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Case Report

Acute neurological deterioration as a result of two synchronous hemorrhagic spinal ependymomas

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Abstract

Background: Ependymomas are the most common intramedullary tumors in adults and are the most common in mid-adult years. The presence of synchronous ependymomas in different sites of the spine is not common and it is even more infrequent to find hemorrhage from a spinal ependymoma as a cause of neurological deterioration.

Case Description: A 32-year-old man presented with back pain and progressive paraparesia. Magnetic resonance (MR) showed two intradural extramedullary lesions on spinal canal with signs of acute hemorrhage. The patient underwent emergent surgical decompression and resection. Pathology revealed myxopapillary ependymomas.

Conclusion: To our knowledge, we report the first case of a patient with acute neurological deterioration as a consequence of synchronous bleeding of two spinal ependymomas located at different levels in the spinal cord. This study illustrates the importance of recognizing the rare, but known occurrence of acute neurological deterioration after spontaneous hemorrhage in spinal ependymomas.

Key Words: Deterioration, ependymoma, hemorrhage, spine, tumor bleeding

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INTRODUCTION

Spinal cord tumors can be classified as extradural (55%) or intradural (45%), the latter being intramedullary (5%) or extramedullary (40%).[16] Ependymomas are the most common intramedullary tumors in adults and are the most common in mid-adult years. Although this pathology can occur anywhere within the Central Nervous System, nearly half of all ependymomas originate within the spinal canal. The cervical region is the most common level of true intramedullary occurrence; however, 40% occurs within filum terminale. A variety of histological ependymoma subtypes may be encountered, the myxopapillary being the most common within cauda equina or filum terminale.[8] Typical presentations for these tumors include pain, lower-extremities paresthesias, lower-extremities weakness, or bladder dysfunction.[18] Although the presence of hemorrhage on imaging or anatomopathological studies is not uncommon,[23] it is rare to see it as a cause of acute neurological deficit.[2] Several case reports have described an acute neurological decline as the result of hemorrhage in ependymoma.

We report a patient who presented after spontaneous bleeding from two separate spinal ependymomas.

CASE REPORT

A 32-year-old man presented with a history of lumbar pain without irradiation and numbness on the right leg. He was a healthy patient without any important bleeding risk factor, as anticoagulation, trauma, or heavy lifting. He was admitted to the neurosurgical department because of acute progressive paraparesia and urinary incontinence. Magnetic resonance (MR) showed two lesions on the spinal canal [Figure 1]. The higher one was located at the ninth dorsal level and was isointense on T1-weighted images, hypointense on T2-weighted images, and enhanced after contrast administration. The other lesion was located at L2–L3 lumbar level and presented similar characteristics to the former, but with hyperintense signal on T1- and T2-weighted images in its caudal part, demonstrating signs of recent hemorrhage. Due to the presence of multiple tumors, a complete craniospinal axis MR was performed including cerebral and cervical MR, with no evidence of other lesions. The patient was operated on an emergency basis and a lumbar L1–L3 and dorsal D8–D11 laminectomy was performed [Figure 2]. On opening the dura mater, blood products and clots were visualized in both levels. After removal of the hematoma, tumors were evident both on dorsal spine and cauda equina, and they were removed. Pathology revealed both tumors to be myxopapillary ependymomas [Figure 3]. The patient was discharged in 10 days after he recovered strength and the urinary function. At 2 months of follow-up, he had regained normal motor function and was able to walk unassisted. Nevertheless, the patient noticed a deficit in propioceptive sensitivity and vibration perception. The follow-up MR showed complete resection of two main lesions.

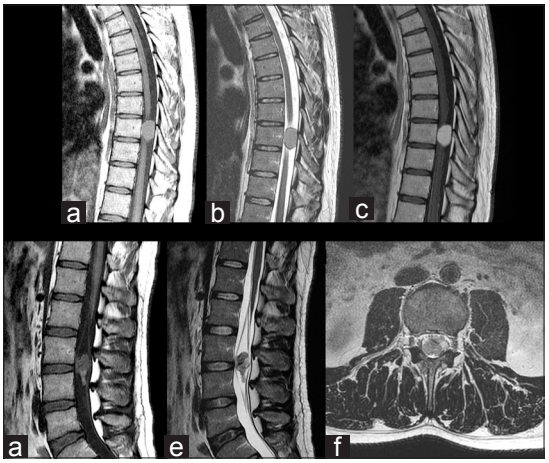

Figure 1: Preoperative MR. (a–c) Dorsal lesion. (a) T1-weighted image showing an isointense extraxial mass. (b) T2-weighted image. Isointense mass with a hyperintense ring surrounded by a better well-defined hypointense area, representing different stages of bleeding. (c) T1-weighted image with contrast administration, where the lesion enhances homogenously. (d–f) Lumbar lesion. (d) Sagittal T1-weighted image. Isointense mass viewed between cauda equina roots with a hyperintense caudal part, as a sign of acute hemorrhage. (e) Sagittal and (f) axial T2-weighted images, where the lesion is heterogeneous suggesting different stages of bleeding

