CASE REPORT

Solitary spinal epidural cavernous haemangiomas as a rare cause of myelopathy

Ying Meng,¹ Mohammed F Shamji²¹Department of Faculty of Medicine, University of Toronto, Toronto, Ontario, Canada²Department of Surgery, Toronto Western Hospital, Toronto, Ontario, Canada**Correspondence to**
Dr Mohammed F Shamji,
mohammed.shamji@uhn.ca

Accepted 12 September 2015

SUMMARY

Cavernous haemangiomas rarely occur in the spinal epidural space. We report the case of a 27-year-old man who presented with myelopathy secondary to spinal cord compression from a purely epidural lesion. The imaging characteristics of cavernous haemangiomas are unique, reflecting a highly vascular lesion. Key differentiating features from intracranial or intramedullary lesions include the lack of a surrounding hemosiderin ring and popcorn appearance. An urgent referral to a neurosurgeon is recommended given the possibility of acute neurological deterioration from intralesional haemorrhage, and good recovery from early surgical resection. Preoperative planning with thorough patient counselling and availability of matched blood is important, and an en bloc resection approach should be taken to minimise blood loss. In this case, the patient experienced complete recovery after surgical resection. No recurrence after complete resection has been reported in the literature. This suggests a good long-term outcome for the patient and that no early adjuvant therapy is necessary.

BACKGROUND

Cavernous angiomas are, most commonly, intracranial pathologies, but they also present in the spine, however, with an incidence of only 0.22 per million per year.¹ Many of these originate from the vertebral body with extension into the epidural space. Very rarely, cavernous haemangiomas are located in the epidural spinal canal without any involvement of the overlying bone. On MRI, these lesions appear homogenous and brightly enhanced with contrast. They are benign lesions but can cause chronic progressive or acute neurological deficits. Surgical resection is the mainstay treatment. They are highly vascular and, whenever possible, an en bloc gross total resection approach should be attempted to avoid unnecessary blood loss and future recurrence.

We describe a case of solitary spinal epidural cavernous haemangioma in a young man presenting with early myelopathy, with no evidence of erosion or scalloping of the surrounding lamina. This case highlights a rare cause of myelopathy. We will review the differential diagnosis, and discuss the clinical findings as well as the principles behind the management considerations. A better understanding of the radiological features of this tumour will help improve patient care.

CASE PRESENTATION

A 27-year-old otherwise healthy man presented to his doctor with cough-induced back pain

accompanied by bilateral lower extremity paraesthesia, numbness, and progressive weakness and stiffness over the course of months. The back pain was localised to the mid-thoracic region. There was no radicular leg pain. The patient denied any fever, chills or weight loss, and he did not have bowel, bladder or sexual dysfunction, nor did he have any significant medical history, or history in his immediate family members.

On physical examination, he had decreased sensation on pinprick, from umbilicus downwards, indicating a sensory level at approximately T10. He was not able to differentiate light touch from right and left. Motor examination revealed normal power in the upper and lower extremities, which was graded at 4/5, with the exception of bilateral plantar flexion. Patellar and ankle reflexes were brisk. The patient's gait was steady, but he could not perform tandem gait.

Based on the overall clinical picture, this patient appeared to have a subacute presentation of myelopathy. We localised the lesion to the mid-thoracic spine around T10 level and, given the timeline and relevant negatives, suspected neoplasm to be the most likely aetiology.

INVESTIGATIONS

Our patient's routine blood work was unremarkable. In this case, medical imaging would be key in assisting with diagnosis. MRI of the thoracic spine revealed a T7–8 lesion occupying a significant portion of the dorsal spinal canal (figure 1). It appeared extramedullary and likely extradural. It measured 5 cm longitudinally and was compressing the spinal cord without causing significant cord oedema or T2-weighted signal change, which indicated cord injury. Remarkable homogenous gadolinium contrast enhancement suggested that this was a highly vascular tumour. CT of the same region did not demonstrate any bony erosion or scalloping around the lesion (not shown).

DIFFERENTIAL DIAGNOSIS

Spinal tumours can be classified as either bony, epidural, intradural extramedullary or intradural intramedullary (ie, inside the spinal cord). Common pathology in the epidural compartment includes metastasis, lymphoma, lipoma and other rare tumours. Intradural extramedullary lesions include schwannoma, meningioma, neurofibroma, myxopapillary ependymoma and leptomeningeal metastasis. Intradural intramedullary lesions are most frequently astrocytoma or ependymoma, and, rarely, hemangioblastoma or metastasis. The differential diagnosis for this homogenous T1

CrossMark

To cite: Meng Y, Shamji MF. *BMJ Case Rep* Published online: [please include Day Month Year] doi:10.1136/bcr-2015-211644

Figure 1 (A) Sagittal T1-weighted MRI demonstrating T7–8 epidural lesion isointense with the spinal cord; (B) the lesion enhances brightly with gadolinium; (C) axial T2-weighted MRI, the epidural lesion displaces the spinal cord anteriorly; (D) sagittal T2-weighted MRI showing a hyperintense epidural lesion. No significant T2 cord signal change was found.

hypointense and T2 hyperintense lesion with peripheral contrast enhancement and no bony erosion on CT scan is of benign neoplastic aetiology, differentiating lymphoma as a common entity from the rarer cavernous angioma. A spinal epidural haematoma would be expected to have a heterogeneous appearance and a spinal epidural abscess would be expected to have more loculations and substantial rim enhancement. Given the potential of a vascular lesion in the epidural space, the decision against percutaneous biopsy was taken in deference to surgical intervention.

