CASE REPORT

Acute compressive myelopathy due to vertebral haemangioma

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SUMMARY

A 47-year-old woman with a history of anaemia presented to the emergency room with an acute onset of leg weakness. Physical examination of the bilateral lower extremities was significant for 0/5 muscle strength in all muscle groups with decreased pinprick and temperature sensation. A sensory level at the umbilicus was appreciated. Fine touch and proprioception were preserved. Bowel and bladder function were intact. CT revealed several thoracic, vertebral haemangiomas. An MRI was suggestive of an epidural clot at the T8–T10-weighted posterior epidural space. At the level of the lesion, the cerebrospinal fluid space was completely effaced, and the flattened spinal cord exhibited signs of oedema and compressive myelopathy. The patient immediately underwent surgical decompression of the spinal cord. An epidural clot and vessel conglomeration were identified. A postoperative spinal angiogram confirmed the diagnosis of vertebral haemangioma. At 1-month follow-up, the patient regained strength and sensation.

to the emergency room, the patient developed a sudden onset of bilateral, lower extremity weakness. She denied similar occurrences in the past as well as any recent fevers, chills or trauma. The patient is married with children. Aside from the anaemia, she is otherwise healthy with no occupational exposures or smoking history. Family history is not significant for any cardiac, haematological or neurological disease. On physical examination, the patient had intact reflexes, sensation, tone and bulk in the upper extremities with 5/5 strength on manual motor testing. Strength in all muscle groups of the lower extremity were 0/5. Sensation was decreased to pinprick and temperature in the lower extremities. Fine touch and proprioception were preserved. A sensory level was appreciated at the level of the umbilicus. Beever's sign was positive, but the abdominal cutaneous reflex was negative. These findings localise the lesion to the thoracic spine. Reflexes could not be elicited in the lower extremities (0/4). Abnormal reflexes were likely due to the acute onset of spinal shock.

BACKGROUND

Vertebral haemangiomas are typically benign vascular tumours, incidentally identified in the spinal column in 10–12% of the population.¹ Traditionally associated with the vertebral body, the tumour rarely exhibits posterior element involvement or extraosseous expression.¹ Although vertebral haemangiomas are commonly asymptomatic, association with the thoracic spine and extension into the neural arches increase the likelihood of the lesion compressing the spinal cord.² In this case report, we present a patient with anaemia with an acute onset of compressive myelopathy localised to an epidural bleed from a thoracic haemangioma invading the posterior elements. This case underscores the importance of an appropriate assessment of vertebral haemangiomas, particularly of the thoracic spine, in order to prevent prolonged compressive myelopathy and permanent neurological sequelae.

INVESTIGATIONS

Complete cell count and differential were consistent with anaemia: low red blood cell count, haemoglobin, haematocrit and red blood cell distribution width. Laboratory findings were consistent with microcytic, hypochromic anaemia. An urgent CT angiography was negative for vascular injury of the aorta or its branch vessels. However, the spinal cord was compressed from T8 to T10 by a posterior hyperdensity, concerning for an epidural or subdural haematoma. A simultaneous CT revealed multiple vertebral haemangiomas with the largest lesion within the T8 vertebral body followed by a smaller lesion at T9. A subsequent MRI revealed a mass in the posterior epidural space between T8 and T10. Measuring 4.4 cm in length, 1.5 cm in transverse width and 9 mm in anteroposterior width, the epidural mass demonstrated heterogeneous STIR hyperintensity, T1-weighted hypointensity and a thin rim of peripheral contrast enhancement—all of which suggest an epidural haematoma. The abutting spinal cord was reduced to 4.5 mm in anteroposterior diameter with corresponding diffuse STIR and T2-weighted hyperintensity, concerning for cord oedema and compressive myelopathy (figures 1A, B and 2). Moreover, a diffusely T1-weighted hypointense bone marrow suggested a haematological disorder. A spinal angiogram after addressing the patient's symptoms confirmed the diagnoses of vertebral haemangioma.

CASE PRESENTATION

A 47-year-old woman with a medical history of anaemia presented to the emergency room reporting epigastric and back pain for 2 days. She described an intermittent, pressure-like discomfort, 'like a knot in my stomach.' After walking in order to alleviate her abdominal pain, she experienced bilateral lower extremity sensory disturbances lasting several minutes. The symptoms prompted the patient to seek urgent medical care. On arrival

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Figure 1 Preoperative T2-weighted MRI. (A) The sagittal image reveals a posterior epidural clot extending from T8 to T10. (B) The axial image demonstrates anterior spinal cord compression against the T7–T8-weighted herniated disc. Although the blood clot is dorsal to the spinal cord, cerebrospinal fluid effacement and cord flattening is most prominent in the anterior epidural space. Postoperative T2-weighted MRI. (C) The posterior epidural clot is evacuated, and postsurgical changes, namely the laminar decompression, are appreciable in the posterior spinal elements. (D) Axial image at the T8-weighted spinal levels demonstrates resolution of the posterior extradural blood clot and free cerebrospinal fluid circumferentially bathing the spinal cord.

