CASE REPORT

Spinal dural AV fistula: an unusual cause of chest pain

Gabriel Bioh, Richard Bogle

Department of Cardiology,
St Helier Hospital, Surrey, UK

Correspondence to
Gabriel Bioh,
gabioh@doctors.org.uk

SUMMARY

A 22-year-old man presented with 6 months of sudden onset, incapacitating, left-sided chest pain occurring 1–2 times a week. The severity of the pain caused loss of consciousness several times leading to multiple fractures. Investigation with echocardiogram, exercise tolerance test, Holter monitor, chest X-ray and V/Q scan revealed no abnormality as did EEG and 48 h video telemetry. MRI of the thoracic and lumbar spine showed a spinal dural arteriovenous fistula (SDAVF) at the level of T6, confirmed on angiogram. The patient underwent division of the left T6 AV fistula. Following the operation, the patient has been completely pain free. Our patient, presenting in his early 20s does not fit the usual age demographic for SDAVF. A second atypical feature is his presentation with chest pain alone and no neurological symptoms. This case represents a rare presentation of SDAVF.

BACKGROUND

This case is particularly interesting because the spinal dural arteriovenous fistula (SDAVF) presented with severe chest pain, a symptom encountered by general physicians on a regular basis, and no neurological symptoms as would be expected. It illustrates the need for persistence in pursuing a diagnosis for a symptom seen so commonly but having a profound effect on the patient's life. On review of the English literature, no other such case could be found, signifying a rare presentation of chest pain.

CASE PRESENTATION

A 22-year-old man presented to the emergency medical clinic with a 6-month history of excruciating left-sided chest discomfort, sudden in onset, stabbing in nature with no radiation but made worse by breathing and moving. He also experienced palpitations described as fast and regular but no associated shortness of breath, presyncope or syncope. He had no significant medical or family history, worked as a manual labourer, smoked 15 cigarettes a day and consumed 40 units of alcohol a week. Clinical examination was unremarkable.

Over the following 3 years, the patient had further episodes of chest pain of increasing frequency, 1–2 times a week lasting from a few minutes to half an hour. He also had multiple episodes of syncope, with falls resulting in multiple fractures. The patient was referred to a cardiology clinic for further investigation of this pain. A non-cardiac diagnosis was suspected prompting referral to a pain clinic where a cervical and thoracic spine MRI was booked, a neurology referral made, and amitriptyline was started for neuropathic pain.

INVESTIGATIONS

All initial investigations were normal including blood tests, chest X-ray, ECG, V/Q scan, echocardiogram, exercise tolerance test and 24 h Holter monitor. The patient was discharged with a diagnosis of musculoskeletal pain and managed with analgesia.

The neurologist ruled out epilepsy with an EEG and video telemetry of an episode. A diagnosis of vasovagal syncope secondary to severe chest pain was made.

In the cardiology clinic, the MRI of the thoracic spine was noted to show an SDAVF at the level of T6 and thought likely to be the cause of the chest pain (figure 1A).

Figure 1 Spinal dural arteriovenous fistula. (A) Sagittal T2-weighted MRI demonstrating flow void due to dilated intradural veins (white arrows). (B) Selective angiography of left T5–8 intercostal arteries demonstrates dilated intradural veins (white arrow).

To cite: Bioh G, Bogle R. *BMJ Case Rep* Published online: [please include Day Month Year] doi:10.1136/bcr-2013-202038

A vascular neurosurgical referral led to a spinal angiogram confirming the SDAVF to be supplied by a dural branch of the left T6 intercostal artery only (figure 1B).

DIFFERENTIAL DIAGNOSIS

The differential diagnosis for sudden onset stabbing chest pain is vast. In this case of a young active man with a physically demanding job, the initial diagnoses to be excluded were pulmonary embolus, spontaneous pneumothorax and pneumomediastinum, and lower respiratory tract infection. Myocardial infarction, most likely secondary to illicit drug use such as cocaine, must be considered as should aortic dissection. Other differentials to be considered once more severe pathology has been excluded include musculoskeletal pain and neuropathic pain.

With a history of palpitations, a symptomatic tachycardia required exclusion as well as underlying cardiomyopathy, although cardiomyopathy was unlikely with a completely normal ECG.

Subsequent episodes of syncope required the exclusion of epilepsy. Other differentials to be considered include orthostatic syncope, arrhythmogenic, for example, brady/tachyarrhythmias, cardiopulmonary, for example, aortic stenosis, and neurocardiogenic syncope thought most likely in this case secondary to pain.