TREATMENT

Given the patient's progressively worsening symptoms, surgical intervention was indicated to acutely decompress the spinal cord. A secondary surgical indication was to obtain tissue for definitive diagnosis. Percutaneous biopsy was deemed dangerous by the consulting interventionist, given the highly vascular nature of the lesion and the attendant risk of haemorrhage. Lastly, surgical resection provides cytoreduction for adjuvant therapies if indicated.

The patient underwent thoracic laminectomy for lesion resection, with intraoperative findings of an epidural mass easily differentiated from the underlying dura. Grossly, it appeared red, friable and globular. A gross total resection was achieved with confirmation of no intradural disease and effective decompression of the spinal cord using intraoperative ultrasound.

OUTCOME AND FOLLOW-UP

Postoperatively, the patient reported immediate resolution of his sensory symptoms and moderate improvement in his gait instability. He was discharged 2 days after surgery to continue his recovery at home. Follow-up spine MRI demonstrated no

residual tumour. Final pathology of the surgical specimen revealed large diameter blood vessels filled with blood, and intervening connective and adipose tissue typical of cavernous haemangiomas (figure 2). Given that cavernous haemangiomas are benign lesions, the patient did not require further adjuvant therapy. His neurological examination had normalised at 1-year follow-up, with no clinical evidence of myelopathy.

DISCUSSION

Cavernous haemangiomas are malformations of the microcirculation, and are composed of mature, thin-walled vessels and sinuses lined by endothelial cells. These can affect any part of the neuroaxis and are most commonly found in the intracranial compartment and are intra-axial in location. Nevertheless, approximately 5% of cases manifest in the spine, where they are most frequently located in the spinal epidural space.² Spinal cavernous haemangiomas account for 4% of all spinal epidural tumours, and typically involve the vertebral bodies from where they extend into the epidural space.³ Alternatively, solitary spinal epidural cavernous haemangioma without any bony involvement is a rare cause of myelopathy.⁴ A review of the current literature shows only a hundred reported cases,⁵ this is in marked contrast to the prevalence of myelopathy as the most common cause of acquired spinal cord dysfunction worldwide. Grasso and *et al*⁶ have reported on an intramedullary lesion case and summarised the literature around spinal lesions in all compartments, finding that half of patients present with subacute progressive myelopathy and half of patients present with acute deficits. Li *et al*⁷ reported a series of 14 patients with epidural cavernous haemangiomas, finding that 50% were misdiagnosed for other entities when surgery was undertaken.

Figure 2 Histology of SSECA, H&E stain, demonstrating (A) large diameter blood vessels filled with blood; (B) intervening fibroconnective and adipose tissue and (C) under high power (40×) magnification.

Preoperative haemorrhage was seen in two of the cases and neurological improvement observed in 50% of those reaching a mean of 34 months follow-up, after which three patients exhibited recurrent disease.

Based on a review of 54 reported cases of spinal epidural cavernous haemangioma, there was a 66% male predominance. The mean age at presentation was 48 years in males and 45 years in females. These lesions are most commonly localised in the thoracic (80%) spine, more specifically posterior to the spinal cord in 93% of the cases.^{5 8} Khalatbari *et al*⁹ reported a 58% predilection for the thoracic spine, 26% for the cervical and 16% for the lumbar spine. They typically present in an indolent fashion with progressive myelopathy. The pathophysiology of myelopathy is postulated to be secondary to direct compression or vascular steal phenomenon. Radiculopathy can also be present secondary to foraminal extension of the tumour. Few cases describe sudden or intermittent symptoms due to haemorrhage within the lesion causing acute expansion.^{10 11} Pathological features include prominent thin-walled blood-filled vascular spaces within the lesion and, under high magnification, small foci of adipocytes within irregular fibrous intravascular septations. Furthermore, immunohistology stains are positive for CD34 but negative for CD31, which is consistent with the literature.¹¹

MRI is the diagnostic modality of choice. Spinal epidural cavernous haemangiomas are isointense with the spinal cord on T1, and hyperintense on T2. They brightly enhance homogeneously with contrast.¹² They do not resemble intracranial cavernomas, which have a characteristic popcorn appearance due to T2 signal loss from hemosiderin deposit after previous haemorrhages. Similarly, cavernous angiomas within the spinal cord may have a surrounding hypointense T2 hemosiderin rim. These features are rarely seen for epidural cavernous haemangiomas. Finally, overlying bony changes (eg, erosions, scalloping) may be appreciated on computer tomography, which may help in the differential diagnosis and surgical planning.³ In the case of our patient, his tumour did not cause any bony abnormalities.

