Good Lord, deliver us” [13].

CONCLUSION

I appreciate the limited space in print-journals as well as I recognize the large volume of case reports seeking publication. What I fail to comprehend is the rather miscalculated, business-sense informed, case report aversion by major medical journals. Case reporting has earned its place in the annals of medical literature: many medical breakthroughs hinged on its shoulders. It is an old and trusted method of medical reporting. The founding fathers of medicine believed in it, I will believe in it too.

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Author Contributions
Segun Adeoye – 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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Proliferative vitreoretinopathy secondary to degenerative retinoschisis: A case series

Mansvkhlal Amer, Safia Ahmed, Carpi Olali, Sohail Ahmed, Gupta M

ABSTRACT

Introduction: We report two cases proliferative vitreoretinopathy that occurred as complication of degenerative retinoschisis. Case Series: Two patients; a male and a female with vitreo-retinal hemorrhage seen in our clinic were evaluated and retinoschisis found as the only cause of the bleeding. Both were treated with Argon barrier photocoagulation which arrested the hemorrhage and resolved the visual symptoms. Conclusion: It has been suggested that retinal ischemia occurs in retinoschisis and this triggers upregulation of growth factors and subsequent formation of new vessels. This hypothesis would need to be tested to ascertain the particular growth factors, that trigger this process and if other cytokines and interleukins also play a role as the process is a vitreoretinopathy.

Keywords: Retinoschisis, Vitreous hemorrhage, Growth factors, Vitreoretinopathy

INTRODUCTION

Degenerative retinoschisis rarely present with symptoms because of their peripheral location [1]. Sometimes, however, advanced cases may present with a large peripheral visual field defect corresponding to the area of retinoschisis and in some cases, it could be confused with retinal detachment by the referring individual, [1, 2]. Patients who present with vitreous haemorrhage related symptoms on the other hand usually have vitreo-proliferative conditions such as diabetes mellitus, retinal vascular occlusion, hemoglobinopathies, coagulopathies or tumors [3–5].

We present two cases of retinoschisis who had ocular symptoms related to vitreous hemorrhage which occurred as a complication of the retinoschisis only and the treatment we instituted to arrest this complication.

CASE SERIES

Case 1: A 58-year-old male presented with a week history of triangle-like floater in his right eye. His vision was otherwise normal and there was no medical history of significance. His corrected visual acuity in both eyes was normal at 20/15 in the right and 20/20 in the left eye. Dilated fundoscopy revealed what looked like three closely spaced macroaneurysms in the inferior-temporal periphery with retinoschises adjacent to the macroaneurysms in the right eye (Figure 1A). The neovessels were located in the inner layer of the...
retinoschisis. Retinoschisis was also found in the fellow eye located inferiorly, but there was no associated neovascularisation.

He was kept on annual follow up, but two years after the initial diagnosis, developed proliferative vitreoretinopathy with new vessels and some fibrous tissue at the edge of the schises in the right eye. Fluorescein fundus angiography showed COATS-like disease (Figure 1B).

Clinically, there was progression of neovascularisation and glial tissue proliferation along the retinoschises. There were some vascular anomalies noted in the inner layer of the retinoschisis, but no holes in either layer. A few months later, repeat retinal angiograms showed areas of non-perfusion in and around the retinoschisis with hyperfluorescence of the new vessels. He had barrier argon laser photocoagulation and has since remained stable with normal vision and no vitreous hemorrhage.

**Case 2:** A 60 year old female patient was referred because of bleeding in the lower mid-peripheral retina of the left eye noticed by an optician during routine visit. She had had no ocular problems but was on treatment for hypertension. Her vision with correction was normal at 20/25 in the right eye and 20/20 in the left eye.

Dilated fundoscopy, revealed bilateral retinoschisis located inferior-temporally in the right eye and inferorly in the left. There was evidence of neovascularization in the inner layer of the retinoschisis associated with small vitreous hemorrhage in the left eye (Figure 2A).

The fluorescein angiography showed hypoperfusion in the area of the retinoschisis in the left eye and hyperfluorescence in the location of the new vessels (Figure 2B).

In order to prevent further vitreous hemorrhage, the patient underwent barrier argon laser photocoagulation. The patient has remained symptom free with no further vitreous hemorrhage in the last 18 months after the laser treatment.

**DISCUSSION**

Degenerative retinoschisis affects about 5-7% of the adult population and are usually incidental findings in clinical settings [6]. Indeed, in Bayer’s [2] studies of 108 cases of retinoschisis, none had symptoms primarily due to the disease.

Complications however, can sometimes result in retinal detachment or rarely vitreous hemorrhage producing symptoms. These are usually limited to the reticular (bulla and cyst in nerve fibre layer) form rather than the typical (flat and cyst in outer plexiform layer) form of the disease. Retinoschisis is responsible for less than 2.5% of all rhegmatogenous retinal detachments and the presence of holes in both outer and inner layers of the retina is the greatest risk factor as this can allow a large amount of fluid from the vitreous cavity to migrate into the potential space, resulting in the detachment [7–9].

Vitreous hemorrhage resulting from retinal neovascularisation as seen in our two cases is rare. Campo [10] and colleagues using retinal fluorescein fundus angiography demonstrated that the neovascularization was on the surface of the cyst and this accounted for the vitreous hemorrhage. They also suggested that the neovascularization in bullous retinoschisis may occur because of hypoxia induced by the combination of chronic retinal elevation and capillary non-perfusion.

Growth factors are known to mediate various inflammatory and angiogenic processes in biologic systems and of particular importance is vascular endothelial growth factor (VEGF) because of its clinical relevance. In the eye, the potent angiogenic factor VEGF has since been recognized to be produced by many retinal cells such as endothelial cells, pericytes, retinal pigment epithelial cells, Muller cells and astrocytes [11–12]. VEGF is upregulated in retinal ischaemic conditions such as diabetic retinopathy, retinal vein occlusion, ruberosis irides, and retinopathy of prematurity and is recognized as the main cytokine inducing the retinal neovascularization that results [13–14]. Although it has five isoforms (121, 145, 165, 189, and 126), the 126 is the predominant. It has high affinity for the 180-kDa fms-like tyrosine kinase (also called Flt-1 or VEGFR1) and 200-kDa kinase insert domain-containing receptor (KDR) also known as fetal liver kinase Flk-1 or VEGFR2. KDR transduces the signals for endothelial proliferation and chemotaxis [15].

Hypoxia stimulates VEGF mRNA expression through binding of Hypoxia-inducible factor 1 alpha subunit (HIF –α) to consensus and ancillary hypoxia –response elements (HREs) in the VEGF promoter and once expressed, VEGF stimulates endothelial cell proliferation and neovascularisation via the mitogen-activated protein kinases (MAPK) dependent pathway [16–17].
VEGF may also promote endothelial cell migration and vascular permeability with the resulting vessel leakage leading to interstitial edema and hydrops which in turn stimulates further VEGF production [18]. It is thought that VEGF promotes vascular leakage by several mechanisms including endothelial cell fenestrations, damage to tight junctions and the upregulation of the expression of Intercellular Adhesion Molecule 1 (ICAM 1) [19–20] (This protein is encoded by the ICAM 1 gene which encodes a cell surface glycoprotein that is typically expressed on endothelial and leukocyte-associated transmembrane protein).

Kun and colleagues [21] demonstrated high levels of interleukin 6 (IL-6) and protein in patients who had vitrectomy as part of the retinal detachment surgical procedures and suggested that these may play a role in the pathobiology of proliferative vitreoretinopathy (PVR). The role of platelet derived growth factor C (PDGF-C) in pathological angiogenesis independent of VEGF was shown by Li and colleagues in their study [22]. PDGF-C as they reported may have effect on vascular cells, tissue stroma fibroblasts and macrophages among others and have therefore been described as pleiotropic and versatile with potential for angiogenic therapy.

It is known that in the presence of a retinal hole, the PVR could be explained as the vitreous humour contains a number of cytokines including TNFa, TGFβ2, PDGF and interleukins; which when in contact with the RPE could stimulate RPE migration (and proliferation) and angiogenesis. In our two cases, no hole was seen in either side of the retinoschisis and therefore this may not be the reason for the neovascularization. Rather, retinal ischemia as earlier suggested may be the trigger for the upregulation of the growth factors and subsequent formation of new vessels.

This hypothesis would, however, need to be tested to ascertain the particular growth factor (s) that triggers this process and if other cytokines and interleukins also play a role as the process is a vitreoretinopathy.

Also, in proliferative retinopathies, VEGF dependent angiogenesis can be induced by inflammation, alteration of shear stress on blood vessels, and glycosylation of proteins; mechanisms that are independent of retinal hypoxia [23–24]. In retinoschisis, the splitting of the retina results in the retinal vessels attached to the inner layer floating freely in the vitreous cavity and being subjected to tractional forces, with eye movements. This could exhibit and alter the shear stress on these blood vessels and thereby result in upregulation of VEGF, with subsequent angiogenesis once the critical level of the cytokine is attained.

Our two patients responded to Argon laser treatment. As demonstrated by Lip et al. [14] as well as Aiello et al. [25], VEGF levels reduced markedly following laser treatment in their studies of patients with proliferative diabetic retinopathy. The effect of argon laser treatment on the level of any mediator found in retinoschisis related vitreoretinopathy would equally be a significant revelation.

**CONCLUSION**

Retinal ischemia, hypoxia and subsequent neovascularization is a cascade mediated by cytokines of which VEGF is the most widely studied and the one with current clinical application in ophthalmology. It has been suggested that retinal ischemia occurs in retinoschisis and this triggers upregulation of growth factors and subsequent formation of new vessels. This hypothesis would need to be tested to ascertain the particular growth factors that trigger this process and if other cytokines and interleukins also play a role as the process is a vitreoretinopathy. Factors which can trigger vascular endothelial growth factor upregulation independent of hypoxia such as alteration of shear stress on blood vessels could also play a role in neovascularization in retinoschisis.

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**Author Contributions**

Mansvkhal Amer – 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**

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Bilateral traumatic distal femoral physeal slips: A very rare case presentation

Anil Mehtani, Jatin Prakash, Suresh Chand, Abhinav Sinha, Ajeet Singh, Harvinder Dev

ABSTRACT

Introduction: Salter–Harris type 1 injuries of distal femur are very rare accounting for a mere 7.7% of all distal femoral injuries. No report of bilateral slips in otherwise normal child has been ever reported. Case Report: We herein present a case of a six-year-old boy with one month old trauma that caused bilateral distal femoral physeal slips. Patient was managed conservatively to avoid any further physeal damage. The slips healed completely with excellent range of motion. Due to late presentation, the fracture was already in stage of healing and was not frankly mobile and therefore open reduction or closed manipulation of physis was not possible without causing additional injury to growth plate. Therefore, slips were managed conservatively. Conclusion: This case presents a number of unique features. Salter–Harris type 1 injury is uncommon in the age group presented in our case. Secondly, bilateral distal femoral slips in otherwise normal child have never been reported. Thirdly, it further shows that remodeling of slips presenting late is possible if the physeal damage is prevented and open reduction in late presenting slips is not required.

Keywords: Paediatric, Trauma, Distal femur, Physeal

INTRODUCTION

The distal femoral epiphysis is the largest and fastest growing epiphysis in the body. There is no inherent protection to the physis with all muscles and ligaments inserting directly to the epiphysis. Distal femoral injuries account for 7% of all pediatric trauma [1]. Distal femoral physeal injuries are even more rarer accounting for 1–6% of all physeal injuries and less than 1% of all fractures. Among these most common is Salter–Harris type 2 injuries. Salter–Harris type 1 injuries of distal femur are very rare accounting for a mere 7.7% of all distal femoral injuries [2, 3]. Also these injuries are common in newborns or adolescents [3]. Direct trauma to distal femur is uncommon mode of injury but may occur in road traffic accident or with falls. Such injuries are mostly seen in adolescents. In newborns, it is mostly due to birth trauma secondary to breech injury. Indirect injuries are most common mode of injury, mostly varus or valgus strains resulting in simultaneous compression of one side with distraction of other cause physeal separation. Most typically these are Salter–Harris type 2 with separation beginning in tension side and exiting from metaphysis of compression side. In most cases patients present with history of trauma with inability to bear weight on.
affected extremity. The knee is typically in flexion owing to hamstring spasm. Mostly the epiphyseal displacement is in the coronal plain producing a varus or valgus deformity. This is confirmed by a plain radiograph and treatment consists of closed manipulation followed by a long leg cast till fracture healing. In fractures which fail to reduce or with unstable reductions may be taken up for open reduction with 4.0 or 6.5 mm screws.

A bilateral distal femoral Salter–Harris type 1 injury has never been reported in literature. Multiple slips have been reported in metabolic disorders like scurvy, [4] or conditions causing generalized bone weakening like leukemia or myelodysplasia. However, we herein present one case of a six-year-old male child with bilateral traumatic distal femoral slips with normal metabolic parameters.

CASE REPORT

We herein present a case of a six-year-old boy, presented to us with mild knee and thigh swelling of left side and inability to walk for one month. Child’s parents gave history of fall from height of about 4–5 meter, 1 month back, following which patient was unable to stand and complained of pain in bilateral knee. There was history of massage and visits to professional bone setter. No history of any medical intervention was elicited. No history of bracing or plaster cast immobilization was given.

On examination, tenderness was present in bilateral thigh. Patient was bed ridden for one month. No abnormal mobility or crepitation was seen. There was swelling in bilateral distal thigh, left more than right. Range of motion at hip was normal. There were bilateral flexion deformity of about 20 degrees with further movement painful and attempt of any movement caused muscle spasm (Figure 1).

Radiographs of bilateral knees were taken (Figure 2). These demonstrated bilateral Salter–Harris type 1 physeal injuries. Routine laboratory investigations were essentially normal except for anaemia (hemoglobin 9.8) and mildly raise alkaline phosphatase (ALP-564). Calcium profile was normal, so were serum vitamin C levels (0.84 mg/dL).

Since patient came to us after one month of injury, fracture was already in the stage of healing and was not mobile. Therefore, closed manipulation could not re-align the physis. Open reduction could further damage the physis in the growing child. So the child was managed conservatively on long leg cast and non-steroidal anti-inflammatory drugs (ibuprofen 100 mg b.d.) for two weeks. After pain decreased, patient was given intermittent skin traction for flexion deformity with range of motion exercises. Both slips healed within four months. Even these completely displaced slips remodeled very well. At six month of follow-up, all the slips were completely remodeled without obvious deformity (Figure 3). Patient gained bilateral excellent range.

