Unusual presentation of more common disease/injury

Spinal extradural cavernous haemangioma in an elderly man

Asifur Rahman, Saif Ul Hoque, Paawan Bahadur Bhandari, Abu Saleh Md Abu Obaida

Department of Neurosurgery, Bangabandhu Sheikh Mujib Medical University, Dhaka, Bangladesh

Correspondence to Dr Asifur Rahman, bijoun14@yahoo.com

Summary

Cavernous haemangiomas are vascular malformations that may affect any part of the central nervous system. Epidural haemangiomas are rare and constitute ~4% of all epidural tumours and 12% of all intraspinal haemangiomas. These tumours enlarge slowly and produce symptoms of progressive myelopathy or radiculopathy or both. History, clinical examination, routine radiographs, MRI and histopathological studies are the aids for a definitive diagnosis. Surgery can give a very beneficial result with good functional and neurological improvement. Chance of recurrence is less after a good surgical removal. Here we present a case of spinal extradural cavernous haemangioma in a 65-year-old man who had a good functional and neurological recovery after surgery. At 9 months postoperative follow-up, he did well without any new problems with regard to recurrence. We report this case for its rarity.

BACKGROUND

Spinal cavernous haemangioma without any primary involvement of the vertebrae is a very rare condition and only about 80 cases have been reported so far in English language literature.

CASE PRESENTATION

A 65-year-old man presented with painless progressive paraparesis for 9 months which was ascending in nature, starting from the right lower limb, and not associated with trauma, fever, weight loss or night pain. He developed loss of sensation of both lower limbs gradually for the last 2 months and became paraparetic with muscle power of grade 2 for the last 15 days. Muscle tone in the lower extremities was increased bilaterally with brisk deep tendon reflexes, extensor plantar responses and patellar and ankle clonus. All modalities of sensation were diminished from the level of dorsal eight downwards. He had no bladder or bowel dysfunction.

INVESTIGATIONS

Plain x-ray of dorsal spine showed mild scoliosis. T1-weighted (T1WI) MRI of the dorsolumbar spine revealed an extradural fusiform isointense lesion extending from D7 to D9 level with extension into the intervertebral foramen at the lower level of D7 vertebra (figures 1A and 2A). The lesion was hyperintense in T2WI with a hypointense rim between the lesion and the dura (figures 1C and 2C) and had intense and uniform contrast enhancement (figures 1B and 2B).

DIFFERENTIAL DIAGNOSIS

Angiomatous meningioma, metastasis, pure epidural haematoma, granulomatous infections, neurogenic tumours, lymphomas, angiolioma and epidural extramedullary haematopoiesis.

TREATMENT

The patient underwent laminectomy of D7–D9. Total excision of the extradural, well circumscribed, red, fleshy, soft to firm, moderately vascular lesion with moderate

vascular adhesion was accomplished en-mass. The lobulated part in the foramen was also removed after widening of the foramen (figure 3A,B). Histopathology revealed cavernous haemangioma (figure 4) which was confirmed with immunohistochemistry.

OUTCOME AND FOLLOW-UP

The postoperative period was uneventful and the patient was discharged on ninth postoperative day. By 9 months, follow-up the patient was ambulatory with power of Medical Research Council (MRC) grade 4+ in both lower limbs but he was lost to follow-up thereafter. Because of financial constraint of the patient we could not do any follow-up MRI.

DISCUSSION

Haemangiomas are congenital vascular malformations that pathologists frequently consider to be hamartomatous malformations. Affecting any part of the neuraxis, they are usually localised in the hemispheres or in the brainstem and may be associated with haemorrhages, focal deficits and seizures. Haemangiomas of the spine are usually lesions of the vertebral bodies, and purely epidural haemangiomas are rare.^{1–8} They constitute ~4% of all epidural tumours and 12% of all intraspinal haemangiomas. Spinal epidural cavernous haemangioma without a primary origin in the vertebral bone is rare, with only 80 cases reported in English literature.^{5–7 9 10} The patients with this type of lesion usually present in the fifth or sixth decade.¹¹ These lesions most commonly develop at the posterior aspect of the epidural space with an affinity for the thoracic or lumbar regions.^{2 5 6}