Given these radiological findings, it was wise to consider spinal epidural cavernous haemangiomas in the differential diagnosis. Despite the benign nature of these tumours, surgical

resection is the treatment of choice to remedy the ongoing symptomatic spinal cord compressive myelopathy, and to avoid the risk of haemorrhage and acute spinal cord. While the MRI findings are not definitive, its consideration is important as it guides further diagnostic steps away from percutaneous biopsy, which risks periprocedural lesional haemorrhage, in deference to surgical intervention for both diagnostic and thus driven therapeutic objectives. Rarely, these tumours have been managed non-operatively in the setting of normal neurological examination or improvement.¹³ Despite this, surgical resection is still recommended, given the potential acute neurological deterioration due to intralesional haemorrhage.^{8 11}

Owing to the tumour's vascular nature, the initial dissection of surrounding structures as well as tumour resection can incur significant bleeding. A thoughtful surgical approach involves identifying and coagulating the feeding vessels, after which the lesion is peeled away from the dura en bloc. Piecemeal resection should be avoided to reduce uncontrolled blood loss. A good plane often exists between the lesion and dura, thus making gross total resection a realistic goal. Based on the previous literature, solitary spinal epidural cavernous hemangiomas have an indolent natural history,⁸ with no reported recurrence after gross total resection. This suggests that a complete resection is curative and no further adjuvant therapy is necessary.

Learning points

- ▶ Solitary spinal epidural cavernous haemangioma is a rare cause of myelopathy with characteristic radiological findings.
- ▶ Preoperative planning with thorough patient counselling and availability of matched blood is important given the highly vascular nature of the tumour.
- ▶ An en bloc resection approach should be taken to minimise blood loss.
- ▶ Based on case series, these tumours are benign. The patient's long-term outcome is good and no further adjuvant therapy is necessary if a gross total resection is achieved.

Competing interests None declared.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

REFERENCES

- 1 Hemalatha AL, Ravikumar T, Chamarthy NP, *et al.* A pure epidural spinal cavernous hemangioma—with an innocuous face but a perilous behaviour!! *J Clin Diagn Res* 2013;7:1434–5.
- 2 Goyal A, Singh AK, Gupta V, *et al.* Spinal epidural cavernous haemangioma: a case report and review of literature. *Spinal Cord* 2002;40:200–2.
- 3 Rovira A, Rovira A, Capellades J, *et al.* Lumbar extradural hemangiomas: report of three cases. *AJNR Am J Neuroradiol*. 1999;20:27–31.
- 4 Akiyama M, Ginsberg HJ, Munoz D. Spinal epidural cavernous hemangioma in an HIV-positive patient. *Spine J* 2009;9:e6–8.
- 5 Sharma MS, Borkar SA, Kumar A, *et al.* Thoracic extraosseous, epidural, cavernous hemangioma: case report and review of literature. *J Neurosci Rural Pract* 2013;4:309–12.
- 6 Grasso G, Alafaci C, Granata F, *et al.* Thoracic spinal cord cavernous angioma: a case report and review of the literature. *J Med Case Rep* 2014;8:271.
- 7 Li TY, Xu YL, Yang J, *et al.* Primary spinal epidural cavernous hemangioma: clinical features and surgical outcome in 14 cases. *J Neurosurg Spine* 2015;22:39–46.
- 8 Aoyagi N, Kojima K, Kasai H. Review of spinal epidural cavernous hemangioma. *Neural Med Chir (Tokyo)* 2003;43:471–5; discussion 6.
- 9 Khalatbari MR, Abbassioun K, Amirjmsidi A. Solitary spinal epidural cavernous angioma: report of nine surgically treated cases and review of the literature. *Eur Spine J* 2013;22:542–7.
- 10 Caruso G, Galarza M, Borghesi I, *et al.* Acute presentation of spinal epidural cavernous angiomas: case report. *Neurosurgery* 2007;60:E575–6; discussion E6.
- 11 Jang D, Kim C, Lee SJ, *et al.* Pure spinal epidural cavernous hemangioma with intralésional hemorrhage: a rare cause of thoracic myelopathy. *Korean J Spine* 2014;11:85–8.
- 12 Shivaprasad S, Shroff G, Campbell GA. Thoracic epidural cavernous hemangioma imaging and pathology. *JAMA Neurol* 2013;70:1196–7.
- 13 Clarke DB, Bertrand G, Tampieri D. Spontaneous spinal epidural hematoma causing paraplegia: resolution and recovery without surgical decompression. *Neurosurgery* 1992;30:108–11.

Copyright 2015 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