DISCUSSION

Distal femoral injuries account for 7% of all pediatric trauma [1]. Salter–Harris type 1 injuries are very rare accounting for only 7.7%. [2, 3] It is most often seen in two age groups, newborns and adolescents [3]. We here have presented a neglected case of bilateral slips in a six-year-old child. The mechanism of injury has been mostly a road traffic accident, sports injury or a fall from height. In our case it was a fall from height on bilateral knee in flexed position.

Bilateral slips have been noted in scurvy in literature [4]. However, our case did not show any evident metabolic abnormality. The case had normal calcium profile, and serum vitamin C levels.

The recommended treatment in acute slips has been close reduction with pinning. Closed reduction under general anesthesia with long leg casts has been tried for displaced fractures. However, series have reported rates of 43–70% of distal femoral fractures treated without internal fixation have displaced [5]. Unless a fracture is truly nondisplaced and stable, immobilization without fixation is no longer the treatment of choice [6].
However, Salter et al. have stated that when excessive manipulation appears to be necessary to achieve acceptable reduction, it is better to maintain growth potential and perform corrective osteotomy at a later date than to overstress the physis and cause more injury [7, 8]. Also, it has been recommended that a reduction should not be performed more than 10 days after the original injury. Since the injury was one month old a closed manipulation was not possible. Any attempt could have damaged the physis and this could have led to growth disturbances and angular deformities. In a younger child, acceptable alignment includes up to 20 degrees of angulation in the sagittal plane, less than 5 degrees of varus or valgus angulation, and no rotational deformity. Since the child in our case was young, had a sufficient remodeling potential, we managed the child conservatively. Once the pain subsided, the child was started on aggressive range of motion exercises, with intermittent skin traction for flexion deformity and lesion healed in four months.

CONCLUSION

This is a rare case report both in terms of presentation and management. Unusual presentation at six years of age with bilateral slips is not reported in literature to date. Also internal fixation had been treatment of choice in such cases. But excellent results in above case with conservative management demonstrates that if physeal damage is prevented, the bone has a great remodelling potential and open reduction might not always be required, especially in late presenting cases.

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Author Contributions
Anil Mehtani – 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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Abhinav Sinha – Acquisition of data, Revising it critically for important intellectual content, Final approval of the version to be published
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Syphilitic paroxysmal cold hemoglobinuria associated with peripheral gangrene: A diagnostic dilemma

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ABSTRACT

Introduction: Paroxysmal cold hemoglobinuria is a rare clinical entity; syphilitic association is even rarer. The lack of awareness amongst healthcare providers, and its fleeting course often mean that many a time, paroxysmal cold hemoglobinuria is not diagnosed. Case Report: Herein, we present a case of syphilitic paroxysmal cold hemoglobinuria with peripheral gangrene that necessitated amputation. We describe the atypical presentation, emphasize or diagnostic dilemma and report our therapeutic considerations. Conclusion: The report raises two important points (i) the lack of awareness of the clinical entity amongst healthcare providers (ii) the sub-optimal work-up done for late-onset dementia. We conclude by making the case for including the screen for syphilis in their work-up of patients presenting with aortic valvular disease, chronic dementia (especially of the early-onset type) and hemolytic anemia of the cold antibody variety.

Keywords: Syphilitic paroxysmal cold hemoglobinuria, Syphilis, Cold hemoglobinuria

INTRODUCTION

Paroxysmal cold hemoglobinuria (PCH) is a relatively rare clinical syndrome first described in 1854 by Dressler [1], its association with syphilis is even rarer; the first description of this association is credited to Goetz, in 1885 [1]. The causative biphasic hemolysin was first described by Donath and Landsteiner in 1904 [2]. The incidence of syphilitic PCH is higher in males, probably reflective of the greater prevalence of syphilis in males. It may complicate congenital as well as acquired syphilis. A number of cases of syphilis associated PCH have been documented, some of these have reported events of peripheral gangrene [3].

In the latter half of the 19th century, congenital and adult tertiary-stage syphilis were the most common cause of PCH. However, with the decreased prevalence of syphilis due to better population education and awareness, improved identification-contact tracing, reporting and monitoring, as well as the availability more effective treatment options for syphilis, incidence of the syphilis-induced PCH has declined significantly. In the present time, PCH is most often related to infections and neoplasms. Identified pathogens include: measles, mumps, influenza, varicella-zoster virus (VZV), cytomegalovirus (CMV), Epstein–Barr virus (EBV), adenovirus, parvovirus B19, Coxsackie A9, Haemophilus influenzae, Mycoplasma pneumoniae, and Klebsiella pneumonia [4–7]. Implicated neoplasms include: solid organ malignancy like small-cell lung carcinoma,
and hematopoietic disorders such as non-Hodgkins lymphoma (NHL), chronic lymphocytic leukemia (CLL), multiple myeloma and monoclonal gammapathies [8–12].

A study of children found that as many as 40% of immune hemolytic anemias were due to the Donath-Landsteiner (D-L) antibody [8, 13]. In the adult population, infections and neoplasms have been associated with the development of D-L antibody [3]. PCH is thought of as a unique form of immune hemolytic anemia (IHA) characterized by paroxysms of severe anemia and hemoglobinuria upon exposure to cold temperatures, due to massive intravascular hemolysis. The hemolysin antibody attaching to red blood cells (RBCs) in the cold and inducing hemolysis due to complement activation when the RBCs are warmed [1, 2, 4, 14]. Episodes are heralded by a combination of the following: sudden onset of back and abdominal pain, headache, leg cramps, fever, rigors, chills, nausea, vomiting, diarrhea, and esophageal spasms. The associated hemoglobinuria is often severe, hematuria is generally minimal or absent. Oliguria or anuria is indicative of renal insult. Cold urticaria and jaundice may also occur [15]. These generalized symptoms are attributable to the release of large quantities of hemoglobin from hemolyzed RBCs, which then act as an irritant to various tissues or precipitate causing renal tubular obstruction. Gangrene rarely occurs in cold autoimmune hemolytic anemia; occurring, it is usually associated with primary cold agglutinin disease [15].

The mainstay of management of PCH is avoidance of cold exposure, supportive care and treatment/control of the associated primary disorder (when applicable and feasible). Administration of warmed, packed RBC units for life-threatening hemolysis and symptomatic anemia is a common event in management. Use of washed RBC units has not been shown to improve transfusion safety, but may be employed if condition remains refractory to standard warmed products. Though corticosteroids have been used, it has not been shown to shorten the clinical course of PCH. Hydration, urine alkalinization and temporary hemodialysis may be required to enhance elimination of hemoglobin, and to prevent or treat complicating renal failure. Antihistamines may ameliorate symptoms of cold urticaria in PCH.

The lack of awareness amongst healthcare providers, and its fleeting course often mean that many a time, like in the case we present, PCH goes undiagnosed. We present a case on patient with PCH secondary to syphilis, and complicated by peripheral gangrene necessitating an amputation.

CASE REPORT

A 91-year old female was presented to the emergency room with complaint of a two-week history of abdominal pain and vomiting which started soon after left trans-metatarsal amputation for gangrene of unknown cause in another hospital. The history was obtained from her daughter due to the patient’s cognitive impairment from baseline dementia. A 10-day history of intermittent passage of red-colored urine was also noted. Abdominal pain was diffuse, dull-to-achy, associated with constipation, vomiting was non-projectile, related to and aggravated by meals and contained recently ingested meals, gastric secretions and phlegm. No relieving factor was identified. No hemoptysis, melena, hematochezia were reported. No other organs or system-related symptoms. Upon further enquiry, daughter recounted were present a remote history of similar passage of red urine that resolved spontaneously. Medical history was significant for hypertension and dementia. As indicated above she underwent left trans-metatarsal amputation for a gangrenous foot two weeks before presentation. Family and social history was unremarkable. She had no known drug, food or environmental allergies. Home medications included amlodipine, metoprolol, omeprazole and laxatives. She reported no history of transfusion of blood or blood products.

On examination, vital signs of the patient were essentially normal, but she was pale, icteric and cachectic. HEENT, chest, cardiovascular and abdominal (no organomegaly) with rectal exam was normal, neurological exam was consistent with mild-moderate dementia, initially suggestive to Alzheimer’s or vascular etiology. Cutaneous and integumentary exam were significant for reddish blue nose tip coloration and acrocyanosis of left index and ring fingers, and a healed post-amputation stump on the left foot (Figure 1). Significantly, no telangiectasia, peri-ungual lesions, or clubbing of extremities was noted.

Initial laboratory tests reported anemia ([Hb]/Hct=6.1/18), hemoglobinuria (without hematuria), reduced haptoglobin, increased LDH, reticulocytopsis (7.8%) peripheral film revealed normochromia, normocytosis, without polychromasia, few schizocytes and neutrophils with toxic granulations were present, and indirect hyperbilirubinemia (3.5 g/dL), all suggestive of intravascular hemolysis. Abdominal ultrasound reported some thickening of gall bladder wall associated with biliary sludge. EKG showed normal sinus rhythm, with a ventricular rate of 67 bpm. Echocardiogram was essentially normal. An initial impression of intravascular hemolysis (autoimmune versus drug-induced hemolytic anaemia) with uremic gastritis/gastropathy was made. Patient’s daughter was unsure of medication exposure during the patient’s admission in the previous hospitals. Gastroenterology and hematology consults were called and obtained. Continuity care and medical records on her recent admission for gangrenous foot (especially antibiotic exposure and vascular studies) were not available at that point. Electronic medical records (EMR) are currently not integrated or cross-accessible across hospital. There is often a lag between need for, patient’s
signing of release document, call for and access to records on prior hospitalization in other hospitals.

Patient was transfused with six units of packed red blood cells to keep hemoglobin >8g/dL, as intravascular hemolysis continued. Consistent with the initial impression of an autoimmune etiology, corticosteroid (prednisone) therapy was instituted. During the hospital course, the acrocyanotic digits progressed to dry gangrene with sharply demarcated borders (Figure 2). Later in the course of her hospital stay, the daughter gave a history associating her prior gangrenous episode with the onset of winter, as well as a remote history of passage of red-colored urine. In view of the interval events and new history obtained, an impression of PCH was made and work-up to confirm the diagnosis and to identify the trigger was pursued. Repeat enquiry did not elicit a remote or recent history of syphilis.

The patient was subsequently kept warm using blankets, mittens and stockings. The Donath–Landsteiner reaction was positive, as was a direct Coombs test performed at room temperature. Screening and confirmatory test for syphilis was positive (significant RPR titer of 1:8 and a positive FTA test), rheumatology test battery was negative (rheumatoid factor, ANA, IgG/M/A, β₂microglobulin), hepatitis profile and HIV screen were negative. Hematological tests including bone marrow aspiration and serum electrophoresis excluded any underlying lymphoproliferative disorder or monoclonal gammopathy. A rheumatology consult was called to assist with evaluation for autoimmune and collagen vascular disorders.

Lumbar puncture to evaluate for neurosyphilis was not performed as her healthcare proxy requested only comfort care. Pulsed vascular recording (PVR) and arterial duplex of the extremities showed mild tibial and disease in the right leg, as well mild femoro-popliteal and tibial disease in the left leg. A bone scan study was suggestive of Paget’s disease; no malignancy pattern was discerned. The patient was treated with penicillin 2.4 M IU IM weekly for three weeks, Ceftriaxone 2 g daily for 10 days. The gangrenous digits (right index and ring fingers) remained well demarcated, requiring surgical amputation. The dose of prednisone was gradually tapered with sustained control of hemolysis. As no other pathological cause of vascular occlusion was identified, patient was discharged to center for sub-acute rehabilitation and advised to avoid future cold exposure. At discharge, she was no longer pale or icteric, intravascular hemolysis had completely resolved, this was evident in her stable Hb/Hct of 10.3/29, reticulocyte count of 1.9% and an essentially normal urinalysis. The patient was lost to follow-up after sub-acute rehabilitation.

**DISCUSSION**

We lay no claim to the first description of syphilitic paroxysmal cold hemoglobinuria, nor to the first report on a complicating gangrene, but our report raises two important considerations.

Firstly, the lack of awareness amongst healthcare providers of this diagnostic entity, and the transitory nature of PCH often mean that many a time, like in the case we present, PCH is often not diagnosed. The patient had prior episodes of passage of red-colored urine, she also had gangrene of her left foot necessitating a transmetatarsal amputation two weeks before presentation (all indicative of a possibility of PCH). In hindsight, the entire symptomatology, (gastrointestinal symptoms inclusive) was consistent with PCH. Taking a detailed history of presenting complaints, utilization of all diagnostic clues, and informational continuity of care are invaluable to making accurate diagnoses.

Secondly, the patient was not diagnosed of syphilis, even in the setting of moderate dementia. Syphilitic dementia complicates 25–40% of untreated syphilis [16]. The report makes the case for investigating every case of dementia, irrespective of the age of presentation or presumed surety of diagnosis. Many physicians (like we did initially at presentation) often attribute dementia to an Alzheimer’ or vascular etiology, especially when dementia-related symptoms is not the presenting complain or when dementia or cognitive decline appear age-appropriate. It is logical to infer that early-onset dementia or cognitive decline and acute changes in mental state...
status are more likely to receive extensive investigation. In this patient, physical examination did not reveal any sign directly relatable to syphilis, and absent a past medical history of syphilis, considering a diagnosis of syphilitic-PCH was far-fetched. The only clues were the presence of dementia and on hindsight, syphilis-relatable PCH.

We did not confirm syphilitic dementia in this case since family refused diagnostic lumbar puncture, the diagnosis is highly likely though. This impression informed our choice of, and duration of antimicrobial therapy. Neuroimaging findings in neurosyphilis are non-specific, these include cortical and subcortical infarcts, cortical atrophy, hydrocephalus, leptomeningeal enhancement associated with a clinical meningitis, and arteritis [16]. The more specific finding of leptomeningeal and cerebral gummas are also not diagnostic. We did not perform neuroimaging on the patient, as dementia was not the presenting complaint. Furthermore, absent the consent to perform lumbar puncture in the setting of dementia in a syphilis sero-positive patient, commonsense advised assuming a neurosyphilis diagnosis and treating as such.

Though the patient was eventually lost to follow-up, it is our expectation that with clinical cure of syphilis, the frequency and severity of her bouts of PCH, as well as vaso-occlusive events will reduce over time, and assuming syphilis is the sole cause of her PCH, a complete resolution is likely. We had no cause to suspect another causation giving the extensive investigation we conducted.