DIFFERENTIAL DIAGNOSIS

The patient's abdominal pain in the setting of acute paraplegia prompted an immediate workup for dissection of the descending aorta that may compromise the artery of Adamkiewicz. Preservation of the dorsal column sensory modalities further supports the presumption of an ischaemic infarction of the anterior spinal artery. Transverse myelitis also presents with an acute onset of myelopathic symptoms. Epidural bleeds are always considered in acutely symptomatic patients, especially in lieu of the radiographic findings. However, the peripheral contrast enhancement of the thoracic mass in the posterior spinal canal insinuates an epidural abscess or metastatic tumour. Finally, a diffusely T1-weighted hypointense bone marrow entertains a potential haematological malignancy whose subtle indications account for its misleading diagnosis.

TREATMENT

In the setting of acute paraplegia and a likely epidural haematoma on imaging, the patient was immediately taken to the operating room within 3 h of symptom onset. The neurosurgeon performed en bloc T8–T10-weighted laminectomies (figure 1C,D). An epidural clot was identified along with a conglomeration of arterial and venous vessels. After evacuating the blood clot and coagulating the tortuous vessels, the spinal cord was adequately decompressed. Neither active pus nor cancerous

tissue was appreciated, and the surgical pathologist confirmed the diagnosis of an epidural clot and an unorganised collection of blood vessels.

OUTCOME AND FOLLOW-UP

After surgery, the patient was transferred to the neurointensive care unit. One postoperative day 7, she developed an episode of tachycardia and dyspnoea. An urgent chest CT was positive for a pulmonary embolus, and a lower extremity duplex ultrasound then localised the deep vein thrombosis to the left femoral vein. Although the threshold for diagnosing a haematological malignancy is much lower in the setting of a blood clot, the diagnosis is unlikely without haematological abnormalities on blood counts or microscopic examination of the blood smear. The patient was immediately started on an anticoagulant regimen. She was discharged to an inpatient rehabilitative facility with an unchanged physical examination—persistent weakness and sensory disturbances in the bilateral lower extremities. At 1-month follow-up over the telephone, the patient reported complete relief of her symptoms, including regaining lower extremity strength and sensation.

DISCUSSION

Vertebral haemangiomas are angiogenic tumours commonly found in the vertebral body of the spinal column. With an

Figure 2 (A) CT scan reveals a posterior epidural clot with anterior flattening (compression) of the spinal cord. (B) CT reconstruction of the thoracic aorta clearly illustrates the vessels supplying the vertebral haemangioma. (C) Prominent blush in the right hemivertebra at the T8 level concordant with haemangioma on MRI. Right and left T8 share a common trunk.

estimated prevalence of 10–12%, the vascular tumours account for 2–3% of all spinal tumours.³ Despite a strikingly high prevalence in the population, their benign character precludes adequate medical attention. Although the tumour lacks malignant or metastatic potential, physicians often overlook the possibility for growth of the haemangioma, albeit at an unappreciable growth rate.³ Nevertheless, expansion of this vascular tissue may result in grave neurological deficits, including compressive myelopathy in which patients clinically present with bilateral lower extremity symptoms consistent with upper motor neuron disease.^{4–6} The thoracic haemangiomas, in particular, have the highest propensity for extraosseous protrusion and thus symptomatic sequelae.⁷ Moreover, vascular tumours invading the posterior spinal elements are ominous for a more sinister haemangioma that may contact the neighbouring neural elements or bleed into the epidural space—both of which can directly compress the spinal cord.⁸ Some authors argue that indirect mechanisms for neurological compromise, such as compression fractures of the affected vertebra and a narrowed spinal column vis-à-vis widening of the pedicles and lamina, play a more important role in symptomatic vertebral haemangiomas.⁹ In this case presentation, although the epidural clot was identified in the posterior space, the patient symptoms were consistent with anterior spinal cord syndrome. These seemingly inconsistent findings are attributed to cord oedema and the anterior

compression of the spinal cord along the anterior spinal column. This can be seen in the sagittal MRI which reveals that the anterior cerebrospinal fluid is completely effaced while the anterior cord flattening (compression) is most appreciable on the CT images (figures 1 and 2). Moreover, a T8–T9-weighted disc herniation applies a superimposed anterior compression.