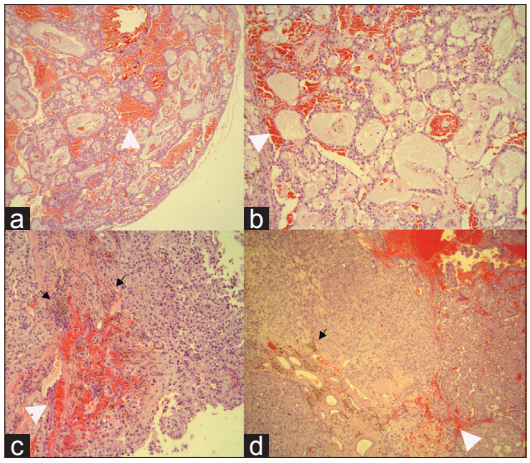

Figure 3: Microscopy (a, HE 4×; b, HE 10×). Both tumors are formed by papillary structures surrounded by a single layer of columnar cells with round nuclei and delicate chromatin. The cores of the papillae have a central blood vessel surrounded by a mucinous/myxoid matrix. There is extensive thickening and hyalinization of vessel walls. Mitotic activity is absent. Well-defined margins are present. (c, HE 10×) Dorsal and (d, HE 2.5×) lumbar lesions. Histological signs of acute and chronic bleeding can be seen within the tumor, with fresh red blood cells (white arrow) and hemosiderin-laden macrophages (black arrows)

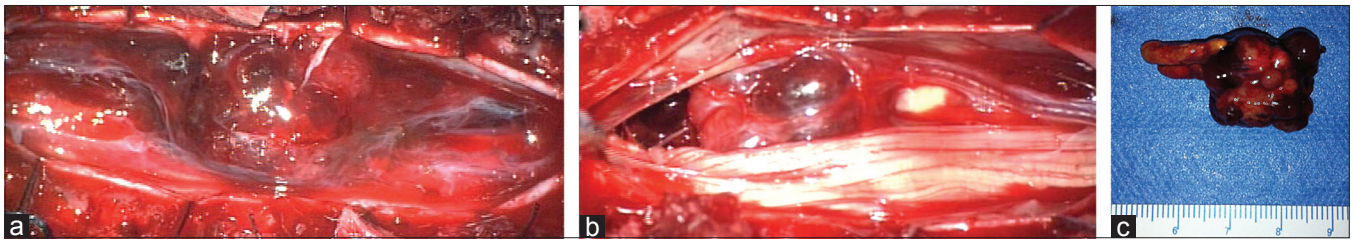

Figure 2: Intraoperative photographs: (a) Dorsal and (b) lumbar lesions were both seen as well-defined extramedullary masses after opening the dura mater, with blood products and clots in both levels. There are signs of acute and chronic hemorrhage surrounding the tumor, as can be seen in a macroscopic image after the resection of the caudal lesion (c)

DISCUSSION

Here, we report a case presenting with two synchronous spinal ependymomas, both with acute hemorrhage and resulting in spinal cord syndrome, which regained motor function after emergency decompression. Ependymoma is a histological benign and slow-growing tumor, with a mean duration of symptoms before diagnoses between 28 and 36 months.[18] There are eight reports of nine patients with acute neurological decline after hemorrhage into a spinal ependymoma[6,9,10,12,13,15,17,19] [Table 1]. Hematoma can be seen easily on pathological studies or MRI; however, this is a rare cause of acute neurological deterioration. In fact, in our case, radiological and operative findings demonstrate bleeding from two different intradural extramedullary lesions, one situated at the dorsal level and the other at the cauda equina, which represents an even more unusual feature. Although case reports of multiple ependymomas involving the caudal spinal canal have been described, ependymomas arising in this region are tipically solitary.[5] It is even more infrequent to find synchronous hemorrhage from different tumoral locations as a cause of neurological deterioration. None of the cases of acute neurological deterioration because of spinal tumor bleeding reported concomitant hemorrhage in various ependymomas located at different levels.