CONCLUSION

In the light of our discussion primary syphilis may be on the decline, but the same cannot be said of tertiary syphilis from untreated/sub-optimally treated syphilis acquired many years (sometimes decades) earlier, when primary syphilis was prevalent. Sporadic cases of tertiary syphilis have been reported in medical literature in the recent past. Physicians should have a low threshold for entertaining the diagnosis and especially including screening for syphilis in their work-up of aortic valvular disease, chronic dementia (especially of the early-onset type) and hemolytic anemia of the cold antibody variety. We highlighted the importance of exploiting all diagnostic clues when cases present a diagnostic dilemma. In concluding, we hope this paper re-brings to limelight the existence of a seemingly forgotten disease entity- syphilitic paroxysmal cold hemoglobinuria.

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Author Contributions

Segun P Adeoye – 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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Prabhjot Manes – 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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A rare case of myositis ossificans progressiva presenting as multiple progressive contracture

Anil Mehtani, Jatin Prakash, Suresh Chand, Abhinav Sinha, Ajeet Singh, Harvinder Dev

ABSTRACT

Introduction: Myositis ossificans progressiva is a rare disease characterized by formation of areas of calcification in soft tissue such as ligaments, muscles and tendons. There are a few sporadic case reports all over the world. The disease has an incidence of less than 1 in 10,000,000 population. Myositis ossificans progressiva is a disease of early childhood. The disease is often progressive with multiple soft tissue contracture and subsequent death by third or fourth decade of life. There is no effective treatment till date. Case Report: We herein present a case report of myositis ossificans progressiva presented to us with numerous lumps and shoulder and hip contracture. Patient was treated conservatively on bisphosphonates. No progression of lumps or swelling were seen after one year of follow-up. Conclusion: This presents a case report of a very rare disease. In most cases there is history of any trauma or inciting factors that result in formation of myositis mass. This case, however, presents a very aggressive form of disease with patient developing spontaneous swellings and progressive contractures.

Keywords: Myositis ossificans progressive, Multiple contractures, Pediatric

INTRODUCTION

Myositis ossificans progressive also known as fibrodysplasia ossificans, Münchmeyer syndrome, stiff-man syndrome, and progressive ossifying myositis is a very rare and crippling disorder. The disease mostly involves patients in their first decade, progressing rapidly to involve muscles, tendons and ligaments. Patients are generally, confined to wheel chair life and mostly live till fourth to fifth decade [1]. The disease is very rare with an incidence of less than 1 in 10,000,000 population and around 700 cases have been reported in literature to date. We report a young boy who presented with very rapid progression of disabling muscle contractures diagnosed clinicoradiologically as myositis ossificans progressiva.
by more lumps in back and lower legs causing restriction of motion of lower spine and hip joint. Patient developed ulcers in the skin overlying lump at scapular region (Figure 1). There was no history of fever, bleeding tendencies, hematuria, seizures, deafness, mental retardation, joint swelling, rash, abdominal colic, fractures, thyroid swelling, or any drug intake.

Physical examination revealed multiple lumps in the neck, scapula, back, iliac region, knee and trunk. There was kyphotic deformity in dorsal spine and muscle contractures involving the sternocleidomastoids, latissimus dorsi, pectoralis major (Figure 2), and the cervical muscles, with restricted abduction and internal rotation of both shoulders. There were bilateral flexion deformities of hip joint. He also had short great toes (Figure 3). Chest expansion was restricted in spirometry testing.

Routine laboratory investigations were normal. Serum calcium chemistry was also normal. The spirometry showed moderate restriction and electrocardiogram cardiac echo were essentially normal.

X-ray of cervical spine showed calcific strands around both shoulder joints (left more than right) and in the paraspinal regions (Figure 4). A detailed skeletal survey of the body revealed calcification in the soft tissues surrounding the cervical region, left shoulder, in the anterior chest wall, the thorax, and the paraspinal muscles and knee (Figure 5). Considering both the clinical

and the radiological features, sporadic myositis ossificans progressiva was diagnosed. The child was treated with graded physiotherapy. Bisphosphonates were added. As there was no acute flare-up, steroids were not given. The patient has been followed-up for one year. No new lumps have been noticed after starting of bisphosphonates.
Myositis ossificans progressiva is a rare, progressive, crippling disorder, with an incidence of less than 1 in 10,000,000 population. The condition has a male preponderance. This is a mesodermal disorder with defect in reparative process [2, 3] causing heterotopic ossification which usually begin in 5–7 years of life [4]. Our case however, has history of ossifications from first year of life.

The case presented late to us with all characteristic features of short great toe, multiple contractures and multiple ossifications. The X-rays were also characteristic in showing the lesion. Based on these characteristic findings, the diagnosis was pretty straightforward. Otherwise diagnostic errors have been documented in up to 87% of myositis ossificans progressiva cases worldwide with cancer being the most common erroneous diagnosis [5]. This is very important to note as error in diagnosis would lead to unwanted biopsies doing more harm than good.

Initial symptoms include painful lumps, mostly starting cranially in neck and shoulder region and progressing caudally involving scapula, trunk and hip regions. This is sometimes associated with stiffness and decreased mobility at joint site resulting in progressive contractures [2]. This case also had this characteristic pattern. Mostly these swellings are preceded by local trauma, injection site, biopsy or a venipuncture site, however, no such inciting factor was observed [6].

Associated skeletal features of great diagnostic significance include short hallux with synostosis, hallux valgus (75–90%), and short thumbs [2]. Kyphoscoliosis, with restricted shoulder and pelvic girdle movements and restrictive pulmonary disease, can occur. Mental retardation, alopecia, and cardiac conduction defects are other associations [4]. This case had short great toes, no mental retardation or conductive deafness.

Radiological investigations are characteristic in myositis ossificans progressive. There is microdactyly of big toes (90%) and thumbs (50%), progressive fusion of the posterior arches of the cervical spine, narrowed antero-posterior diameter of lumbar vertebral bodies, with or without bony ankylosis with soft tissue calcification at multiple sites [7].

There is no effective treatment till date. Multiple treatments have been tried. Steroids are useful in acute flare-ups, bisphosphonates are thought to decrease ectopic calcifications. This was observed in our case as well when oral bisphosphonates have stopped further progression in a short follow-up of one year. However, longer follow-up would be more helpful. Some newer and investigational drugs include antiangiogenic agents such as squalamine, thalidomide, COX-2 inhibitors, BMP4 antagonists, and noggin and gremlin gene therapy. However none of them has a proven efficacy. Surgeries at large are contraindicated including procedures like biopsy. Surgical release of contractures is recommended.
only if joint movement is severely impeding movement or there is nerve impingement and this is not without increased risk of further ossification [8, 9].

CONCLUSION

This presents a case report of a very rare disease. In most cases there is history of any trauma or inciting factors that result in formation of myositis mass. This case however presents a very aggressive form of disease with patient developing spontaneous swellings and progressive contractures. The disease was controlled on bisphosphonates and no new swelling developed in follow-up of one year.

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Anil Mehtani – 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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Imaging findings in a case of cystic neoplasm of pancreas: A case report

Pooja Gupta, Devika Gupta, Kamlesh Kumar Singh, Satish Mendonca

ABSTRACT

Introduction: The spectrum of cystic neoplasms of the pancreas encompasses a wide range from benign to malignant. Serous cystadenomas are benign cystic tumors of the pancreas.

Case Report: A 54-year-old female presented with pain in epigastrium with a lump in mid upper abdomen. On imaging there was a large, lobulated, well defined, multicystic, heterogeneously enhancing mass arising from the body of pancreas which was subsequently excised and on histopathological examination was found to be a serous cystadenoma of pancreas.

Conclusion: The aim of this case report is to highlight the characteristic radiological features in cystic neoplasms of the pancreas along with a review of the relevant literature.

Keywords: Serous cystadenoma, Micocystic, Oligocystic

INTRODUCTION

There are various cystic neoplasms of the pancreas which can be benign to malignant. The cystic masses of pancreas can be pseudocysts, mucinous cystic neoplasms, serous cystadenoma and intraductal papillary mucinous neoplasms. The most common cystic neoplasms are mucinous followed by the serous cystadenomas. With the recent advances in imaging techniques more cases of pancreatic cystic neoplasms are being identified. Herein, we have a case of a middle aged female who presented with pain and lump in the epigastrium and on subsequent evaluation was found to have a cystic neoplasm in the body of pancreas.

CASE REPORT

A 54-year-old female was presented with pain in epigastric region of six months duration and nausea, anorexia, weight loss and lump in mid upper abdomen of two months duration.

On admission to the hospital, chest and abdominal X-rays were performed which were inconclusive. Routine laboratory studies including hematological and biochemical parameters were normal. Serum amylase was within normal limits. Following this an ultrasound of the abdomen was performed. Ultrasound showed a well-delineated, multi-loculated, cystic lesion of size 5.2x6.2x7.7 cm in the body of the pancreas. The cysts were all subcentimetric giving a honeycomb appearance to the
lesion (Figure 1). Subsequently, a barium meal study was done which showed a smooth extrinsic indentation on the lesser curvature of the stomach (Figure 2). Contrast-enhanced computed tomography scan showed a large, lobulated, well defined, lesion with multiple subcentimetric non enhancing hypodense cystic areas with enhancing hyperdense walls in the body of pancreas (Figure 3). Multiple discrete foci of calcifications were seen within the lesion (Figure 4). Anteriorly the lesion was reaching till anterior abdominal wall, posteriorly it was abutting the splenic vein, superiorly it was abutting the segment IV of liver and inferiorly it was abutting lesser curvature of stomach. After complete investigations, patient was diagnosed as a case of cystic tumor of pancreas and she was operated upon with resection of the lesion. Histology of resected pancreatic tissue revealed multicystic lesion in the body of pancreas containing cysts less than 1 cm in size lined by small flat to cuboidal cells (Figure 5). There was no architectural or

Figure 1: Ultrasound of the abdomen showing a 5.2x6.2x7.7 cm (APxTRxCC) multi-loculated, cystic mass in the body of the pancreas.

Figure 2: Barium meal showing a smooth indentation on lesser curvature of the stomach.

Figure 3: Contrast enhanced computed tomography of the abdomen showing a multicystic structure with enhancing walls and septae in the body of pancreas.

Figure 4: Non-contrast computed tomography scan of abdomen showing a multicystic lobulated mass in the body of pancreas with calcifications within the lesion.

Figure 5: The resected lesion showing multiple cysts lined by a single layer of flat to cuboidal low epithelial cells having pale to clear glycogen-rich cytoplasm (H&E stain, x400).
cytological atypia noted. Based on the imaging findings and further confirmation by histopathology a diagnosis of serous cystadenoma of pancreas was made.

DISCUSSION

Serous cystadenomas are benign tumors of the pancreas. There is strong female predominance with a sex ratio of 2:1. The most common presenting symptom is vague abdominal pain. A palpable epigastric mass is present in nearly two thirds of cases.

Serous cystadenomas constitute 25% of all cystic tumors of the pancreas the majority of which are found in females [1]. The most common site of serous cystadenomas is the head and tail of pancreas. In our case, the lesion was present in the body of the pancreas. They are seen almost exclusively after 35 years of age and 82% occurs after 60 years of age. The diagnosis of serous cystadenoma can be done by various imaging modalities such as ultrasound, computed tomography (CT) scan, and endoscopic ultrasound. On CT scan 30% of cases have a fibrous central scar with or without a characteristic stellate calcification which is considered pathognomonic for serous cystadenoma [2]. On contrast-enhanced CT scan, there is enhancement of the septae [3]. Serous cystadenomas have three morphologic patterns: polycystic, honeycomb, and oligocystic [4]. In 70% of cases, serous polycystic adenomas consist of a collection of cysts (usually more than six) which range from a few millimeters up to 2 cm in size [5]. On non-contrast CT, they appear as hypodense, cystic masses that frequently show calcifications [6]. The honeycomb pattern, seen in approximately 20% of patients, is characterized by numerous subcentimeter cysts that cannot be individually distinguished by cross-sectional imaging. Therefore, cysts having the honeycomb pattern are seen as well defined lesions with soft-tissue attenuation seen on CT [7]. Oligocystic have fewer larger cysts which are usually greater than 2 cm in diameter.

Differential diagnostic features that help in distinguishing serous from mucinous cystic tumors include older age group, and presence of multiple (>6) small cysts in case of serous cystadenoma as opposed to mucinous cystadenomas in which cysts are larger and fewer in number.

Patients with serous cystadenoma are thought to have an excellent long-term prognosis. Hence is important to differentiate serous cystadenoma from other cystic tumors.

CONCLUSION

Serous cystadenomas are benign tumors of pancreas which present as well defined multicystic lesions predominantly in the body of the pancreas. They have a good long term prognosis and hence need to be well differentiated from other cystic lesions of the pancreas on various imaging modalities.

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Urinary bladder perforation due to encrusted cystitis: A rare entity

Faouzi Mallat, Wissem Hmida, Khaled Ben Ahmed, Sarra Mestiri, Faouzi Mosbah

ABSTRACT

Introduction: Encrusted cystitis is extremely rare and severe chronic infection of the bladder causing intolerable function and serious consequences for the patient. Case Report: Herein, we report a new case of encrusted cystitis complicated by rupture of the bladder, septic peritonitis and uroperitoneum, and digestive fistula in a 57-year-old male and review literature. Conclusion: Encrusted cystitis is a severe entity that its evolution is unpredictable and it may be complicated by urinary bladder rupture that should be considered.

Keywords: Encrusted cystitis, Bladder rupture, Septic peritonitis, Uroperitoneum, Digestive fistula

INTRODUCTION

Encrusted cystitis is a severe chronic infection of the bladder. It is a very rare form of chronic cystitis [1]. It is characterized from a functional perspective by the usual symptoms of cystitis and inconstantly by the elimination of encrusted crystals and even stones during micturition. Calcified plaques composed of calcium salts including phosphate, carbonate, and ammonium-magnesium salts in the bladder mucosa accompanied with inflammation and ulcerations characterize pathologically encrusted cystitis. Despite its increasing incidence in nineties, especially in immunodepressed patients and renal transplant recipients, less than 100 patients have been reported in the published articles, mainly single case reports. Etiology of this rare entity is presently controversial. A few hypotheses exist to explain this phenomenon. The most popular of them proposes combined action of mucosal alterations and microorganisms that split urea forming ammonia and thus creating an alkaline environment leading to the disease by precipitation of calcium salts. Corynebacterium group D2 was denoted as the most frequent culprits [2]. Therapeutic strategies are not well-defined owing to the rare occurrence of this entity. No cases of urinary bladder rupture due to encrusted cystitis have previously been reported to our knowledge.

