

Sudden-onset paraplegia during pregnancy caused by haemorrhage in a spinal cord haemangioblastoma: A case report

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Abstract

Spinal cord haemangioblastomas are rare central nervous systems tumours, and haemorrhage. It is an uncommon occurrence.

We report a 28-year-old pregnant patient who presented with paraplegia due to acute haemorrhage of a spinal haemangioblastoma. Magnetic resonance imaging showed extensive syrinx cavities, an intramedullary lesion at the T4-T5 spinal cord level, and a subarachnoid haemorrhage. Digital subtraction angiography showed the feeding artery and dilated tortuous draining vein within the dural sac. The lesion was deemed a haemangioblastoma. The histopathological examination confirmed the diagnosis. Postoperatively, the paraplegia improved and the patient was able to walk within 2 weeks.

Imaging is important for early diagnosis to prevent patients persistent neurological deficits.

Keywords: Spinal cord haemangioblastoma, Haemorrhage, Paraplegia, Magnetic resonance.

Introduction

Haemangioblastomas (HMBs) are benign vascular tumours that account for about 3% of all central nervous system tumours. They primarily present in the posterior cranial fossa.^{1,2} HMBs can arise sporadically or can be associated with von Hippel-Lindau (VHL) disease. Spinal cord HMBs are rare, comprising 1.6-6.4% of all spinal cord tumours, and are usually located intramedullary.^{3,4}

Intramedullary HMBs can give rise to motor and sensitive deficits related to the tumour characteristics, such as the location, size, syringomyelia, and peripheral oedema, despite their benign histological features.⁵ Haemorrhage due to a spinal cord HMB is rare and

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generally is a subarachnoid haemorrhage.⁶

We present the case of a young pregnant patient with sudden-onset paraplegia due to acute haemorrhage involving a spinal HMB.

Case Presentation

A 28-year-old female at 30-weeks' gestation presented with rapid-onset paraplegia that occurred while sitting in an armchair. Until then, she had been well, with no neurological complaints. She reached a local hospital immediately for an initial assessment and was transported soon to our centre in April 2014. The family history did not include any evidence of VHL disease. The physical examination showed complete motor loss in her lower extremities bilaterally. Below the T5 vertebral level, sensation to pinprick was absent. There was bilateral loss of the patellar and Achilles tendon reflexes. The physical examination revealed no abnormal findings in the upper extremities.

Magnetic resonance imaging (MRI) showed multiple



Figure-1a: A post-gadolinium sagittal fat-saturated T1-weighted image showed heterogeneous enhancement of the tumour.)



Figure-1b: A T2-weighted image showed a subarachnoid haemorrhage above the T4 level.

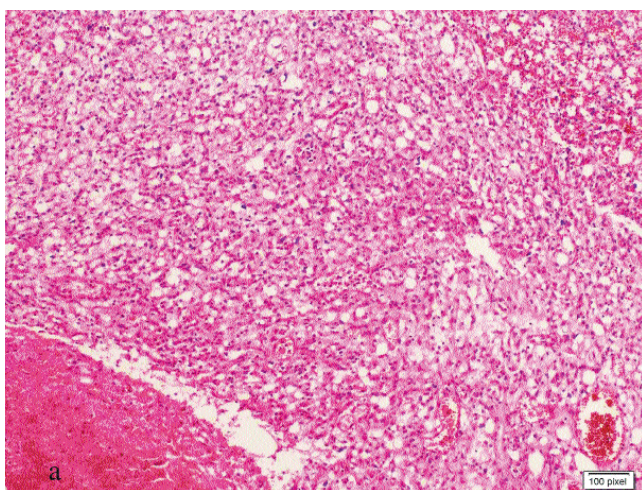


Figure-2a: Hemangioblastoma is characterized by foamy stromal cells and thin-walled capillary vessels (hematoxylin and eosin staining).

syrinx cavities from the brainstem to the lumbar spinal cord. An intramedullary lesion at the T4-T5 level was isointense on T1-weighted images and iso-hyperintense on T2-weighted images. On contrast-enhanced fat-saturated T1-weighted images, the tumour showed intense, but heterogeneous (caused by vascular flow voids), enhancement and was well demarcated from adjacent tissues (Figure-1a). On T2-weighted images, a

