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Spinal cord ependymoma presenting as acute paraplegia and subarachnoid hemorrhage: a case report and review of literature

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Abstract

Acute paraplegia is a rare presentation for a spinal cord ependymoma. Subarachnoid hemorrhage (SAH) due to spinal ependymoma is also very rare. We report a 32-year-old woman who presented with acute paraplegia and typical clinical signs of SAH with normal cerebral angiography, and further diagnostic work-up revealed an spinal cord ependymoma as the source of the hemorrhage. There is evidence that some spinal cord ependymomas have intratumoral hemorrhage, but most of these bleedings occur without symptoms. We discuss the clinical and neuroradiological findings of this rare case and review the literature related to this unusual presentation.

INTRODUCTION

Acute paraplegia is a rare presentation for a spinal cord ependymoma. There is evidence both from surgery and magnetic resonance imaging (MRI)[1] that some of the spinal ependymoma have intratumoral bleeding. However, most of these bleedings occur without symptoms. Based on Pubmed, there were only two cases of the spinal cord ependymoma presenting with acute paraplegia due to intratumoral and subarachnoid hemorrhage being reported since 1990.[2,3] In the series by McCormick *et al*., of the 23 patients with ependymoma of the filum terminale, only two suffered acute neurological deterioration. In these two cases, MRI was not performed and intratumoral hemorrhage was confirmed at surgery.[4] Subarachnoid haemorrhage (SAH) due to spinal ependymoma is also very rare. A review of the literature based on Pubmed showed that only 17 patients with spontaneous SAH due to a spinal ependymoma had been reported since 1958.[5]

We describe here a case of spontaneous intratumoral bleeding from a spinal ependymoma presented with acute paraplegia and SAH with normal cerebral angiography.

CASE REPORT

A 32-year-old woman was admitted to the Emergency Department of our Hospital because of numbness and weakness in both legs. The weakness developed 24 h prior to admission, when she felt sudden onset of waist soreness. Within 6 hours of onset, she developed numbness of the right gluteal region, then to the left side, and progressive worsening with numbness and weakness both legs. Since the 10th hour of onset, she lost all sensation, and had complete weakness below the buttocks, and had difficulty passing urine. Five hours before she reached the emergency department, she felt severe headache and presented to the Hospital with convulsive seizures. She had past history of ependymoma of the fourth ventricle, treated with surgical ablation, adjuvant radiotherapy and chemotherapy 8 years earlier. There was no history of trauma, lifting of heavy weights or fever in the last 3 months. She also did not have history of taking anticoagulants. Her mother died of brainstem hemorrhage at the age of 49 years.

She was confused and had paraplegia with neck rigidity when she arrived in the emergency room, and she was not cooperative with motor power and sensory examination. The deep tendon reflexes were present in both upper limbs, but were diminished in the lower limbs. The Babinski sign was negative bilaterally. Pulsation of the dorsalis pedis artery were equal both feet.

CT scan of the spine at the T7-S1 level showed no abnormality. CT brain showed SAH in the sulcus of the cerebellum and the occipital lobe (Figure 1). The patient immediately underwent cerebral angiography but there was no aneurysm or malformation seen.

She became alert on the 6th day after onset of symptom. Neurological examination then

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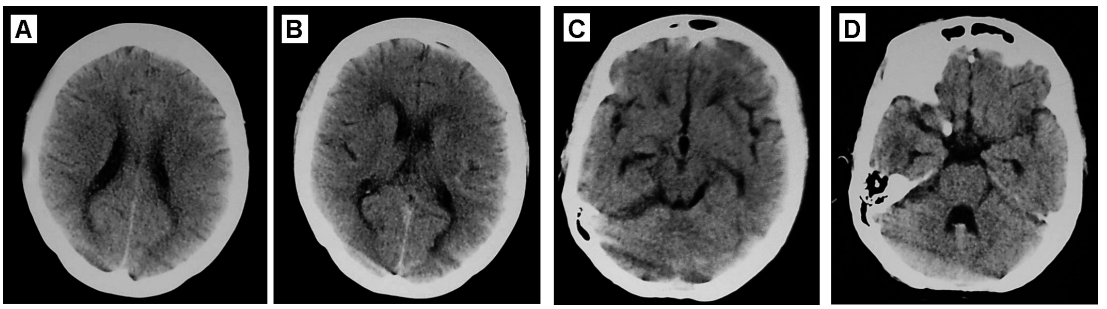