Radiographic identification ensures vascular tumours are not mistaken for their metastatic counterparts; however, the haemangiomas still warrant an adequate investigation on imaging modalities. On plain spine films, the affected vertebra characteristically exhibit vertical striations or a ‘honeycomb’ appearance. Similarly, CT demonstrates a ‘polkadot’ pattern on axial cuts of the vertebral body as well as the extent of the vertebral tumour and spinal cord compression (if present). Contrast enhancement of the haemangiomas is particularly valuable in identifying extraosseous expression.¹ The high resolution quality on MRI, on the other hand, clearly delineates the tumour boundaries. Intraosseous tumours are described as having ‘fat-signal intensity’ on both T1-weighted and T2-weighted images.^{1–10} However, posterior element invasion and extraosseous expression demonstrate T1-weighted isointensity and T2-weighted hyperintensity owing to the low-fat content and flow-void areas, both of which contribute to the lesions’ propensity to haemorrhage.¹

In an institutional review, Acosta *et al*³ proposed a treatment algorithm for the management of vertebral haemangiomas.

While asymptomatic lesions are simply observed, symptomatic tumours depends on clinical presentation coupled with radiographic findings. Symptomatic patients with lesions confined to the vertebral body are treated with percutaneous vertebroplasty or transarterial embolisation.³ Extraosseous extension, on the other hand, entails both a transarterial embolisation and vertebrectomy, as both the provocative mass and the parent tumour are addressed, respectively.¹¹ Now considered a second-line intervention, radiation therapy is reserved for patients with refractory haemangiomas.¹² Neurological deficits, namely weakness and cauda equina syndrome, warrant immediate surgical decompression of the spinal cord.¹³ When the intervention is not imminent, preoperative embolisation has been shown to reduce intraoperative blood loss.¹¹

In summary, compressive myelopathy signifies an alarming medical complication that is rarely prescribed to the insidious haemangiomas of the thoracic spine. A compromised spinal cord may have devastating consequences, especially in the face of weakness or cauda equina syndrome. Oftentimes, the workup of these symptoms exceeds the narrow window of opportunity to surgically decompress the spinal cord before permanent paraplegia ensues. Therefore, a high index of clinical suspicion is required to diagnose an active bleed from these deceitful tumours and, most importantly, to provide the best standards of care for patients affected by vertebral haemangiomas.

Learning points

- The differential diagnosis for an extradural mass in the posterior spinal canal includes abscess, haematoma and primary/metastatic tumour. Radiographic imaging will help differentiate between these conditions.
- Although the CT angiogram may be negative, further imaging should be pursued with a high index of clinical suspicion for a neurovascular injury.
- Immediate surgical intervention is required in the presence of acute neurological decline before permanent damage ensues.

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Competing interests None.

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REFERENCES

- 1 Fox MW, Onofrio BM. The natural history and management of symptomatic and asymptomatic vertebral hemangiomas. *J Neurosurg* 1993;78:36–45.
- 2 Laredo JD, Reizine D, Bard M, et al. Vertebral hemangiomas: radiologic evaluation. *Radiology* 1986;161:183–9.
- 3 Acosta FL Jr, Sanai N, Chi JH, et al. Comprehensive management of symptomatic and aggressive vertebral hemangiomas. *Neurosurg Clin N Am* 2008;19:17–29.
- 4 Krueger EG, Sobel GL, Weinstein C. Vertebral hemangioma with compression of spinal cord. *J Neurosurg* 1961;18:331–8.
- 5 McAllister VL, Kendall BE, Bull JW. Symptomatic vertebral haemangiomas. *Brain* 1975;98:71–80.
- 6 Nguyen JP, Djindjian M, Gaston A, et al. Vertebral hemangiomas presenting with neurologic symptoms. *Surg Neurol* 1987;27:391–7.
- 7 Templin CR, Stambough JB, Stambough JL. Acute spinal cord compression caused by vertebral hemangioma. *Spine J* 2004;4:595–600.
- 8 Kalina P. Spinal cord compression by thoracic vertebral hemangioma—a case report. *Open J Med Imaging* 2012;2:29–31.
- 9 Healy M, Herz DA, Pearl L. Spinal hemangiomas. *Neurosurgery* 1983;13:689–91.
- 10 Ross JS, Masaryk TJ, Modic MT, et al. Vertebral hemangiomas: MR imaging. *Radiology* 1987;165:165–9.
- 11 Acosta FL Jr, Dowd CF, Chin C, et al. Current treatment strategies and outcomes in the management of symptomatic vertebral hemangiomas. *Neurosurgery* 2006;58:287–95; discussion-95.
- 12 Faria SL, Schlupp WR, Chiminazzo H Jr. Radiotherapy in the treatment of vertebral hemangiomas. *Int J Radiat Oncol Biol Phys* 1985;11:387–90.
- 13 Baker ND, Klein MJ, Greenspan A, et al. Symptomatic vertebral hemangiomas: a report of four cases. *Skeletal Radiol* 1986;15:458–63.

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