CASE REPORT

A case of a 57-year-old male without pathologic past history was presented with a peritonitis secondary to rupture of an encrusting and necrotizing cystitis. He had experienced dysuria, suprapubic pain, intermittent macroscopic hematuria, elimination of stones, anorexia and weight loss of about 12 kg over the previous four months.

Our patient initially presented to the accident and emergency department early in the morning with a sudden onset of sharp and constant lower abdominal pain, mostly located in the hypogastrium. The pain became worse and was associated with symptoms of oliguria and dribbling. On examination, there was generalized tenderness,
guarding and rigidity in the lower abdomen with positive rebound tenderness.

All the initial blood results were normal but the white blood cell count was $32 \times 10^9$/L. Urinalysis revealed a pH of 8.5 and was positive for the presence of blood and leukocyte esterase.

Abdominal radiograph was inconclusive. Sonogram of bladder revealed thickening of bladder wall with two distinct layers. Superficial layer (white arrow) is echogenic, corresponding to encrustation of urothelium. Underlining layer (black arrow) is hypoechogenic, corresponding to detrusor.

Computed tomography (CT) scan showed free fluid in the abdomen with a thickening of a limited segment of the small bowel, thick calcification of urothelial wall, not mobile in prone position (Figure 1) and moderate bilateral hydronephrosis.

Based on the clinical and radiological findings, the patient underwent laparotomy.

Exploratory laparotomy showed the cause of the patient’s symptoms—intraperitoneal rupture of the urinary bladder. We revealed an area of focal necrosis, thickening and calcification of the bladder wall leading to peritonitis and uroperitoneum (Figure 2). The lesion was found to be completely adhered to the bladder wall making resection difficult. It had necrotic edges and a petrous consistency when cut. A fistula between the bladder and the sigmoid has been identified (Figure 3). Partial resection of the lesion was carried out. The defect in the wall of the urinary bladder was sutured and a colostomy was associated there.

Definitive histopathological study reported calcic crystal deposit with data of non-specific chronic cystitis resulting in diagnosis of encrusting and necrotising myositis (Figures 4–6).

In postoperative period, the patient was managed with third generation cephalosporins and aminoglycoside. Corynebacterium (group D2) was present in the specific culture.

The evolution was rapidly fatal on the fifth postoperative day by septic shock.

**DISCUSSION**

Encrusted cystitis is a rare and severe inflammation of the bladder mucosa [1, 2]. It was described first in 1914 as a more or less localized ulcerated inflammation of the
bladder wall with calcium phosphate deposits on the ulcerated surface and walls [3].

The fundamental factor for its existence is a precipitated salt deposit which requires alkaline urine. Currently *Corynebacterium urealyticum* is almost exclusively described in this disease [4]. *Corynebacterium urealyticum* is a gram-positive commensal microorganism of the skin [5]. Urinary infections due to this bacterium require three conditions to cause alkaline-encrusted cystitis: a clinical context with immunosuppression or prolonged antibiotic therapy; urologic procedures either surgical or endoscopic, and an inflammatory or neoplastic pre-existing lesion of the urothelium [5, 6]. The delay between the urologic procedure and the diagnosis can vary up to several years [5]. It was demonstrated that its active osteogenic process is reversible, probably by changing the tissue’s environmental conditions (reducing inflammation and eradicating infection), suggesting that *Corynebacterium urealyticum* infection can be considered the ‘primum movens’ of pathogenic mechanisms behind the bladder tissue calcification [4].

In some reported cases, this bacterium was not found because it was not specifically sought [7]. Our patient had all the clinical and histological features of *Corynebacterium urealyticum* infection.

Mitomycin C as a bladder chemotherapeutic agent can be a factor in the development of the disease [8].

On cystoscopy, encrusted cystitis is characterized by a marked inflammatory appearance of all or part of the bladder mucosa with ulcerations and whitish plaque corresponding to multiple calcified encrustations [2]. Ultrasound is sensitive for encrusted cystitis diagnosis and shows thickening and calcified lesions in the bladder wall. Computed tomography scan is a more sensitive technique to detect calcification even if it is thin or radiolucent on radiographs. It provides excellent visualization of the urothelial wall and calcification. It should be considered in deciding optimal treatment and in monitoring the regression of calcified plaques [9].

Encrusted cystitis is a severe chronic infection of the bladder causing intolerable function and serious consequences for the patient. It may be revealed by some non-specific complications such as urinary tract obstruction, macroscopic hematuria, renal failure. Encrusted cystitis may be complicated by rupture of the bladder, and to the best of our knowledge no previous cases of urinary bladder rupture due to encrusted cystitis have previously been reported.

In this case, the severe chronic inflammation, important edematous mucosa, huge calcifications associated with obstructive urinary problems long neglected may contribute to the fragilisation of the urinary bladder wall and its rupture.

Treatment consists of three complementary elements: treatment of infection, urine acidification and chemolysis, and elimination of calcified plaque that contains microorganisms. All Corynebacterium group D2 strains are sensitive in vitro to glycopeptides, vancomycin, and teicoplanin and have a similar effect. Calcified plaque contains high levels of microorganisms and limits in vivo antibiotic effectiveness. Surgical or endoscopic plaque removal is suggested. Transurethral resection of bladder plaque remains difficult with risk of segment rupture.

Bladder wall healing depends on calcium precipitation prevention and therefore on the prevention of calcium salt oversaturation in alkaline urine [10].

**CONCLUSION**

Encrusted cystitis is a rare and severe chronic inflammatory disease of the bladder. Its diagnosis is difficult and should be considered when presented with a long history of symptoms associating dysuria, suprapubic pain, intermittent hematuria, persistent elimination of stones. Urinary bladder rupture as a complication and a
circumstance of discovery of the encrusted cystitis is an exceptional condition.

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REFERENCES
Retroperitoneal desmoid tumor with unusual appearance as a spontaneous ureteral rupture and rare association with desmoid tumor of the breast

Faouzi Mallat, Wissem Hmida, Sarra Mestiri, Badreddine Sriha, Moncef Mokni, Faouzi Mosbah

ABSTRACT

Introduction: Desmoid tumors of retroperitoneum and breast are rare. Their association is exceptional and their management is controversial. Spontaneous ureteral rupture as a complication and a circumstance of discovery of the retroperitoneal desmoid tumor is a rare entity and poses management difficulties. To the best of our knowledge, this case is the first to present this exceptional association, and to report the ureteral rupture as a circumstance of discovery of the retroperitoneal desmoid tumor. Case Report: We present the first reported case, seen in a young girl without Gardner’s syndrome, in which upper ureteral rupture was the primary symptom of an aggressive retroperitoneal desmoid tumor. She underwent laparotomy for septic peritonitis secondary to rupture of the right ureter, in which initially right ureterocutaneostomy was done, later right nephrectomy was made; for recurrent right ureteral rupture. Imatinib (Glivec) 200 mg was prescribed. However, due to intractable diarrhea, the target therapy was discontinued. At follow-up, left breast desmoid tumor was diagnosed by mammography, sonography and magnetic resonance imaging scan; and confirmed by sonographically guided core biopsy; and large excision was made. At the last follow-up, radiological exams did not show any significant change in size of the retroperitoneal mass or recurrent tumor in the breast or other sites. Conclusion: Desmoid tumors can occur synchronously or metachronously in many locations, even the absence of Gardner’s syndrome. Their evolution is unpredictable and it may be complicated, in the case of retroperitoneal desmoid tumor, by spontaneous ureteral rupture that should be considered.

Keywords: Retroperitoneal desmoid tumor, Spontaneous ureteral rupture, Breast desmoid tumor

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INTRODUCTION

Desmoid tumors are benign myofibroblastic neoplasms originating from the muscle aponeurosis and classified as deep fibromatoses [1]. They consist 3% of all soft tissue tumors and 0.03% of all neoplasms [2].

We present a very particular case of a young female patient, without Gardner’s syndrome, presented with spontaneous rupture of the right ureter due to a retroperitoneal desmoid tumor. At follow-up a second location in the left breast was discovered to the best of our knowledge, these aspects were never reported in literature.
CASE REPORT

A 24-year-old female patient was initially admitted to the emergency department with persistent right abdominal pain 48 hours. The patient had been suffering vague upper abdominal pain and discomfort for two years with fullness of upper abdomen part for the preceding one month; and had undergone two colonoscopic examinations in those two years, which did not reveal any colorectal polyps and hence excluded familial adenomatous polyposis.

There was no history of weight loss, loss of appetite, abdominal trauma or oral contraceptive intake. No family history of similar ailment or malignancy was reported.

An abdominal computed tomography (CT) scan (Figure 1) was performed which revealed large, hypodense and heterogeneously enhancing mass (138×98×112 mm) with an exophytic component arising from retroperitoneal space, extending up to the mesenteric vessels and encased them, compressing the right kidney, and also abutting the anterior abdominal wall. Both ureters were encased in the infiltrative tumor, with extravasation of radiocontrast from the ureterocutaneostomy catheter showed no extravasation of radiocontrast from the ureter. The CT scan. The patient underwent laparotomy which revealed large, hypodense and heterogeneously enhancing mass (138×98×112 mm) with an exophytic component arising from retroperitoneal space, extending up to the mesenteric vessels and encased them, compressing the right kidney, and also abutting the anterior abdominal wall. Both ureters were encased in the infiltrative tumor, with extravasation of radiocontrast from the ureterocutaneostomy catheter showed no extravasation of radiocontrast from the ureter.

The lesion was found to be completely adhered to many structures making total resection impossible. It had a petrous consistency when cut and partial resection of the lesion was carried out. At this site, the ureteric wall was hard, thickened, and adherent to the surrounding tissue; both ureters were encased in the infiltrative tumor and the right ureter was completely obstructed and ruptured 3 cm below the point where it crossed the common iliac artery. Right ureterocutaneostomy was performed.

After 16 days, antegrade opacification performed through the ureterocutaneostomy catheter showed no extravasation of radiocontrast from the ureter. The percutaneous retroperitoneal drain tube was removed. The next day, she was discharged without complications.

Histological and immunohistochemical findings confirmed the diagnosis of desmoid tumor.

The evolution was characterized by a rapidly septic peritonitis and uroperitoneum secondary to recurrent rupture of ureterostomy one month later confirmed by the CT scan. The patient underwent laparotomy which includes a radical nephrectomy plus removal of the entire upper ureter, and the postoperative course was uneventful. Since that, a double-J catheter (6F, 24 cm) was placed in the left ureter for prevention.

After the operation, imatinib (Glivec) 200 mg was prescribed. However, due to intractable diarrhea, the target therapy was discontinued and the patient refused any other treatment.

After 10 months of follow-up, the patient presented with the chief complaint of a palpable lump in her left breast. Mammography revealed a 2-cm spiculated mass in the upper inner quadrant of the left breast. Sonography showed a suspicious hypoechoic mass with irregular margins (Figure 2A). In magnetic resonance imaging (MRI) scan, the tumor was found to be of iso-intensity in the T1-weighted phase and of homogeneously low intensity in the T2-weighted phase and slow enhancement after contrast administration, and the marginal part showed very low intensity in both phases (Figure 2B–C). Sonographically guided core biopsy was performed, and the histopathologic findings observed in this case were typical of desmoid tumor of the breast (Figure 3). The patient underwent wide excision of the lesion.

Figure 1: (A) Computed tomography scan showing the ureterocutaneostomy, the site of the rupture of the right upper ureter with pelvic urinoma, (B) Contrast-enhanced Computed tomography image of the lower abdomen showing a soft-tissue mass in the mesentery (arrow), (C) The attenuation of the mass is similar to that of the psoas muscles. Coronal reformatted image demonstrates that the mesenteric vessels are displaced and encased by the mass.

Figure 2: (A) Sonography image showing irregular, hypoechoic mass with indistinct margins. Tumor is confined to breast parenchyma and does not involve retromammary structures. (B, C) In magnetic resonance imaging scan, the central part of the tumor was found to be of iso-intensity in the T1-weighted phase and of homogeneously low intensity in the T2-weighted phase and slow enhancement after contrast administration. The marginal part showing very low intensity in both phases.
Currently, the patient is doing well. Follow-up CT scan, done at interval of three months, did not show any significant change in the size of retroperitoneal and mesenteric mass or left ureteral obstruction or other complications.

There is no recurrent tumor in the breast or other sites. The patient changes the double-J catheter of the left ureter every six months and still refuses any medical or surgical treatment.

DISCUSSION

Desmoid tumors which belong to a family of fibroblastic proliferations that include a variety of fibromatoses, are benign tumors composed of fibrous elements. Desmoid tumors have an estimated incidence of 3.7 new cases per million people per year [3]. Desmoids may occur in the abdominal wall, the mesentery, or the retroperitoneum. Extra-abdominal desmoids may involve shoulder, thigh, buttock, or trunk whereas only a few cases describe a location within the breast [3, 4]. To the best of our knowledge, this is the first documented case of three locations of desmoid tumors in retroperitoneal space, mesentery and breast.

Desmoids are the most common primary tumor of the mesentery [5]. Most of these tumors occur sporadically; however, patients with Gardner’s syndrome are at higher risk than others. The incidence of abdominal wall and mesenteric desmoids in patients with Gardner syndrome ranges between 4–29%, and the tumors typically occur after abdominal surgery [3]. Desmoid tumors may be also associated with trauma and estrogen therapy [5]. In the present case, symptoms were sporadic, and the patient had no previous history of any of the above conditions. In addition, family history, upper gastrointestinal endoscopy, colonoscopy, and ophthalmoscopy were normal suggesting that our patient may be negative for the Gardner syndrome.

Desmoids may occur in all age groups but they are typically seen in the third and fourth decades of life [3, 6] and have no significant racial or ethnic predilection. The difference in sex distribution is statistically insignificant, but there is a slightly higher incidence of this tumor in women than in men [5].

Symptoms of these fibromatoses depend on the site of the tumor. The signs are insidious and usually manifest when there is a large palpable tumor, or with abdominal discomfort or pain, nausea, vomiting, weight loss, and fever [4, 5, 7]. Desmoids can be locally aggressive and may invade contiguous structures. Some complications that have been reported and may lead to severe morbidity and mortality [8], include small-bowel obstruction and hydronephrosis [9], or ureteric obstruction, intestinal perforation, entero-cutaneous fistula, and intestinal hemorrhage. However, we have not seen any previous case of spontaneous ureteral rupture in the literature, such as seen in the patient presented in this report, leading to septic peritonitis and uroperitoneum and aggressive surgical operations for management and a loss of the right kidney.