Figure 1. Brain CT scan indicated SAH in the sulcus of the occipital lobe and the cerebellum.

showed the muscle power of the lower limbs to be 0/5, and 5/5 in the upper limbs. Sensory examination showed a decrease in pain and temperature sensation below the groin. There was also impairment of the position and vibration sensation in the lower limbs. MRI of the same day showed multiple SAH lesions involving T6-T10, conus medullaris, filum terminale, and posterior cranial fossa. The spinal lesion was slightly hyperintense on T1 weighted images, hyperintense on T2 weighted images (Figure 2), and had partial enhancement following GD-DTPA administration at the spinal meninges.

The patient underwent laminectomy from T12-L5 two weeks after the onset of symptoms. On opening the dura, the spinal cord appeared enlarged. A myelotomy was done and the tumors surrounding conus medullaris and filum terminale were removed. Histology examination showed cellular ependymoma with perivascular pseudo-rosette pattern (Figure 3). Chemotherapy and radiotherapy to the spine were performed after surgery. She discharged from hospital 1 month after treatment without having any neurologic recovery of the lower limbs.

DISCUSSION

Ependymoma is the most common intramedullary tumor of the spinal cord in adults.[4] The typical clinical course is slowly progressing with pain and paresthesias. Most tumors are histologically benign with little infiltrative potential and slow growth, which is consistent with the long duration of symptoms and good long-term survival.

The vast majority of patients with acute onset of SAH are found to have intracranial lesions, most often aneurysms or arteriovenous malformations, and less often due to . brain tumours, vasculitis, and secondary to cerebral infarct.[6] SAH due to spinal lesions is very rare. Walton[6] analysed 312 patients with SAH and found the bleeding originating from intraspinal lesions in only 2, both were angiomas. Other authors report the incidence of SAH due to spinal lesion to be less than 1%[7-10], most commonly due to trauma, arteriovenous malformations, and aneurysms of spinal arteries.[11]

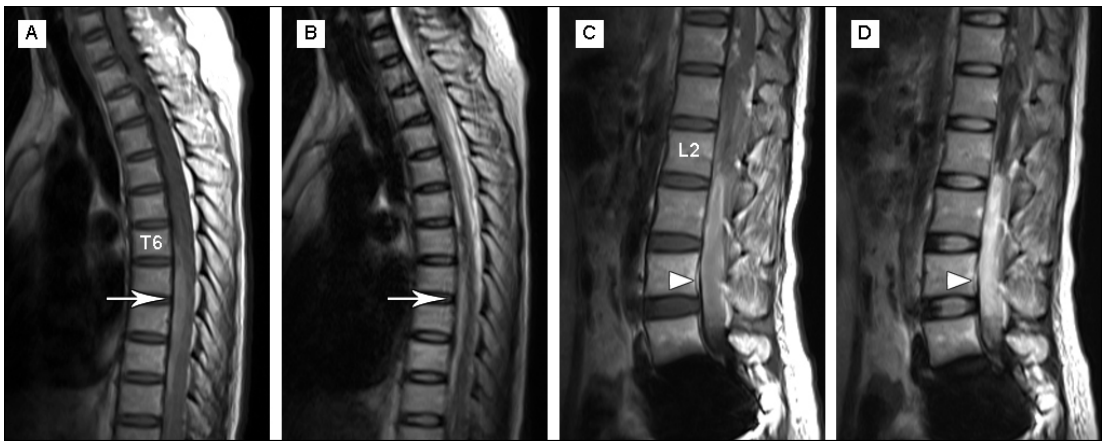

Figure 2. MRI scan showed that the lesion at T6-10 (arrow) was hyperintense on T1 (A) and T2 weighted images (B); and the lesion at conus medullaris and filum terminale (arrowhead) was of mixed intensity on T1 (C) and hyperintense on T2 weighted images (D).