TREATMENT

Division of the left T6 AV fistula resulted in relief of the pain with resolution of the dilated intradural veins (figure 2).

OUTCOME AND FOLLOW-UP

Follow-up of sagittal T2 and gadolinium-enhanced T1 spinal angiogram confirmed no evidence to suggest a recurrent dural fistula and no cord signal abnormality.

At 1 year cardiology follow-up, the patient remained pain free.

DISCUSSION

SDAVF was first described by Foix and Alajouanine in 1926.¹ It is a rare cause of myelopathy typically presenting in middle

aged men with a slowly progressive course. Symptoms include weakness and sensory loss ascending from the feet, bladder and bowel dysfunction. Median time to diagnosis ranges from 12 to 44 months.²

It is thought to be an acquired condition caused by arterialisat-ion, by a feeding radicular artery, of the normal perimedullary venous plexus. The arteriovenous shunt leads to venous congestion due to loss of the normal arteriovenous gradients, resulting in ischaemia.^{3 4}

The treatment of SDAVF involves either microsurgical open laminectomy with division of the draining fistula or endovascular embolisation of the fistula. There is currently much debate over the optimal treatment modality. However, a recent meta-analysis suggested superiority of surgery with a 98% successful fistula obliteration rate at follow-up compared with 46% with embolisation.⁵

It can be seen that our patient, presenting in his early 20s does not fit the usual age demographic for SDAVF. A second atypical feature is his presentation with chest pain alone and no neurological symptoms, correlating with the T2-weighted MRI (figure 1A) which demonstrated the cord to be intact with no signal abnormalities or swelling. It is possible that the SDAVF was incidentally identified while investigating the patient's chest pain, before it could cause a myelopathy. However, a causal link to the chest pain seems likely by virtue of the complete cessation of chest symptoms following obliteration of the SDAVF. This would then represent a very rare presentation of SDAVF with no other such recorded presentation in the literature to the authors' knowledge.

Learning points

- ▶ Chest pain has a wide and varied differential bridging multiple medical and surgical specialties.
- ▶ Persistence of symptoms despite therapy requires continuous revision of the differential diagnosis often necessitating further investigation.
- ▶ Spinal dural arteriovenous fistula can rarely present with severe chest pain.

Figure 2 Postoperative sagittal T2-weighted MR image demonstrating resolution of the dilated intradural veins.

Contributors GB was involved in the writing of article, figures, literature search and RB contributed to conception of article and critical revision of article.

Competing interests None.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

REFERENCES

- 1 Foix C, Alajouanine T. La myélite nécrotique subaigue: melite central angio-hypertrophique a evolution progressive: paraplégie amyotrophique lentement accendante, d'abord spasmodique, puis 15 flasque, s'accompagnant de dissociation albumin-cytologique. *Rev Neurol (Paris)* 1926;33:1–42.
- 2 Jellema K, Tijssen CC, van Gijn J. Spinal dural arteriovenous fistula: a congestive myelopathy that initially mimics a peripheral nerve disorder. *Brain* 2006;129:3150–64.
- 3 Aminoff MJ, Barnard RO, Logue V. The pathophysiology of spinal vascular malformations. *J Neurol Sci* 1974;23:255–63.
- 4 Kendall BE, Logue V. Spinal epidural angiomatous malformations draining into intrathecal veins. *Neuroradiology* 1977;13:181–9.
- 5 Steinmetz MP, Chow MM, Krishnaney AA, et al. Outcome after the treatment of spinal dural arteriovenous fistulae: a contemporary single-institution series and meta-analysis. *Neurosurgery* 2004;55:77–87.

Copyright 2014 BMJ Publishing Group. All rights reserved. For permission to reuse any of this content visit <http://group.bmj.com/group/rights-licensing/permissions>.
BMJ Case Report Fellows may re-use this article for personal use and teaching without any further permission.

Become a Fellow of BMJ Case Reports today and you can:

- ▶ Submit as many cases as you like
- ▶ Enjoy fast sympathetic peer review and rapid publication of accepted articles
- ▶ Access all the published articles
- ▶ Re-use any of the published material for personal use and teaching without further permission

For information on Institutional Fellowships contact consortiasales@bmjgroup.com

Visit casereports.bmj.com for more articles like this and to become a Fellow