On gross histopathologic examination, desmoid tumors are usually circumscribed lesions, but they may have irregular or infiltrating borders. On the cut surface, they are white and coarsely trabeculated, resembling scar tissue. Desmoids are usually larger than 5 cm when they are discovered, and they may be larger than 15 cm. In 10–15% of cases, desmoids are multiple [10].

Histologically, desmoid tumors are lesions composed of bland spindled or stellate fibroblastic cells embedded in a collagenous stroma, without evidence of muscular or neural differentiation and with little or no inflammatory component. The tumor may infiltrate adjacent viscera and tissues at the periphery [5].

The imaging appearance of these tumors is variable and depends on fibroblastic proliferation, fibrosis, collagen content, and vascularity. On sonography, desmoid tumors have variable echogenicity, appearing as masses of low, medium, or high echogenicity with smooth, defined margins [6]. On CT scan, most desmoid tumors appear as well-circumscribed homogeneous masses that may be isodense or hyperdense relative to muscle. Some cases of heterogeneous masses with infiltrative outer margins are seen. Desmoid tumors may enhance after injection of IV contrast material [3], localizes the tumor and excludes metastasis. Malignant rhabdoid tumors (MRT) reveals the tumor’s hypointensity on T1 and demonstrates variable signal intensity on T2 weighted imaging, depending on the accumulation of mucoid structures [6]. Therefore, a differentiation from other solid tumors is impossible using morphological criteria [6].

Figure 3: Histological and immunohistochemical findings of the retroperitoneal and breast desmoid tumor showing (A) Mesenchymatous tumor in contact with the normal tissue, (B) largely dissecting the parenchyma, (C) High magnification showed that the mass consisted of spindle cells, with a regular nucleus, separated by large amounts of collagen fibers in edematous tissue with rare inflammatory cells (H&E stain, x400) (D). All tumor cells were negative with monoclonal anti-CD34 antibody (E). Moderate nuclear staining was detected with beta-catenin antibody (F). All tumor cells were negative for Bcl-2.
A multidisciplinary approach including surgery, chemotherapy, and radiation therapy is required for treatment. Complete resection is the therapy of choice for this type of tumors, but these tumors are often unresectable because of their aggressive and local infiltration, compression of surrounding structures and massive involvement of adjacent vital structures and sometimes a complete resection is only possible through organ transplant. The high recurrence rate even after complete surgical resection favors the use of nonsurgical therapy, such as nonsteroidal anti-inflammatory drugs and antiestrogens [3]. In our case due to extensive fibromatoses of the retroperitoneal space and the mesentery, excision was impossible, but wide excision of the breast lesion with clear margins was made without recurrence.

Distant metastases have not been reported [5], but in our present case, the second location in the breast seen 10 months after the diagnosis of the intraabdominal desmoid tumor But with the same Histological and immunohistochemical features can be a simple concomittent location not be diagnosed at the same time of the other lesions; or sporadic location appeared after the other locations.

CONCLUSION

Spontaneous ureteral rupture as a complication and a circumstance of discovery of the retroperitoneal desmoid tumor is a rare condition. Its diagnosis is difficult and should be considered when presented with symptoms of acute abdominal pain or renal colic. Computed tomography scan remains quite valuable to confirm the diagnosis whereas the treatment is difficult. Desmoid tumors are benign neoplasms but are often unresectable because of their aggressive and local infiltration, compression of surrounding structures and massive involvement of adjacent vital structures; especially in the retroperitoneal space, as seen in our case. To the best of our knowledge, no cases of retroperitoneal and breast desmoid tumor association have previously been reported. Our purpose is to increase awareness of this condition so that clinicians will consider diagnosing it. This will facilitate prompt diagnosis and treatment.

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Self resolving non-parasitic splenic cyst: A case report

Meetal Shah, Abdul Quyyum Khan, Maria Vittoria Cavalletti, Luciano Perrone, Chenji Ratnavel, Giovanni Domenico Tebala

ABSTRACT

Introduction: Splenic cysts are uncommon findings whose treatment is usually surgical if more than five centimeters. Case Report: A case of a self resolving symptomatic large non parasitic splenic cyst is reported. Conclusion: Even if general guidelines prescribe a surgical treatment for >5 cm splenic cyst, sometimes a wait-and-see strategy could be adopted, with the consent of the patient and keeping an eye on his/her general conditions.

Keywords: Spleen, Splenic cyst, Splenectomy, Non-surgical treatment

INTRODUCTION

Splenic cysts are uncommon findings. Their treatment is usually based on their size and probable etiology. Open or laparoscopic splenectomy remains the treatment of choice in particular for large cysts, even if less invasive procedures, such as partial splenectomy, marsupialization or simple drainage, are gaining popularity as they allow maintaining splenic function. Conservative non-surgical treatment has been proposed for cysts whose size is less than 4 cm, whereas larger lesions would require an operative approach to avoid complications such as rupture or infection, as they are supposed to be unlikely to regress spontaneously. Herein, we report a case of a big splenic cyst that resolved without any treatment but observation. Current policies and indications for surgery in case of splenic cyst should be revised.

CASE REPORT

A 42-year-old female (anesthetist by profession) presented with sudden onset of upper left abdominal pain, left loin pain and left lower chest pain, without any history of trauma, even minimal. She did not complain of any urinary or bowel symptom and had never seen blood in her urine or stools. She had no angina-like pain or any breathing problem. Her past medical history was completely unremarkable and she had not had any trip abroad in the last five years.

General conditions were good and vitals were normal. Physical examination revealed an area of tenderness...
at the upper left quadrant of the abdomen, without any guarding or rebound, where a deep mass could be palpated. Urine test and culture were negative. Blood test showed no abnormality, in particular no anemia or eosinophilia. Serologic tests for parasites were negative. Ultrasound scan revealed an 8-cm cystic mass of the spleen but no free fluid. Computed tomography (CT) scan (Figure 1) confirmed the presence of an 8-cm cyst of the medial aspect of the spleen without any other relevant abdominal finding. The cyst was apparently simple and thin-walled, without any internal sept or vegetation. The CT scan was unenhanced as patient declined contrast.

Following the guidelines, we proposed a laparoscopic total or partial splenectomy, depending on the intraoperative findings, to avoid complication as rupture or infection. After an in-depth discussion with the patient, she was adamant that she would like to avoid any surgery, even laparoscopic.

For this reason we agreed to adopt a wait-and-see strategy. She was monitored for 24 hours in the surgical ward, than discharged. Pain subsided gradually in a week or so. Three months later, a repeat CT scan (Figure 2) revealed that the cyst had completely disappeared. Now, more than 2 years after that episode of pain, the patient is completely fine and asymptomatic. A recent ultrasound scan confirmed the absence of any splenic cyst.

**DISCUSSION**

Splenic cysts are generally uncommon findings which tend to present with a variety of nonspecific symptoms such as abdominal fullness, generalized pain, shortness of breath and back pain [1–3]. Incidence in Europe recently went up to 1% due to the widespread use of ultrasound [4], but due to the nature of symptoms, a great deal of splenic cysts are chance findings on ultrasound, abdominal CT or even on autopsy. A study carried out by Robbins et al. and published in 1978, revealed 32 benign splenic cysts found incidentally at autopsy in a total of 42,327 patients’ autopsy records [5].

While hepatic cysts are mostly congenital or parasitic and rarely malignant or infective and renal cysts are mostly congenital or occasionally related to systemic diseases, splenic cysts are categorized into two groups based on the absence or presence of an epithelial wall. Primary cysts, also known as ‘true cysts’, have a complete epithelial wall and secondary cysts, or ‘false cysts’, have an incomplete or no epithelial lining [1]. True cysts are further divided into parasitic or non-parasitic cysts. Parasitic cysts are rare and usually caused by the Echinococcal infection [6] while non-parasitic cysts can then categorized into congenital or neoplastic cysts. Non-parasitic cysts can be true or false. Congenital cysts can be either epidermoid, dermoid or endodermoid cysts, with endodermoid cysts having the highest prevalence of all nonparasitic true cysts at 90% [7]. Neoplastic cysts can be hemangiomas or lymphangiomomas. These tend to be multilocular and so, can be diagnosed easily. False cysts comprise about 50–80% of non-parasitic cysts [8] and tend to be post-traumatic as a result of blunt injury to the abdomen, or in some rare cases, are caused by mononucleosis, tuberculosis or malaria [9].

Even if they are usually asymptomatic and discovered during routine abdominal imaging, they can present with left upper quadrant pain, which can have a sudden onset as in the reported case. More often, when symptomatic, they can give vague abdominal pain due to capsular distension or pressure on the surrounding structures [10]. Ultrasound scan and CT scan are the procedures of choice to investigate the cyst and the bases of the choice of the treatment. Blood tests can be useful in discriminating a parasitic etiology.

The management of splenic cysts depends on the type of pathogenesis involved, the size and any possible
complications, and while there is still controversy surrounding the optimal management of these cysts, surgery is evidently the first port of call. Research recommends some form of surgical intervention for cysts which are symptomatic or complicated, or greater than 4–6 cm in diameter [11, 12]. This is especially true for post-traumatic cysts, which should be managed when the diameter exceeds 4 cm due to the greater risk of rupture and infection such as hemoperitoneum, abscess formation and chemical peritonitis [9, 13]. Both open and laparoscopic techniques can be used, however, preference is given to laparoscopic methods as they are minimally invasive, with shorter hospitalization time and lower rates of post operative pain [2].

There are a variety of surgical methods involved in splenic cyst treatment and factors influencing the surgical procedure selected are: patient age, size and location of cyst, and type of cyst [3]. These include percutaneous drainage, total splenectomy, partial splenectomy, marsupialization and fenestration [9]. Percutaneous drainage is now considered an effective way of reducing the size of large cysts pre-operatively before a more effective surgical method is used to prevent recurrence [14]. Total and partial splenectomy can both be performed laparoscopically with partial splenectomy being preferred as splenic parenchyma is preserved enabling regular immunological function of the spleen if enough is saved [9, 15]. Marsupialization is a method of separating the splenic tissue from the cyst [9] and due to the low recurrence rate experienced, it is recommended in the management of superficial cysts [16]. Finally, fenestration methods involve resecting only a segment of the cyst wall. This allows for communication between the peritoneal cavity and cyst cavity but has been reported to have a high recurrence rate, irrespective of whether laparoscopic or open methods are used [13]. It is important to acknowledge that every operative approach has a potential for complications, such as pancreatic injury with pancreatitis or fistula, stomach or bowel injury, diaphragmatic injury with or without pneumothorax and loss of splenic function with the risk of overwhelming post-splenectomy sepsis [17].

This case report, however, illustrates the possibility of spontaneous regression of cysts with no surgical intervention being necessary. While a conservative approach was an unusual strategy to adopt, with a multitude of surgical options being available, it was carried out at the request of the patient. A similar case was reported in 2009 involving a female patient who initially presented with a 6-cm true cyst, which regressed to a 1.8x1.4 cm cystic lesion in nine years [1]. While, in general, the first choice of management in non-parasitic splenic cysts should be laparoscopic surgery which preserves spleen parenchyma, it is important to consider the possibility of spontaneous cyst regression [18]. This can occur following parenchymal rearrangement and tissue scarring enabled by the absence of a true epithelial lining. The patient should be informed and given the two possible options, surgery or wait-and-see, in particular in the absence of hemodynamic abnormalities and when the CT scan shows a clear diagnosis of non-parasitic cyst and excludes a malignant etiology. In this regard, an enhanced CT scan is the best diagnostic tool, should the patient agree to receive intravenous contrast.

CONCLUSION

Self resolution of splenic cysts is not a heavily documented phenomenon as most cases are resolved early on with surgery, but should be considered especially in uncomplicated cysts. Regular follow-up and computed tomography scans would enable cyst progression to be tracked and if patients remain asymptomatic, it would eliminate the need for surgery and all associated risks.

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Familial myelinated nerve fibres with proliferative vitreoretinopathy

Mehra A, Safia H Ahmed, Carpi Akikio Olali, Ahmed S, Gupta M

ABSTRACT

Introduction: To report a rare case of familial myelinated nerve fibres with vitreoretinal complications. Case Report: A three and half-year-old boy with reduced vision (right 6/60, left 6/12) was found to have extensive bilateral myelinated nerve fibres with similar lesion in the parent. The child later developed vitreous hemorrhage in the left eye and examination confirmed localized proliferative vitreoretinopathy. Conclusion: This is probably the first reported case of familial myelinated nerve fibres with proliferative vitreoretinopathy, vitreous hemorrhage and bilateral amblyopia. Our case and the few other reported familial ones suggest a dual pattern of occurrence of the myelinated retinal fibres; sporadic and genetic.

Keywords: Myelinated fibres, vitreoretinopathy, Familial, Amblyopia, Vitreous hemorrhage

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INTRODUCTION

Myelinated retinal nerve fibres which occur in 1% of the population are often localized in a segment of the retina and have no effect on vision [1]. We report a rare case of familial myelinated nerve fibres with vitreoretinal complications.

CASE REPORT

A three and half-year-old boy was referred because of reduced vision noticed during routine pre-school screening. He was otherwise well, but his uncle had tuberous sclerosis and grandmother retinitis pigmentosa. His unaided visual acuity was 6/60 in the right eye and 6/12 in the left and fundus examination revealed extensive myelinated nerve fibres along the superior and inferior arcades and involving the optic disc in the right eye and almost total myelination of the nerve fibres on the inferior half of the left eye (Figure 1A–B). The anterior segment structures were normal in both eyes. He was refracted and the following prescription given; Right eye -3.0/+1.50 @ 90° and left eye -1.0/+2.75 @ 80°. An initial subnormal electroretinogram was found to be normal a year later and he has had no problems seeing at night or tuberous sclerosis features. However, the mother was also examined and found to have myelinated nerve fibres between 1.30 and 4 o’clock in right eye (Figure 1C).

The patient was kept on annual review but in December 2006, he complained of seeing white spots in his eyes. His corrected visual acuity was 6/24 in the right eye and 6/12 in the left and dilated fundoscopy revealed extensive vitreous condensation with bands and localized traction in the left eye. There was, however, no retinal tear. In July 2010, he developed vitreous hemorrhage in that left eye and examination confirmed localized proliferative
vitreoretinopathy in the inferior-nasal quadrant (Figure 1D). No active intervention has been initiated as there has since been slow regression of the abnormal vessels and clearing of the hemorrhage.

DISCUSSION

Myelinated nerve fibres usually seen as incidental finding in routine clinics are benign and stationary in most cases though some are associated with complications such as telangiectasia, capillary congestion, neovascularization and recurrent vitreous hemorrhage [2–4]. Bilateral involvement was documented in 7.7% of autopsy cases and common associations include asymmetric myopia, amblyopia, Von Recklinghausen disease and Gorlin’s syndrome and craniosynostosis [5–7].

