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CASE REPORT

Case Report: Presentation of an unusual cause of carpal tunnel syndrome with accompanying literature review

[version 1; peer review: 3 approved with reservations]

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Abstract

Background: Carpal tunnel syndrome (CTS) is a condition seen commonly in clinical practice; high-flow arteriovenous malformations (AVM) can be a rare but important cause.

Case Report: We discuss a case of a patient who had developed left CTS in the fifth decade of life as the result of a progressively enlarging congenital peripheral AVM affecting his left upper limb. This case illustrates the clinical challenges encountered in the surgical and interventional management of this complex issue.

Discussion: High-flow AVMs affecting the extremities may be comprised of a convoluted network of vessels in high-flow, low-resistance systems that often recur despite intervention.

Conclusion: Peripheral AVM affecting the hand can be a rare and therapeutically challenging cause of carpal tunnel syndrome that warrants multidisciplinary team discussion.

Keywords

surgery, neuropathy, arteriovenous, congenital, malformation, interventional

Open Peer Review**Approval Status** ? ? ?

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22 Mar 2019	view	view	view

1. **Su Rak Eo** , Dongguk University Ilsan Hospital, Goyang, South Korea
2. **Guy Rubin** , Emek Medical Center, Afula, Israel
3. **Carlos Henrique Fernandes** , Federal University of São Paulo, São Paulo, Brazil

Any reports and responses or comments on the article can be found at the end of the article.

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Author roles: **Patiniott P:** Conceptualization, Data Curation, Formal Analysis, Investigation, Methodology, Project Administration, Resources, Software, Validation, Visualization, Writing – Original Draft Preparation, Writing – Review & Editing; **Herath M:** Conceptualization, Data Curation, Investigation, Writing – Review & Editing; **Riddell P:** Supervision, Writing – Review & Editing

Competing interests: No competing interests were disclosed.

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Introduction

Carpal tunnel syndrome (CTS) is a condition commonly encountered by physicians and surgeons alike, with the prevalence in the general population ranging from 3–5%¹ and accounting for up to 90% of all entrapment neuropathies². The finding of a congenital arteriovenous malformation (AVM) is a much less common occurrence in clinical practice, with some studies suggesting a prevalence of 1 in 100,000, with the majority of these occurring in the head and neck, with peripheral limb AVMs rarer still³. AVMs have a tendency to grow progressively larger with age, only becoming symptomatic when associated with haemodynamic instability or compression of surrounding structures⁴.

Literature review

Medline, PubMed, Ovid, WorldCat, ProQuest and Access Medicine databases were accessed with the input of the following search terms: arteriovenous malformation; arteriovenous malformation upper limb; arteriovenous malformation flexor retinaculum; compressive lesion upper limb. This initial search yielded 1290 results to which the following filters were applied: Peer reviewed; English Language; date within 25 years, which narrowed the results to 675. To further refine the search, of these articles, only those containing the following terms were included: carpal tunnel syndrome; carpal tunnel; median neuropathy; flexor retinaculum, which resulted in a total of 57 articles all of which were reviewed. Of these articles, 8 were identified as being similar.

The most similar case published in the literature was presented by Krishnamoorthy and colleagues who described a case of an 18-year-old man with persistent median artery and causing compression of the carpal tunnel with the median artery itself behaving as the main feeding vessel to the AVM⁵. Although the pathology described was similar, the lesion was less complex than the one reported in our case and accordingly, attracted a different approach to the management. The remaining 7 cases shared some similarity with respect to the aetiology of symptoms^{6–10} or the underlying pathology^{11,12}; however, in each of these cases, the lesions described were not as progressive or involved as that which was observed in our patient.

Case report

Our patient was a 58-year-old man who had migrated to Australia from South Sudan. He was left hand dominant with no known medical comorbidities or past surgical history. He had been referred to our general surgical outpatients clinic by his general practitioner with a nerve conduction test that had confirmed the presence of a severe left sided CTS.