Epidural cavernous haemangiomas are usually associated with spinal pain, radiculopathy, progressive compressive myelopathy and rarely with signs of acute or remote haemorrhages. Progressive neurological deterioration can be explained by the slow growth rate of this benign lesion. Rarely, there may be acute compressive features because of associated extradural haemorrhage or sudden expansion of the lesion caused by microhaemorrhages or macrohaemorrhages in the haemangioma or thrombosis.^{3 6 7 9–11} Our

Figure 1 MRI sagittal views in T1-weighted images (WI) (A), postcontrast (B) and T2WI (C) showing isointense fusiform extradural lesion (A), homogenous intense contrast uptake with dural tail sign like enhancement (white arrows in B) and hyperintense fusiform extradural lesion with hypointense rim between the lesion and the dura (black arrow in C).

patient had progressive spastic paraparesis which is a feature of progressive myelopathy. He developed it progressively as the lesion increased in size gradually. The progression of paraparesis was rapid at the later stage most likely because of non-compliance of the spinal cord from compression by the lesion any more.

Although plain x-rays may show evidence of compression such as scalloping, erosion or honeycomb appearance, x-ray of the dorsolumbar spine in this patient only showed mild scoliosis. Currently, MRI is the modality of choice.^{6 10} In most of the cases, the T1WI of spinal cavernous haemangioma shows a homogenous signal intensity isointense to that of spinal cord and which might be due to slow blood flow, while on T2WI the signal of the lesion is consistently hyperintense, which may be explained by the high content of stagnant blood.^{2 3 7 10} In our case, the MRI in T1WI showed a typical isointense lesion which was hyperintense in T2WI with a low signal intensity rim and had homogenous brilliant contrast enhancement. Lee *et al*⁵ found a rim of low T2 signal intensity in 57% of cases in their study. The cause of the low signal intensity rim is uncertain. It may be one of or a combination of (1) the fibrotic capsule itself, (2) the interface between a mass and adjacent dura or posterior longitudinal ligament, (3) a chemical shift artefact created by epidural fat and intratumoural fluid artefact and (4) deposition of hemosiderin.^{5 7} We believe that the thin hypointense rim between the lesion and the cord is the interface between the mass and

the dura. It could be the dura itself or deposition of hemosiderin. Homogenous contrast enhancement can be because of the sinusoidal channel structure of the lesion.^{9 10} Frequently, the lesion is found to be extended to the intervertebral foramen, which may give rise to confusion with schwannoma or meningioma.^{3 7} Unlike in the management of capillary haemangiomas, digital subtraction angiography (DSA) as an investigation tool or a preoperative surgical adjunct has no role in the management of cavernous haemangioma.^{8 11}

The treatment for these lesions is total removal of the tumour with microsurgical technique. Because of their hypervascularity, piecemeal resection should be avoided, as excessive intraoperative bleeding resulting in incomplete resection might occur.^{6-8 10} Reoperation for remnant or recurrent epidural haemangioma is very difficult because of peridural or periradicular adhesion and unclear tumour margins; so, complete resection cannot be guaranteed in reoperation.^{1 7} Gross total resection with careful circumferential dissection away from the dura should be the goal. However, the foraminal portion may present a more challenging component.¹¹ When total excision is not possible adjuvant radiotherapy is advised.¹⁰ As we felt that we had achieved a total removal we did not advise the patient for any adjuvant therapy.

The cavernous type of haemangiomas display histologically a large number of closely packed sinusoidal thin-walled channels, lined by a single layer of endothelial cells

Figure 2 MRI axial views in T1WI (A), postcontrast (B) and T2WI (C).