There are very few reported cases of familial myelinated retinal nerve fibres in literature [8–9]. In our patient, the mother similarly had bilateral involvement though less extensive. The patient presented with reduced vision and the lesions later complicated in the form of proliferative vitreoretinopathy.

The mechanism of retinal ischemia and secondary vascular changes is not known. Minning and Davidorf [4] suggested that the myelinated nerve fibres may induce mechanical disruption of retinal vasculature. Myelinated nerve fibres on account of increasing diameter and metabolic activity compete for available oxygen in tissue causing relative ischemia. When the inner retinal ischemia occurs, the mechanism of neovascular activity is the same as in the other retinal vascular diseases such as retinal vein occlusion and diabetic retinopathy, possibly due to the release of angiogenic factors like vascular endothelial growth factors (VEGF). Generally, the vitreous hemorrhage is self-limiting. Rubeosis was not a reported complication. Although experience with laser treatment is limited, it does help in regression of neovascularization.

Aberrant myelinated retinal nerve fibres are thought to form because of failure to prevent oligodendrocyte lineage cells from passing through the lamina cribrosa or optic nerve head. Astrocytes in the lamina cribrosa have been postulated as specialized, acting as a barrier through the orientation and number of their glial filaments [10]. However, it is not known how inheritable defect could affect this process.

This is probably the first reported case of familial myelinated nerve fibres with proliferative vitreoretinopathy, vitreous haemorrhage and bilateral amblyopia.

CONCLUSION

Our case and a few other reported familial ones suggest a dual pattern of occurrence of the myelinated retinal fibres; sporadic and genetic. There is thus the need for further study to elucidate the inheritance pattern if any in the familial form.

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ABSTRACT

Introduction: As laparoscopic cholecystectomy is one of the most common procedures done worldwide. Although anomalies are rare but are associated with congenital malformations of gallbladder, bile ducts and vascular system. Case Report: Herein, we present a case of young female presenting with symptoms of pain in right hypochondrium with ultrasonographic diagnosis of cholelithiasis undergone successful laparoscopic cholecystectomy with intraoperative findings of: 1. The length of the gallbladder was measured to be 25.8 cm. 2. The fundus of the gallbladder was placed to the left of the falciform ligament. The gallbladder then extended to the right of the falciform ligament, reached up to the liver margin before taking a ‘U’-turn to lie in the usual gallbladder fossa. Conclusion: Anomalies of gallbladder present an important hurdle in successful laparoscopic cholecystectomy. Most of time not known preoperatively encountered during surgery. Isolated left-sided gallbladders are rare and found in 0.04–0.3% of cases. When there is question about anatomy of biliary tract intraoperatively one should consider for anomalies. A habit of calm and slow dissection with precautions should be developed. Clearance of the anatomical structures with limited use of electrocautery should be done before proceeding towards ligation or clip applications to structures. A surgeon should be well equipped with knowledge of anomalies of gallbladder and meticulous dissection with good exposure of structures should be done when an anomaly found.

Keywords: Left-sided gallbladder, Longest gallbladder, Laparoscopic cholecystectomy, Anomaly gallbladder

INTRODUCTION

As laparoscopic cholecystectomy is one of the most common procedures done worldwide. Although anomalies are rare but are associated with congenital malformations of gallbladder, bile ducts and vascular system. Knowledge of anomalies before going for laparoscopic cholecystectomy is essential for safe and successful surgery. These can be dealt with meticulous dissection and appropriate identification of structures before applying clips and cutting structures.
CASE REPORT

We present a case of a young female presenting with symptoms of pain in right hypochondrium with ultrasonographic diagnosis of cholelithiasis. Laparoscopic cholecystectomy was planned. After creating pneumoperitoneum standard four ports were placed. On inspection of gallbladder findings noted were:

1. The fundus of the gallbladder was placed to the left of the falciform ligament. The gallbladder then extended to the right of the falciform ligament, reached up to the liver margin before taking a ‘U’ turn to lie in the usual gallbladder fossa. The infundibulum and neck of the gallbladder were placed in the location of the normal gallbladder. The cystic duct entered the common hepatic duct from the right side, forming the Calot’s triangle. The surgery was completed laparoscopically in four hours, using the fundus-first approach (Figure 1). The gallbladder was retrieved from the epigastric port (Figures 2 and 3).

2. The length of the gallbladder was measured to be 25.8 cm. The measurement was made in the operation theatre, using a transparent straight ruler with centimeter markings on one side and inch markings on the other side. Microscopy showed normal histology of the gallbladder (Figure 4).

Patient undergone uneventful laparoscopic cholecystectomy and was allowed oral intake in evening and discharged on next day. Patient followed for 30 days with no significant complaints.

DISCUSSION

Anomalies of gallbladder presents an important hurdle in successful laparoscopic cholecystectomy. Most of time not known preoperatively encountered during surgery. A prenatal study done by Bronshtein et al. 1993 on 10,016 fetal examinations after the 14th week of gestation reported 17 cases of anomalous gallbladder a 0.15% incidence of gallbladder malformations [1]. As per study of 500 subjects by Carbajo et al. Congenital gallbladder malformations were diagnosed in 1% of the cases, all cases were intraoperatively diagnosed and only two patient have to be converted to open cholecystectomy [2]. Isolated left-sided gallbladders are rare and found in 0.04%-0.3% of cases [3]. Two possible embryological etiologies for left-sided gallbladder are suggested.

1. Gallbladder is attached to the left lobe and migrates in front of common duct to come in left sidein which case the cystic duct is in a normal anatomic position.

2. Gallbladder is entirely formed from bud from the left side in which case the cystic duct joins the CBD or left hepatic duct from the left side [4, 5].

As in our case gallbladder was of 25.8 cm which is largest as certified by Guinness book of world record with association of anomalous location of fundus of gallbladder to left side. Dr Naeem Taj operated a case at CDA hospital.
Islamabad, Pakistan, Rasheeda Bibi, 70-year-old female with a 25.5 cm long gallbladder.

When there is a question about anatomy of biliary tract intraoperatively one should consider for anomalies. There are four types of aberrant gallbladder: (1) intrahepatic, (2) left-sided, (3) transverse, and (4) retrodisplaced illustrated. The aberrant gallbladder produced false positive liver scans which were correctly diagnosed by hepatic angiography [6]. A habit of calm and slow dissection with precautions should be developed. Clearance of the anatomical structures with limited use of electrocautery should be done before proceeding towards ligation or clip applications to structures. An intraoperative cholangiography can be used to further delineate details of anatomy [7]. If surgeons’ experience allow, one can proceed with laparoscopic surgery. Fundus first approach can make access easy in tricky situations. If surgeon is not experienced enough, conversion to open procedure should be done.

CONCLUSION

A surgeon should be well equipped with knowledge of anomalies of gallbladder and meticulous dissection with good exposure of structures should be done when an anomaly found. The importance of identifying the gallbladder at hepatic angiography by observing the cystic arteries in arterial phase and the gallbladder wall stain in hepatogram phase is stressed.

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REFERENCES


Primary aggressive non-Hodgkin lymphoma of the parotid gland in a young individual: A case report

Devika Gupta, GPS Gahlot, Vandana Rana, Rajat Jagani, Davendra Swarup

ABSTRACT

Introduction: Extranodal non-Hodgkin lymphomas constitute 25–40% of all lymphomas. The most common site is gastrointestinal tract followed by head and neck area. Salivary gland is involved in 2–5% of all cases. Case Report: We herein report a case of a 24-year-old male who presented with painless, gradually increasing mass in the region of left parotid. Imaging studies, both ultrasonography and magnetic resonance imaging (MRI), were suggestive of infiltrative mass lesion involving the left parotid gland along with evidence of suspected bony metastasis. Patient underwent total parotidectomy with excision of level II&III cervical lymph nodes. Histopathological examination supported by immunohistochemistry helped us to clinch the diagnosis of primary diffuse large B cell lymphoma of the parotid gland. Conclusion:

Non-Hodgkin lymphoma can mimic a large number of both benign and malignant disease entities of the salivary gland. Hence a high index of suspicion is required for an early diagnosis of non-Hodgkin lymphoma in a rapidly growing mass of salivary gland.

Keywords: Non-Hodgkin lymphoma, Salivary glands, Parotid gland, Diffuse large B cell (DLBCL)

INTRODUCTION

Non-Hodgkin lymphoma (NHL) is a heterogenous disease of lymphoid/extra lymphoid tissues presenting clinically in indolent as well as aggressive forms. Lymphoma constitute 2–5% of all salivary gland neoplasms with the parotid gland being involved most frequently followed by submandibular and then minor salivary glands [1]. These neoplasms may arise from an intraparotid lymph node or in the gland itself. Most primary salivary gland lymphomas are of B cell lineage in which the MALT (Mucosa-associated lymphoid tissue) lymphomas are most common. These arise in a background of benign lymphoepithelial lesions and have known to have an association with Sjögren’s disease. Other NHLs like Diffuse Large B Cell (DLBCL) and follicular lymphomas are less commonly reported. Our case is primary DLBCL arising in the parotid which was diagnosed on histology following total parotidectomy.
A 24-year-old male, non-smoker with no known co-morbidities presented with rapidly progressive, painless swelling of left parotid region which gradually increased in size from about 2x1 cm to 5x4 cm over a short span of four months. There was no history suggestive of fever or any connective tissue disorder. There was no other significant personal/family history. General physical examination revealed a well-nourished individual with no lymphadenopathy. On local examination there was a firm, nontender diffuse swelling measuring approximately 5x4 cm in the left parotid region associated with puckering and discoloration of overlying skin. There were no overlying dilated veins, visible pulsation, sinus fistula seen. Oral examination and laryngoscopy was normal. Systemic examination of the respiratory, cardiac and central nervous systems were within normal limits. All his routine hematological and biochemical parameters were within normal limits except for serum lactate dehydrogenase (LDH) which was raised to 522 IU/L. Fine-needle aspiration cytology (FNAC) attempted twice from the lesion was inconclusive. Ultrasonography of neck revealed a lesion heterogenous in echotexture involving the left parotid gland with multiple enlarged level II, III cervical lymph nodes, largest measuring 1.3x1.7x2 cm at level II (left). The lymph node in left level II appeared necrotic. Magnetic resonance imaging (MRI) scan showed irregularly contoured lesion in left parotid gland involving both the superficial lobe and tail region with extension in to the deep lobe and measuring approximately 32.6x20.4x22 mm (CCxAPxTR). There was focus of T1 and T2 hyperintensity within this lesion in the tail suggestive of hemorrhage. No cystic components were noted within this lesion. Multiple left necrotic lymph nodes were noted in the level I, II, and level III.

Patient was taken up for left extended parotidectomy under general anesthesia. Peroperative findings showed a growth involving anterior one-third upper border of sternocleidomastoid, level II and part of level III lymph nodes. A firm mass measuring 3.5x3x4 cm was also present in deep lobe below cervical and zygomatic temporal divisions of facial trunk in a dumb bell fashion. External carotid artery, internal jugular vein and facial nerve were preserved. Excision of the superficial lobe, deep lobe, part of sternocleidomastoid muscle, level IIb and level III lymph nodes with skin over parotid was done. The procedure was uneventful without any evidence of facial nerve damage. Histopathology of the mass revealed atypical lymphoid cells involving both the superficial and deep parotid lobes. These cells were seen infiltrating through the parenchyma of both lobes and infiltrating the sternocleidomastoid muscle. The cells were large, oval to round had vesicular, hyperchromatic nuclei with conspicuous nucleoli. Mitosis was brisk. Areas of necrosis with numerous scattered apoptotic bodies were seen. No lymphoepithelial lesions were identified. The left juxtraglandular lymph nodes, level III and level IIb lymph nodes showed involvement by similar malignant cells. Immunohistochemistry (IHC) study was performed and the tumor cells were strongly positive for LCA, CD20, CD79a and negative for CD3, CD5, CK, EMA, Tdt. The Ki-67 labeling index was 100%.

Based on histomorphology and IHC a diagnosis of Diffuse large B cell lymphoma of the left parotid was made. After this diagnosis patient underwent systemic imaging studies which showed multiple, ill-defined lytic lesions in the body of C7, DV4-DV6, DV10, LV2, LV4, SV1 and bilateral iliac bones. Bone marrow study was unremarkable. A clinical stage IV disease was established by oncologist and patient started on RCHOP.
chemotherapy. As part of RCHOP therapy patient received eight cycles of injection rituximab (375 mg/m²), injection cyclophosphamide (750 mg/m²), injection vincristine (1.4 mg/m²), injection Adriamycin (50 mg/m²) and Tablet Prednisolone 100 mg per day for five days. Eight cycles of chemotherapy were given each at 21 days intervals. He responded well and six months after treatment he is on follow-up at our hospital.

DISCUSSION

Primary lymphomas of the salivary glands are rare and account for 2–5% of all salivary gland neoplasms. Parotid is most commonly involved in 50–90% cases followed by submandibular gland. In the largest study of 40 salivary gland lymphomas, only three cases were seen arising from submandibular gland [2]. Primary parotid lymphomas account for 0.87% of all NHL cases and 4–5% of all extranodal NHLs [3]. Malignant lymphomas of the parotid are uncommon in patients younger than 50 years with peak age at 55 years [4]. Our patient is young individual of 24 years of age. The lymphomas may arise from intraparotid lymph nodes or in the gland itself. If only the intraglandular lymph nodes are involved with no parenchymal infiltration then it should be considered of nodal origin. The distinction is usually difficult as there is extensive, diffuse parenchymal involvement of intraparotid lymph nodes. So primary salivary gland lymphoma is considered when parenchyma of the gland is involved [5]. Most NHLs arising in the salivary glands are B cell lineage including low grade B cell lymphoma of MALT, follicular lymphoma and DLBCL [6]. The DLBCL is a high grade infiltrative tumor associated with destruction of salivary gland parenchyma with tumor cells invading between residual gland acini. On histology the cells are large and atypical and resemble either centroblast or immunoblast as was seen in our case. On the other hand, low grade MALT lymphomas of parotid gland usually arise in setting of benign lymphoepithelial lesion [7].

Most of the lymphoma cases present with painless, firm swelling in the region of salivary gland mimicking other salivary gland epithelial neoplasms. Hence the patient are subjected to imaging studies and other investigations like FNAC which are not very helpful and cause delay in diagnosis. Most of the lymphomas of the salivary gland are surgically treated because they lack definitive imaging features as happened in our case. Histological examination shows infiltration of monoclonal B cells into the ductal epithelium cells and destroying them. Association between MALT lymphoma and autoimmune diseases like SLE and Sjögren is known [8].