The patient provided us with a 12 month history of transient paraesthesia in the distribution of the median nerve with sparing of the palmar cutaneous region. He reported that his left hand had been pulsatile his whole life and despite being ungainly was, until recently, asymptomatic. On inspection, his symptomatic left hand was 50% larger in comparison to his right. On palpation, thrills were appreciated in association with the multiple aneurysmal bulges. We hypothesised that an AVM was

present deep to the flexor retinaculum and had caused some degree of compression to the median nerve. We subsequently arranged for him to undergo CT angiography, which confirmed our suspicions. (Figure 1)

In an attempt to relieve his symptoms we arranged for the patient to undergo angiography with endovascular embolisation. Multiple large aneurysmal sacs were coiled and feeding vessels embolised; however, the extent of the abnormal vasculature in this instance was considerable and definitive treatment was unable to be established without incurring a significant risk of ischaemia to the patient's hand.

Immediately post-intervention, the patient did not report any difference in his symptoms and this remained to be the case until 6 months post-procedure. At this visit it was observed that his AVM had significantly reduced in size macroscopically and the patient reported improvement in his symptoms. This change correlated with a volumetric reduction on repeat CT angiogram; it was postulated that this could be attributed to a delayed fibrosis following embolisation (Figure 2).

Unfortunately, this improvement was only transient and at 9 months post procedure his AVM had again spontaneously reverted to its original pre-intervention dimensions.

The patient's case was subsequently discussed at several multi-disciplinary meetings, eventually a high-risk surgical procedure involving resection of the AVM with multiple vascular graft reconstruction was offered to the patient. However, he ultimately decided to continue living with his symptoms rather than risk compromising his hand function or viability.

Discussion

Direct compression of the median nerve by a congenital AVM within the flexor retinaculum is a particularly unlikely cause of carpal tunnel syndrome, made even more remarkable by

Figure 1. Coronal and sagittal views on CT angiography pre-embolisation.

Figure 2. Coronal and sagittal views on CT angiography 6 months post-embolisation.

the fact that the patient in this case only became symptomatic in the fifth decade of life.

Low-flow AVMs may be clinically less concerning as they are more amenable to surgical or endovascular intervention and appear to have lower rates of recurrence when compared to high-flow AVMs.

Most AVMs involving the extremities feature low-resistance niduses between the arteries and veins. Rosen and colleagues suggest that in the management of high-flow vascular malformations

if these niduses are not adequately controlled, then attempts at embolising or coiling of the proximal feeding vessels are likely to fail as new collateral feeders will invariably develop to replace them¹³, such as was the experience with our patient.

Conclusion

Peripheral limb high-flow AVM can be a rare and therapeutically challenging cause of carpal tunnel syndrome. It is a complex issue which warrants multidisciplinary team discussion, as complications of treatment and recurrence must be carefully weighed against the likelihood of improvement on a case-by-case basis.

Consent

Formal written consent for publication was obtained from the patient prior to the writing of the case report.

Data availability

All data underlying the results are available as part of the article and no additional source data are required.

Grant information

The author(s) declared that no grants were involved in supporting this work.

Acknowledgements

The authors of this case report would like to acknowledge that the images for this case report were made available to the authors with permission from the patient by the Department of Radiology at Flinders Medical Centre, Bedford Park, South Australia.

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Version 1

Reviewer Report 16 February 2021

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Carlos Henrique Fernandes

Department of Orthopedics and Traumatology, Escola Paulista de Medicina, Federal University of São Paulo, São Paulo, Brazil

Congratulations for the authors, very interesting and challenging case.

I have a few considerations about the case that I believe will be useful for the readers of the journal.

In the text the authors described , "On inspection, his symptomatic left hand was 50% larger in comparison to his right".

Reviewer: I believe that a photo of the patients hands are very important for a good comprehension.

In the text the authors described that the patient was undergo CT angiography, which confirmed their suspicions.

Reviewer: Why the choice by CT angiography? I think that the authors can include a discussion about the methods of diagnose.

What are the differential diagnosis? Please, include in the discussion.

A photo to demonstrate the hand volumetric reduction is necessary too.