Figure 3 Intraoperative pictures showing the interface between the lesion and the dura (white arrow in A), clear dura after removal of the haemangioma (black arrow in B) and space after removal of the transforaminal part (white arrow in B).

in collagenous tissue without interposed neural tissue. Some of these channels are filled with blood. Elastic fibres and smooth muscle fibres within the walls are scant if not absent. Immunohistochemistry shows a positive reaction for CD31 and CD34 but is negative for S100 and epi-

Figure 4 Photomicrograph (H&E, ×100) of the lesion showing variable-sized vascular channels lined by single layer of endothelial cells (black arrow) containing red blood cells (white arrow). Connective tissue stroma (red arrow) and occasional adipocytes (blue arrow) are also seen.

thelial membrane antigen.^{2 3 7 10 12} Histopathology of the excised mass in our case revealed variable-sized vascular channels lined by single layer of endothelial cells in connective tissue stroma. Most of these channels contained red blood cells. Stromal foam cells were seen and intermingled adipocytes were present. It was negative for epithelial membrane antigen.

Learning points

- ▶ Extradural cavernous haemangioma should be kept in mind as a differential diagnosis in a patient with MRI scans demonstrating an isointense lesion on T1WI and hyperintense on T2WI showing homogenous contrast enhancement.
- ▶ Proper preoperative planning and complete resection during the first surgery is essential.
- ▶ Surgery can give a very beneficial result with good functional and neurological improvement and early treatment of extradural cavernous haemangioma is advocated to prevent further deterioration and permanent neurological deficits.

Competing interests None.

Patient consent Obtained.

REFERENCES

1. **Badinand B**, Morel C, Kopp N, *et al*. Dumbbell-shaped epidural capillary hemangioma. *AJNR Am J Neuroradiol* 2003;**24**:190–2.
2. **Rovira A**, Capellades J, Zauner M, *et al*. Lumbar extradural hemangiomas: report of three cases. *Am J Neuroradiol* 1999;**20**:27–31.
3. **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.
4. **Hatiboglu MA**, Iplikcioglu AC, Ozcan D. Epidural spinal cavernous hemangioma. *Neurol Med Chir (Tokyo)* 2006;**46**:455–8.
5. **Lee JW**, Cho EY, Hong SH, *et al*. Spinal epidural hemangiomas: various types of MR imaging features with histopathologic correlation. *Am J Neuroradiol* 2007;**28**:1242–8.
6. **Antunes A**, Beck MF, Strapasson AC, *et al*. Extradural cavernous hemangioma of thoracic spine. *Arq Neuropsiquiatr* 2011;**69**:720–1.
7. **Minh NH**. Cervicothoracic spinal epidural cavernous hemangioma: case report and review of the literature. *Surg Neurol* 2005;**64**:83–5; discussion 5.
8. **Vassal F**, Peoc'h M, Nuti C. Epidural capillary hemangioma of the thoracic spine with proximal nerve root involvement and extraforaminal extension. *Acta Neurochir (Wien)* 2011;**153**:2279–81.
9. **Aoyagi N**, Kojima K, Kasai H. Review of spinal epidural cavernous hemangioma. *Neurol Med Chir (Tokyo)* 2003;**43**:471–5; discussion 6.
10. **Satpathy DK**, Das S, Das BS. Spinal epidural cavernous hemangioma with myelopathy: a rare lesion. *Neurol India* 2009;**57**:88–90.
11. **Hasan A**, Guiot MC, Torres C, *et al*. A case of a spinal epidural capillary hemangioma: case report. *Neurosurgery* 2011;**68**:E850–3.
12. **Nowak DA**, Widenka DC. Spinal intradural capillary haemangioma: a review. *Eur Spine J* 2001;**10**:464–72.

This pdf has been created automatically from the final edited text and images.

Copyright 2012 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.

Please cite this article as follows (you will need to access the article online to obtain the date of publication).

Rahman A, Hoque SU, Bhandari PB, Abu Obaida ASM. Spinal extradural cavernous haemangioma in an elderly man. *BMJ Case Reports* 2012;10.1136/bcr-2012-006453, Published XXX

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