Once the diagnosis of NHL is established it is important to evaluate the patient for any other synchronous systemic involvement or dissemination to decide on the therapy. An Ann Arbor or International Prognostic Index (IPI) scoring system is used to stage the disease. Ours is an aggressive DLBCL of the parotid with metastasis to the vertebrae (stage IV). Treatment depends on clinical staging and irradiation is considered as treatment of choice in localized lesions in early stage. However, it is both or either radiotherapy or chemotherapy which is considered along with surgery in disseminated forms [9, 10]. Our patient underwent RCHOP chemotherapy following total parotidectomy and is on follow-up.

CONCLUSION

Parotid gland swelling is a common presentation in clinical practice and lymphomas affecting parotid gland is clinically indistinguishable from other benign or malignant lesions. Imaging modalities and fine-needle aspiration cytology is not always helpful and hence majority of patients require a parotidectomy for definitive diagnosis.

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Gastric outlet obstruction due to a giant antral polyp which have malignant transformation

Erkan Oymaci, Ali Coskun, Deniz Ucar, Erdem Sari, Nazif Erkan, Mehmet Yildirim, Hale Kizanoğlu

ABSTRACT

Introduction: Gastric polyps are seen 2–3% in all gastrointestinal endoscopic examinations. Hyperplastic polyps, the most common type of gastric polyps, are present 85–90% of cases. Giant villous tumors of the stomach are somewhat rare and the high incidence of malignant transformation of them requires prompt diagnosis. Giant gastric polyps can also cause gastrointestinal bleeding that leads to iron deficiency anemia or partial gastric outlet obstruction. All giant polyps must be resected endoscopically or surgically and still there are no marked guidelines for the optimal management of gastric polyps at the time of initial examination. The point to be taken into consideration for patients is the need for radical surgery or not.

Case Report: A 62-year-old male was admitted to our clinic with complaints of epigastric pain, nausea and vomiting. Upper gastrointestinal endoscopy revealed that 6x7 cm diameter pedunculated polypoid mass which obstruct the pyloric channel in the gastric antrum. The patient is treated by distal subtotal gastrectomy and gastroenterostomy. The histopathologic examination of the specimen confirmed the occurrence of adenocarcinoma.

Conclusion: Giant hyperplastic gastric polyps are fairly uncommon. We presented a case of giant gastric polyp which have malignant transformation and cause gastric outlet obstruction that resected surgically in an attempt to add to the current literature.

Keywords: Gastric polyp, Villous adenoma, Obstruction, Surgery

INTRODUCTION

Gastric polyps are seen 2–3% in all gastrointestinal endoscopic examinations. Hyperplastic polyps, the most common type of gastric polyps, are present 85–90% of cases [1]. Hyperplastic polyps, fundic gland polyps, gastric adenomas and some of gastric carcinoid tumors may present as a gastric polyp. Villous adenomas of the stomach are somewhat rare with approximately 100 cases only reported in literature and have tendency to undergo malignant transformation as high as 72% [2]. They are frequently multiple and associated with other gastrointestinal neoplasms. The high incidence of malignant transformation of gastric villous adenoma requires prompt diagnosis of this rare tumor. Polyps greater than 2 cm are at significant risk for malignancy and they require complete resection [3]. Except fundic gland polyps which have a clear typical feature, upper endoscopy cannot reliably distinguish the type of gastric polyp by gross inspection. Thus, histopathological...
diagnosis is important although whether to biopsy or excise gastric polyps is not always clear [4]. Giant villous tumors of the stomach are somewhat rare and the high incidence of malignant transformation of them requires prompt diagnosis. Giant gastric polyps can also cause gastrointestinal bleeding that lead to iron deficiency anemia or partial gastric outlet obstruction. Gastric outlet obstruction presents with nausea and vomiting and usually develops over weeks to months. It may be complete or incomplete with intermittent symptoms. Gastric villous adenomas have potential for malignant transformation and hence must be excised endoscopically or surgically whichever may be feasible. We presented a case of giant gastric villous adenoma which have malignant transformation and cause gastric outlet obstruction in an attempt to add to the current literature.

CASE REPORT

A 62-year-old male was admitted to our clinic with complaints of epigastric pain, nausea and vomiting. He described two pain and vomiting episodes of similar type, each lasting three or four days, in last two months. His medical background and family history were unremarkable. Physical examination revealed that mild epigastric tenderness and normal bowel sounds. At the admission, laboratory tests revealed hemoglobin of 9.5 g/dL with a mean corpuscular volume of 68.9 fl. Serum iron was 30 μg/dL and total iron-binding capacity was 408 μg/dL. The other laboratory results were also within normal limits. Upper gastrointestinal endoscopy revealed a 6x7 cm diameter pedunculated polyp covered with irregular villous mucosa arising from the antrum of the stomach and which totally obstructed the entrance of the pyloric channel (Figures 1 and 2). This polyp was seen protruding into the bulb of the duodenum causing gastric outlet obstruction. Multiple biopsy were taken from both the polypoid mass and antrum. Histopathologic examination of the biopsy revealed a gastric villous tumor with carcinomatous change. Microscopic findings of the biopsy showed malignant glandular formation invading the lamina propria on the stomach. It also showed finger-like projections covered by dysplastic epithelium (Figures 3 and 4). The patient underwent a distal subtotal gastrectomy with Billroth II gastrojejunostomy reconstruction and lymph node dissection on the third day of admission (Figures 5 and 6). No postoperative complications occurred and the patient is discharged on the postoperative sixth day. Evaluation of the specimen showed that the polyp has the largest diameter of 7 cm. The histopathologic examination of the specimen confirmed the presence of villous adenoma with the occurrence of adenocarcinoma with no morphological and histochemical evidence of Helicobacter pylori infection. There was no lymph node metastasis.
Most of gastric polyps have asymptomatic presentations and are incidentally finding with an incidence of approximately 6% on upper endoscopic examination [5]. Their pathogenesis is unknown but the majority of polyps occur as multiple lesions which arising from inflamed gastric mucosa. Infection of the gastric mucosa with Helicobacter pylori has been reported in up to 90% of the cases [6]. Giant gastric polyps are fairly uncommon. Patients with hyperplastic polyps, greater than 3 cm in largest diameter are more likely to be symptomatic. These giant hyperplastic polyps represent about 2% of all hyperplastic polyps [7]. Gastric polyps can vary in size from a few millimeters to several centimeters. The median size of polyps removed endoscopically was 3 cm while the median size of surgically removed polyps was 6 cm [8, 9]. Kumar et al. have reported the largest endoscopically treated polyp causing intermittent gastric outlet obstruction to date [9]. They removed an 8-cm polyp; two-thirds of the polyp was snared and the remainder excised at a subsequent visit. Aksel et al. reported that the size of the gastric polyp was 12 cm diameter in their study and made radical gastrectomy for the gastric polyp which is in giant diameter and causes severe bleeding [7]. In our case, we also made radical gastrectomy due to malignant transformation which include high grade dysplasia and giant 6x7 cm diameters of polyp. Symptomatic presentations can range from an ulcerated polyp leading to anemia to complete gastric outlet obstruction. Al-Haddad et al. reported that the incidence of hyperplastic polyps in iron-deficiency patients was 1.4% and the largest polyp was 5 cm in diameter [10]. Anemia is the most frequent clinical manifestation of diffuse gastric polyposis. Our patient has both gastric outlet obstruction and iron-deficiency anemia in laboratory tests at admission.

Villous tumors of the gastrointestinal tract are neoplasms that arising from the columnar epithelium because such neoplasms occur infrequently in the upper gastrointestinal tract, the clinical, radiologic, and pathologic features have not been completely defined. Inaba et al. reported that the villous adenoma accompanied by malignant changes was positive by the carcinoembryonic antigen peroxidase-antiperoxidase method [11]. This result showed the biological property of villous adenoma that they can easily change into malignancy. Adenoma of the stomach and duodenum are subject to malignancy more often than colonic adenomas. It might be possible to encounter with dysplasia and carcinoma around the polyp. There is a synchronous adenocarcinoma risk in another part of the stomach after polypectomy in up to 30% of cases [12]. So, it is necessary to examine surrounding tissue and to take multiple biopsies from the gastric mucosa with endoscopic intermittent follow-up. It is quite important because of the possibility of recurrence at the polypectomy site and development of malignancy in the remote gastric mucosa after polypectomy [13].
CONCLUSION

As conclusion, we presented a case of giant gastric villous polyp which have malignant transformation and cause gastric outlet obstruction that resected surgically in an attempt to add to the current literature. The point to be taken into consideration for patients is the need for radical surgery or not. The main disadvantage of endoscopic methods is the risk of incomplete tumor resection. So, it seems to be there is no alternative treatment other than radical gastrectomy for the gastric giant polyp which causing obstruction.

**********

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REFERENCES

CASE REPORT

A rare cause of upper gastrointestinal bleeding: Posttraumatic pseudoaneurysm

Negi RC, Brij Sharma, Bhupender, Gaurav Kapoor, Bal Beer Verma, Ashok Sharma

ABSTRACT

Introduction: Hemobilia is a rare cause of upper gastrointestinal bleeding. It needs prompt diagnosis and immediate management to save the life. Posttraumatic pseudoaneurysm of hepatic artery is seen mostly after abdominal trauma causing liver injury or interventions on hepatobiliary system. On esophagogastroduodenoscopy active bleeding without apparent source is clue to the diagnosis.

Case Report: A 27-year-old male presented with history of one episode of hematemesis and melena. There was history of roadside accident one and a half months before and he had liver injury which was revealed on computed tomography of the abdomen. Patient was managed conservatively and discharged. On examination, patients was anemic and had postural fall in blood pressure. Emergency esophagogastroduodenoscopy revealed blood coming from ampulla. Computed tomography angiography (CTA) revealed pseudoaneurysm in segmental branches of right hepatic artery with active bleeding. Conclusion: Hemobilia is defined as bleeding into the biliary tree from abnormal communication between blood vessel and bile duct. The most common cause of posttraumatic hemobilia is pseudoaneurysm. The classic triad of hemobilia is absent in 70% of cases and in such cases clinical diagnosis is difficult. The CTA is the investigation of choice and embolization is the treatment option. Surgery should be done in cases refractory to embolization.

Keywords: Hemobilia, pseudoaneurysm, embolization, Gastrointestinal bleeding.

INTRODUCTION

Hemobilia is a rare cause of gastrointestinal (GI) bleeding which develops as a result of communication between blood vessel and biliary tract [1]. Posttraumatic pseudoaneurysm of hepatic artery is seen in 1% of hepatic injury [2]. It should be considered in patients presenting with upper gastrointestinal bleeding with prior history of abdominal trauma. The patients commonly present with either hematemesis or melena but may also present with abdominal pain or jaundice. High index of suspicion is required for the diagnosis of hemobilia.

CASE REPORT

A 27-year-old male presented with complaints of one episode of hematemesis seven days back and
Melena for seven days. There was a history of roadside accident one and a half months before with grade IV liver injury, revealed on computed tomography (CT) scan of the abdomen. Patient was managed conservatively in Department of Surgery and discharged from hospital after one week in stable condition. On examination, the patient was anemic, pulse 110/min, blood pressure 80/50 mmHg with postural fall in blood pressure. The systemic examination was normal. Clinical possibility of upper GI bleed was kept. Emergency investigation revealed hemoglobin 4.5 g/dL, TLC 4900/mm³, BU 30 mg%, S. cr.-1.0 mg%, and electrolytes Na-135 mEq /L, K-4.1 mEq/L, Cl-105. The nasogastric tube drainage was persistently revealed altered blood. The patient was stabilized with three units of blood transfusion and emergency esophagogastroduodenoscopy was done. The esophagogastroduodenoscopy revealed altered blood in the stomach with large clot in the fundus but source was not evident, first and second part of duodenum was also normal, but blood was present. On visualizing the ampulla, there was evidence of blood coming from ampulla. Emergency computed tomography angiography revealed pseudoaneurysm in segment VIII of liver involving segmental branches of right hepatic artery with active bleeding with communication with biliary radicals, with blood in gallbladder, common bile duct and second part of duodenum with subcapsular hematoma in segment VIII (Figure 1). The digital selective arteriography revealed pseudoaneurysm from segmental branches of right hepatic artery (Figure 2). Embolization was done and post embolization digital selective arteriography revealed complete non opacification of pseudoaneurysm (Figure 3).

DISCUSSION

Hemobilia is defined as bleeding into the biliary tree from abnormal communication between blood vessel and bile duct [1]. Sandworm and Mirkovitch described hepatic artery pseudoaneurysm formation after blunt trauma or penetrating trauma to liver, after percutaneous diagnostic or therapeutic procedures on hepatobiliary system [2]. The mean time period between traumatic hepatic injury and presentation of patients with hemobilia is reported to be four weeks [3]. The most common cause of posttraumatic hemobilia is a pseudoaneurysm. The classic hemobilia triad described by Quinke in 1871 consists of upper gastrointestinal bleeding (hematemesis 60% or melena 90%) and biliary colic (70%) and obstructive jaundice in 70% of cases [4]. The classic triad is absent in almost 70% of cases and in such cases clinical diagnosis is difficult to suspect [5]. The hemobilia may be minor or major, in minor hemobilia minor bleeding stops spontaneously whereas in hemobilia major the
blood flows rapidly into the duodenum presenting as melena or hematemesis. Earlier the most common cause of hemobilia was accidental trauma but nowadays the common cause is iatrogenic injury to liver due to liver biopsy or percutaneous interventions [6]. In a study done by Srivastava et al. from India the predominant cause of hemobilia was liver injury following roadside accidents [7]. The hemobilia should be suspected in patients with prior history of abdominal trauma presenting with upper gastrointestinal bleeding. The presenting case also had a history of abdominal trauma which was managed conservatively. The patient presented with hematemesis and melena after four weeks of abdominal trauma.

In such patients esophagogastroduodenoscopy will reveal altered blood and clot in stomach and duodenum but without apparent source of active bleeding. The blood may be seen in ampulla. The gold standard diagnostic investigation is angiography to localize the site of bleeding but in the present era CT scan and magnatic resonance imaging (MRI) scan are also used for diagnostic purpose as non-invasive technique [7, 8]. The selective arterial embolization of pseudoaneurysm is the procedure of choice for management of hemobilia. Surgery is required in case refractory to embolization [9]

CONCLUSION

Hemobilia is a rare cause of upper gastrointestinal bleeding due to posttraumatic pseudoaneurysm and it should be diagnosed promptly. High index of suspicion is needed for diagnosis. Angiography is the choice of investigation for localization of bleed and embolization is the treatment of choice.