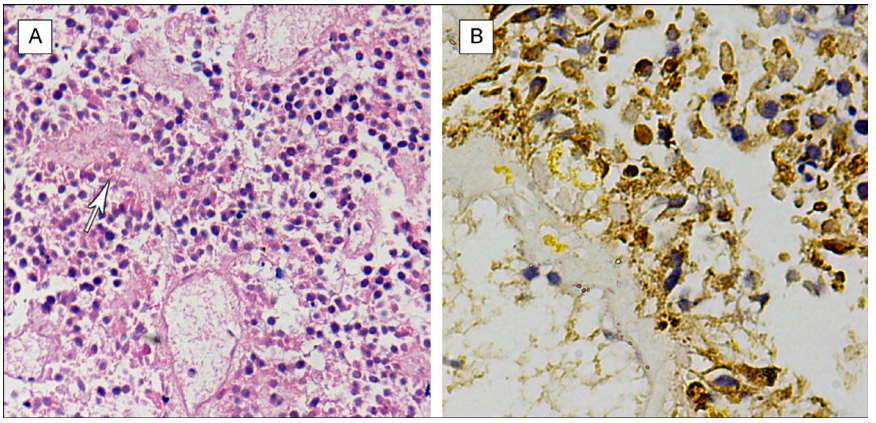

Figure 3. Spinal biopsy specimen revealing diffuse emendymoma cells (hemotoxylin-eosin,×200), the arrow indicated a perivascular pseudo-rosette pattern (A), with neoplastic cells positive for GFAP immunohistochemistry (×400) (B).

Spinal cord masses like ependymomas, nerve sheath tumours, hemangioblastomas, metastasis, meningiomas and others[4] may also cause SAH, with ependymoma being the most common (60%) pathology.[12]

Our patient presented with signs and symptoms of SAH including severe headache, neck stiffness, confusion and seizures. She also presented with paraplegia at the onset. Cranial CT was positive for blood in the sulcus of the cerebellum and occipital lobe. But the cerebral angiography as well as CT scan of the spine at the T7-S1 level showed no abnormality revealed no bleeding source. Cervical, thoracic and lumbar MRI showed SAH in T6-T10, conus medullaris and filum terminale. Combining with her history of ependymoma in the fourth ventricle 5 years ago, we suspected that the abnormal signal in MRI was probably drop metastasis of ependymoma. However, MRI was not typical of ependymoma, as these tumors usually appear as well-circumscribed masses, slight hypointense on T1, hyperintense signal on T2-weighted images with contrast enhancement.[13] The lesion deforming the spinal cord in our patient was hyperintense on T2 but slightly hyperintense on T1, did not show GD-DTPA enhancement except partial for spinal meninges at perilesional areas. It was suggestive of acute hematoma. Furthermore, unlike a spinal cord tumor, our patient was completely free of symptoms just before developing acute paraplegia. There was also no precipitating factors related to tumoral bleed such as excessive physical exercise, pregnancy, anticoagulant therapy or recent epidural anaesthesia. The spinal cord operation confirmed that it was ependymoma. Spinal ependymoma presenting as SAH is rare. Nineteen patients have been reported so far, including our patient. Our patient illustrate that spinal etiology should be considered, when the investigations for intracranial pathology is negative in patients with SAH.

The long term outcome of spinal ependymomas following surgery is generally poor.[4] The most important factor determining the long-term outcome is the preoperative neurological status.[4,14] Patients with severe preoperative neurological deficits are unlikely to make good recovery. Nevertheless, there are exceptions. Hoshimaru *et al.* reported that some of their patients with severe neurological deficits made a full recovery.[13] Most of these patients exhibited their symptoms for a short period, suggesting that a brief duration of symptoms before diagnosis may be a factor for favorable neurological outcome. Prognosis in the presence of acute spinal cord compression is also better if surgery is performed promptly. The delay before surgery in our patient may have contributed to the poor outcome.

DISCLOSURE

Conflict of interest: None

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