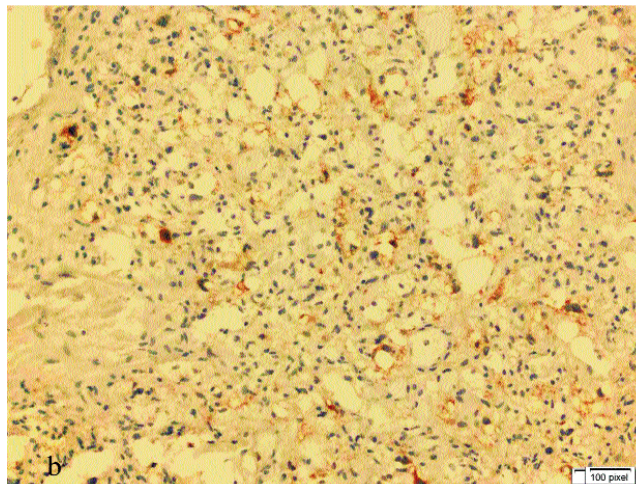


Figure-2b: Tumour cells express inhibin alpha (immunohistochemical staining).

subarachnoid haemorrhage was detected above the T4 level (Figure-1b). Based on the MRI findings, the lesion was thought to be a haemangioblastoma. To plan treatment, digital subtraction angiography (DSA) was used to assess the location of the arterial feeders and venous drainage. The highly vascularised tumour had a feeding artery branch from the right T5 intercostal artery and a dilated draining tortuous vein within the dural sac

After considering pre- or postpartum surgery, the surgeons decided that an urgent operation was required. During the surgical intervention, T4 and T5 laminectomies were performed via a posterior approach under general anaesthesia. After opening the dura and making a subsequent midline myelotomy, a highly vascularised intramedullary mass was exposed. The tumour was dissected with continuous neurophysiological monitoring of somatosensory evoked potentials (SSEPs) and subtotal tumour resection was achieved.

Histopathologically, the tumour was highly vascular and composed of blood vessels lined with endothelium with clusters of vacuolated stromal cells, called foamy cells (Figure-2a). These histological findings were compatible with HMB, which was confirmed immunohistochemically. The stromal cells expressed inhibin alpha and the diagnosis was HMB (Figure-2b).

Any possible association with VHL disease was eliminated in the postoperative clinical assessment. The patient's lower extremity paraplegia disappeared in stages after the surgery and she was able to walk unassisted after 2 weeks. Her baby was born healthy at 39-weeks' gestation.

Discussion

A haemangioblastoma is a benign vascular neoplasm of

the central nervous system. This tumour most commonly occurs in the cerebellum, while it rarely affects the spine. HMB can occur as a component of VHL disease or in isolation. The clinical findings of spinal HMBs resemble those of other spinal tumours and are based on the spinal level.^{7,8} The radiological findings are important for making a preoperative diagnosis.

Chu et al. described the characteristic MRI findings of a spinal HMB as a small superficially located, well-demarcated, intensely enhanced tumour associated with a relatively large syrinx compared with the size of the tumour.⁹ Our patient's MRI images were characteristic. There was also a subarachnoid haemorrhage above the T4 level. Spontaneous haemorrhage is a rare complication of a spinal cord HMB and can result in severe neurological deficits.¹⁰ Reports of a spinal HMB presenting with a haemorrhage describe acute paraplegia that does not recover fully after complete excision of the mass. According to Koda et al., once bleeding occurs, it can lead to paralysis that tumour extirpation cannot rescue; consequently tumour excision should be considered for an asymptomatic HMB with only slight symptoms.¹¹ Our case presented with acute paraplegia due to a subarachnoid haemorrhage. Fortunately, 2 weeks after the surgery, there were no motor or sensory deficits and she could walk. An early diagnosis and appropriate surgery can lead to recovery from a persistent neurological deficit, even in a pregnant patient.

Conclusion

Haemangioblastoma is a rare spinal benign tumor of spinal cord which could be diagnosed by radiological imaging. Early diagnosis and surgical treatment may forestall persistent neurological deficit, even in a pregnant patient.

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