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Neglected telangiectatic osteosarcoma of the femur presenting as surgical emergency

Youssef Mahdi, Lamiaa Rouas, Abdelouahed Amrani, Abderrahmane Malihy, Najat Lamalmi, Zaitouna Alhamany

ABSTRACT

Introduction: Telangiectatic osteosarcoma is a rare and aggressive subtype of intramedullary osteosarcoma. The distal femur is the most common site. Case Report: We present a case of telangiectatic osteosarcoma of the femur in a 15-year-old male. The tumor was neglected for six months. The patient had severe anemia with hemoglobin of 5 g/dL. Radiography showed an expansive lytic lesion of the distal end of the femur with cortical destruction, focal periosteal reaction and a pathologic fracture. It showed massive extension to adjacent soft tissue. Given the advanced stage of the disease, an amputation of the limb was performed. Gross examination of the resected distal femur revealed a 22x5 cm intramedullary multicystic hemorrhagic and destructive tumor with cortical destruction and extension into the adjacent soft tissues. Histologically, the tumor showed blood-filled spaces lined by giant cells, separated by septa with malignant tumor cells. An immature bone osteoid kind was observed. Numerous mitotic figures were noted. The resection margins were negative. The staging showed no metastasis. The decision was to complete by 6 cycles of chemotherapy. The postoperative course was unremarkable. Conclusion: In terms of telangiectatic osteosarcoma, pathological analysis is critical in positive and differential diagnosis.

Keywords: Telangiectatic, Osteosarcoma, Cystic, Malignant

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INTRODUCTION

Telangiectatic osteosarcoma is a rare, aggressive morphologic variant comprising 2–4% of osteosarcomas [1–3]. Pathologic fracture is a clinical feature of this malignant bone-forming tumor [1]. This tumor resembles aneurysmal bone cyst radiologically and microscopically with large cavity containing blood filled cystic spaces separated by septa [4, 5]. There is usually invasion of soft tissues and formation of a large palpable mass resembling a soft tissue tumor at clinical examination. Historically, the preferred treatment for telangiectatic osteosarcoma was amputation. More recently, chemotherapy has changed the prognosis of this tumor, which is considered worse than conventional osteosarcoma in the first observations [1, 4–6].

We present a rare case of telangiectatic osteosarcoma in the femur of a 15-year-old boy with a brief review
of literature and emphasize on diagnostic difficulties, differential diagnosis, and treatment.

CASE REPORT

A 15-year-old boy was presented with a six month history of a growing mass in left thigh. The lump had been neglected. It grew until it reached the subcutaneous zone and ulcerated. The remainder of his medical history was non-contributory. He presented to the Children’s Hospital. He was very pale and asthenic.

He had severe anemia with hemoglobin of 5 g/dL. The rest of routine laboratory tests were within normal levels. Radiography showed an expansive lytic lesion of the distal end of the femur with cortical destruction, focal periosteal reaction and a pathologic fracture (Figure 1). It showed massive extension to adjacent soft tissue. Given the advanced stage of the disease, an amputation of the limb was performed.

At the pathology laboratory, we received a piece of the left lower limb amputation (Figure 2). We opened it. Gross examination of the resected distal femur revealed a 22x5 cm intramedullary multicystic hemorrhagic destructive tumor with cortical destruction and extension into the adjacent anterior and posterior soft tissues. The latter component of the mass was larger than the intraosseous tumor (Figure 3). The articular surface was not involved. We cut the femur in its longest axis with electric saw (Figure 4). Fixation in 10% neutral buffered formalin for 48 hours and decalcification of bone with nitric acid were performed. Slice of bone section was included in full. Many specimens from soft tissues and the surgical margins were also taken. Microscopic view of histological specimens of tumor stained with hematoxylin and eosin showed prominent blood filled cysts with malignant stroma in septa separating cysts (Figures 5 and 6). It contained atypical tumor cells oval or round of variable size with osteoblast-like multinucleated giant cells and a variable amount of immature osteoid (Figures 7–12). Numerous mitotic figures were noted (Figure 13). The tumor was located at 6 cm from the bone limit and 2 cm at the edge of the soft tissues. The resection margins were negative. The marrow, scooped and submitted separately, was negative.

The staging (bone scintigraphy, chest X-ray and abdominal ultrasound) showed no metastasis. The decision was to complete by six cycles of chemotherapy (Adriamycin (60 mg/d) and cisplatin (120 mg/d)). The postoperative course was unremarkable.

DISCUSSION

Telangiectatic osteosarcoma is a very rare tumor that account for less than 4% of all cases of osteosarcoma [1–3]. Paget was the first to describe it in 1854 [4, 5, 7]. Originally, it was thought to be a variant of conventional osteosarcoma, but it has several distinctive features.

It most frequently occurs in the second decade of life and has a male predominance (1.5:1 male/female ratio) [1–4]. The peak incidence and the anatomic distribution are similar to that of conventional osteosarcoma [1–5]. Most tumors occur in the metaphyseal region of long tubular bones [1, 2]. The distal femoral metaphysis is the single most common anatomic site, followed by the upper tibia and proximal humerus or proximal femur [1–4].

Telangiectatic osteosarcoma is rapidly expansile and behave aggressively. Clinical manifestations resemble those of conventional osteosarcoma, including pain and palpable mass [1, 2, 4, 5]. One characteristic clinical finding of this tumor is pathological fracture, being present in one-fourth of the cases, due to massive bone destruction [1].
Delayed diagnosis reported in previous publications varies from 3–4 months [2, 5, 7]. In our case, the causes of the delay in the diagnosis were fear of the diagnosis and the treatment and a low socio-economic status. Indeed, the mother was a housewife and the father did not have fixed income. This made difficult to access to the hospital and the diagnostic tests necessary at first times.

The diagnosis of telangiectatic osteosarcoma requires presence of a predominantly lytic destructive lesion of bone with no or minimal lesional sclerosis on radiographic imaging [1, 4]. Periosteal bone formation is
Figure 8: Septa shows highly pleomorphic stromal cells and tumor giant cells bordering spaces without endothelial linings. Note minimal osteoid.

Figure 9: (A, B) High magnification showing nuclear pleomorphism of sarcomatous septal cells and tumor giant cells (H&E stain, x400).

Figure 10: Tumor osteoid in septum bordering blood-filled space. Higher magnification showing irregular, finely divided (lace-like) strands of osteoid (H&E stain, x400).

Figure 11: Telangiectatic osteosarcoma with blue spiculated bone.

Figure 12: Telangiectatic osteosarcoma. Malignant osteoid is deposited in a lace-like pattern.
absent or minimal and is referred to as onion skin [1, 2]. The tumor commonly shows extension into soft tissues [1, 7]. Imaging features alone would also be compatible with an aneurysmal bone cyst, Ewing sarcoma, Langerhans cell histiocytosis, fibrosarcoma and malignant fibrous histiocytoma [8]. The pathologic findings enable confident exclusion of these radiologic differential diagnosis [4].

In our case, the gross findings were very suggestive of telangiectatic osteosarcoma. We had the gross appearance of an expansile hemorrhagic tumor with predominantly cystic spaces filled with blood, described as “a bag of blood” [1, 2, 4]. The tumor can have a honeycombed appearance with spongy areas containing smaller cysts. There is no fleshy or sclerotic bone formation. Features of invasive growth are often observed with extensive irregular cortical erosion and/or complete disruption of cortical continuity and massive invasion of soft tissue [1, 2, 4].

Microscopic findings revealed highly pleomorphic cells and foci of bone matrix formation between blood filled cyst like spaces, confirming the diagnosis of telangiectatic osteosarcoma. In some cases, where septa have broken down, atypical stromal cells may be identified, free-floating within the blood clot [4]. The amount of osteoid varies, but usually fine and lacelike osteoid is observed in minimal amount [1, 4, 6]. However, this feature is not essential to establish the diagnosis of telangiectatic osteosarcoma [4].

At this advanced local stage, telangiectatic osteosarcoma should be differentiated from an osteosarcoma of soft tissue. In the later, the tumor must arise in the soft tissues without any attachment to periosteum or bone [9].

The most important morphologic differential diagnosis is the benign ABC due to radiographic and gross appearance’s similarities. Microscopic examination is the only definitive diagnostic aid [4]. The presence of atypical or malignant stromal cells identified in telangiectatic osteosarcoma is never seen in aneurysmal bone cyst (ABC) [4]. The presence of atypical and/or overtly malignant cells is enough to rule out the diagnosis of ABC [4]. Furthermore, osteosarcoma is associated with aggressive growth features as indicated by cortical destruction and extension into the surrounding soft tissues [5]. In contrast, aneurysmal bone cysts cause marked expansible remodeling of bone and cortical thinning but lack true soft-tissue involvement [5].

Prognosis in the modern era is similar to conventional osteosarcoma [1, 4–6]. Indeed, Farr et al and Mervak et al found no prognostic differences between the two entities [10, 11].

Surgical resection of the tumor, limb-salvage as possible, is the treatment of choice, preceded by neoadjuvant chemotherapy [4–6]. Following resection, the histological response must be evaluated according to Huvos score based on the percentage of necrosis [12]. Depending on the grade, chemotherapy regimens are adapted after surgery [12]. In our case, given the emergency status, the patient was treated by primary amputation.

CONCLUSION

The case is reported because of the rarity of telangiectatic osteosarcoma along with the important role of pathological examination in making the positive and differential diagnosis. In our case, we had the typical histological appearance. Despite the locally advanced stage, the patient showed no metastasis.

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Hemodialysis catheter malposition: How to prevent this fault?

Fateme Shamekhi Amiri

CASE REPORT

A 26-year-old male, known case of solitary kidney disease presented with respiratory difficulty, sweating and frothy bloody discharge from oral cavity. At eight years of age, left vesicoureteral reflux detected from him and underwent antireflux operation (subureteric transurethral injection with teflon through cystoscopy). Four years later, he underwent antireflux operation of right vesicoureteral reflux. At 13 years of age, renal sonography revealed asymmetric kidney size (right kidney 57 mm, left kidney 85 mm), normal renal function (urea: 50 mg/dL [8.3 mmol/L], creatinine: 0.9 mg/dL [79.56 µmol/l]) and hematuria (RBC 18–20/hpf). The patient had not followed-up until one week before admission when he developed dyspnea on rest and then he referred to emergency room. No history of fever, oliguria, and dysuria was presented. Physical examination showed tachycardia, high blood pressure (150/105 mmHg), distended neck veins and grade ll/VI systolic murmur in apex, left sternal border and pulmonic area. Laboratory analysis revealed leukocytosis WBC 17900×10^3/µL, hemoglobin 10.6 g/dL and elevated ESR 50 mm/hr. Biochemical tests revealed hyperglycemia (FBS 130 mg/dL), elevated urea and creatinine (urea 184 mg/dL, cr 13.3 mg/dL) and (uric acid 9 mg/dL, Na+ 137/L, K+ 5.4 Meq/L), hyperlipidemia (cholesterol: 260 mg/dL, TG:274 mg/dL), increased total CPK and LDH (CPK 1600 U/I, LDH 639 U/I), negative troponin and decreased transferrin saturation percent (TSAT 6%). Urinalysis showed hematuria (RBC 2–3/hpf) and proteinuria (trace). Urine culture was negative. A 24-hour urine protein collection revealed proteinuria (345 mg), urine creatinine (480 mg) and urine volume (1200 cc). Renal sonography revealed small sized kidneys (right kidney 72 mm, left kidney 67 mm). Electrocardiography (EKG) showed inverted T-wave in precordial leads (V4–V6). Transthoracic echocardiography showed normal ejection fraction and 1° mitral regurgitation. On admission, for the patient inserted percutaneous hemodialysis catheter in chest by anesthesiologist and performed hemodialysis for 1.5 hr. One day later, chest X-ray revealed bilateral diffuse patchy infiltration in lungs that was in favor of uremic lung, also it showed hemodialysis subclavian vein catheter misplacement (Figure 1).

Then, hemodialysis catheter removed and percutaneous temporary uncuffed, non-tunneled double lumen catheter inserted in right jugular vein in neck and chest X-ray became normal (Figure 2).

**Diagnosis:** End-stage kidney disease due to chronic pyelonephritis/Reflux nephropathy. The patient is on hemodialysis and is preparing for kidney transplantation.

DISCUSSION

Cannulation of central veins and placement of catheters for temporary hemodialysis is a common procedure in the management of patients with end stage renal failure. The right internal jugular vein is the site of choice for central venous catheter placement, because it provides a more direct route into the right atrium and therefore helps prevent sheath kinking and further catheter secondary shift, being associated with the lowest complication rate. The procedure can be associated with a variety of malpositions of the catheter and rarely, can lead to significant morbidity and even mortality, if this is not recognized and not corrected early [1]. Non-tunneled percutaneous central vein catheter is usually placed at the bedside, while tunneled catheters can be placed in an interventional suite or operating room using fluoroscopic guidance.

Also study by trerotola demonstrated, using non-tunneled catheter, an incidence of central venous
thrombosis and/or stenosis of 40–50% with the subclavian route versus an incidence of 0–10% with the right internal jugular route [2].

A standard teaching and learning process in the medical field involves peer review and evaluation of fundamental mistakes that physicians encounter in clinical practice. These complications can be minimized and managed effectively by an experienced operator having increased awareness and using better technique [3]. Prior to the placement of central catheters, ultrasound imaging evaluates venous patency in patients who have a history of prior deep vein thrombosis in the region of the proposed access site. Familiarity with ultrasound-guided access is a critical aspect for the practitioner performing frequent central venous catheterization. Static ultrasound can be helpful to localize the vein for access when using techniques that rely on knowledge of anatomic landmarks (i.e., landmark technique), or alternatively dynamic ultrasound can be used to guide the needle into the vein in real time. The use of chest radiographs to establish the correct placement of central neck lines is to be routinely practiced [4]. Also the field of interventional nephrology needs to be viewed as a new road towards improving the dialysis access care provided to chronic kidney disease patients across the globe.

**CONCLUSION**

Improper placement technique can lead to early dysfunction and inadequate dialysis treatment. The current images are examples of demonstrating improperly placed temporary hemodialysis catheter by non-nephrologist and correction of this mistake by nephrologist. The advent of interventional nephrology in the world has created a new opportunity for the nephrologists to change their roles from thinkers to doers and leaders. The subspecialty is a step towards not only learning the necessary skills to perform dialysis access related procedures but to assume a leadership role in coordinating a medical team to provide the best the possible care.

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