Authors: The patient's case was subsequently discussed at several multidisciplinary meetings, eventually a high-risk surgical procedure involving resection of the AVM with multiple vascular graft reconstruction was offered to the patient.

Reviewer: A question, a simple decompression of the carpal tunnel could improve the patient symptoms? What are the possible consequences?
Please, include in the discussion section.

Authors: However, he ultimately decided to continue living with his symptoms rather than risk compromising his hand function or viability.

Reviewer: What the authors think about the patient future? What will occur with the patient? What the authors will do when the patient return? I Believe the possibilities can be included in the discussion

I believe that the conclusion can be limited to main purpose of the research, describe the AVM as a possible cause of median nerve compression inside of carpal tunnel.

Is the background of the case's history and progression described in sufficient detail?

Partly

Are enough details provided of any physical examination and diagnostic tests, treatment given and outcomes?

Partly

Is sufficient discussion included of the importance of the findings and their relevance to future understanding of disease processes, diagnosis or treatment?

Partly

Is the case presented with sufficient detail to be useful for other practitioners?

Partly

Competing Interests: No competing interests were disclosed.

Reviewer Expertise: Hand surgery, wrist arthroscopy, carpal tunnel syndrome, congenital hand deformities, fractures, tendon, nerve.

I confirm that I have read this submission and believe that I have an appropriate level of expertise to confirm that it is of an acceptable scientific standard, however I have significant reservations, as outlined above.

Reviewer Report 15 February 2021

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Guy Rubin

Orthopaedic Department, Emek Medical Center, Afula, Israel

This is a rare case report. I think the literature review should be more elaborate. There were 8 papers with similar cases. It is interesting to the reader to get some more information. Maybe a table that can summarize important information from each paper.

The discussion section should also be more informative and compare this case and what we know so far about similar cases.

Is the background of the case's history and progression described in sufficient detail?

Partly

Are enough details provided of any physical examination and diagnostic tests, treatment given and outcomes?

Partly

Is sufficient discussion included of the importance of the findings and their relevance to future understanding of disease processes, diagnosis or treatment?

Partly

Is the case presented with sufficient detail to be useful for other practitioners?

Yes

Competing Interests: No competing interests were disclosed.

Reviewer Expertise: hand surgery

I confirm that I have read this submission and believe that I have an appropriate level of expertise to confirm that it is of an acceptable scientific standard, however I have significant reservations, as outlined above.

Reviewer Report 19 August 2019

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Su Rak Eo

Department of Plastic and Reconstructive Surgery, Dongguk University Ilsan Hospital, Goyang, South Korea

Please add clinical photos of this patient - preop, and post procedural views.
Describe the AVM more in detail in shape, size, and characteristics.

This patients seemed to have persistent symptoms according to author's report. Please describe that in the discussion section.

Describe how the embolization was performed more in detail.

Is the background of the case's history and progression described in sufficient detail?

No

Are enough details provided of any physical examination and diagnostic tests, treatment given and outcomes?

Partly

Is sufficient discussion included of the importance of the findings and their relevance to future understanding of disease processes, diagnosis or treatment?

Partly

Is the case presented with sufficient detail to be useful for other practitioners?

Yes

Competing Interests: No competing interests were disclosed.

Reviewer Expertise: hand surgery

I confirm that I have read this submission and believe that I have an appropriate level of expertise to confirm that it is of an acceptable scientific standard, however I have significant reservations, as outlined above.

Author Response 22 Oct 2019

Paul Patiniott, Modbury Hospital, South Australia, Australia

Dear Dr Su Rak Eo,

Thank you for your review and valuable feedback relating to our case report on an unusual case of carpal tunnel syndrome.

I agree that the article can be improved by including additional pre and post operative clinical photographs, further describing the AVM in addition to elaborating on the technical / procedural details.

We are grateful for your expert peer-review of our literature review and case study.

Best wishes,

Dr Paul Patiniott
Surgical Registrar, SA Health, South Australia

Competing Interests: Nil conflicts of interest to disclose.

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