In our opinion, the most probable hypothesis for the presence of two concomitant ependymomas in the thoracic and cauda equina regions resulted from the

Table 1: Literature review of case reports on spinal ependymomas presenting with hemorrhage as a cause of acute neurological deterioration

	Age / Gender	Predisposing factors	First Symptom	Evolution	Clinical course	Diagnostic Imaging	Number of hemorragic lesion / Spine Level	Timing of surgery	Location / Patological diagnoses	Outcome
Desteé *et al*,[6] 1984	47 / M	Anticoagulation	Low back pain	Cauda equina syndrome	Acute	Myelography	1 / L5-S1	Delay	Ex / NR	Normal function
Herb *et al*,[7] 1990	63 / M	NR	Sciatica	Cauda equina syndrome	Acute	CT scan (1st) + myelography (2nd)	1 / L3	Emergency	Ex / NR	No improvement
Rivierez *et al*,[12] 1990	18 / F	NR	Sciatica	Cauda equina syndrome	NR	Myelography	1 / L1-L5	NR	NR / NR	NR
Malbraine *et al*,[13] 1994	65 / F	Anticoagulation	Low back pain	Cauda equina syndrome	Acute	Myelography (1st) + CT scan (2nd) + MR (after surgery)	1 / L2-L3	Emergency	Ex / MP	No improvement
Lagares *et al*,[9] 2000	24 / M	NR	Low back pain	Paraplegia	Acute	MR	1 / Low dorsal	Emergency	In / II	Walking with aid
Oertel *et al*,[10] 2000	35 / M	NR	Paraplegia	Paraplegia + sensory loss + sphyncter dys	Acute	MR	1 / D9-D11	Emergency	In / II	Walking without aid
Tait *et al*,[11] 2004	57 / F	Anticoagulation	Cauda equina syndrome		Acute	MR	1 / L3	Emergency	Ex / II	Walking without aid
Heuer *et al*,[8] 2007	31 / F	None	Low back pain	Monoparesia + Sphyncter dys	Acute	MR	1 / L1-S2	Delay 1 month	Ex / MP	Normal function
	31 / M	Heavy Lifting	Low back pain	Paraparesia	Acute	MR	1 / D11-L2	Delay 1 week	Ex / MP	Normal function
Present study, 2011	32 / M	None	Low back pain	Progressive paraparesia	Acute	MR	2/ D9 & L2-L3	Emergency	Ex / MP (both lesions)	Walking without aid

M: Male, F: Female, NR: Not reported;, dys: dysfunction, Ex: Extramedullar, In: Intramedullar, MP: Myxopapillary ependymoma, II: WHO Grade II ependymoma (papillary; tanycytic; cellular variants...)

implant of neoplastic cells from a primary filum terminale ependymoma, following the physiologic ascending spinal CSF flow. The same mechanism could account for rare cases of intracranial metastasis,[4,14] sometimes occurring even long time after surgical removal.[1] The detection of the multiplicity of ependymomas is important due to the possible deterioration of the patient. If a dural opening is made in the presence of an intracranial mass, the patient may deteriorate because of cerebral herniation. For this reason, a brain and entire craniospinal axis MR should be performed before performing spinal laminectomy, if multiple ependymomas are detected or CSF seeding is suspected. The reported case showed no intracranial tumors or lesions in the cervical spine in the craniospinal MR done before surgery.

Nevertheless, it is not possible to be sure if multiplicity of these lesions is due to CSF spread or multicentric foci. Genetic analysis of the tumor samples is the only way to know with certainty the mechanism of multiple ependymomas. Vural *et al.*[21] used conventional cytogenetics, multiplex fluorescence *in situ* hybridization (M-FISH), interphase-FISH specific to 22q11, and epidermal growth factor receptor loci analyses of the tumor samples to confirm the clonal origin of ependymomas located at different spinal levels. Unfortunately, these techniques could not be used in our center. An intradural extramedullary location is rare for ependymomas.[11] Though for anatomical and surgical reasons, ependymomas within the filum terminale are considered extramedullary tumors.[16]

Few theories have been described to explain the specific predisposition to bleeding in these tumors, related to their peculiar anatomical location and histological features. Relative to the rostral lesion, abnormal mobility in the thoracolumbar region plays an important role in the development of hemorrhage in tumors situated in this location.[20,22] In relation to the mass located at the cauda equina region, myxopapillary ependymomas are the most prone to bleeding because of the intense physical stress they support in the caudal region and the unique vascular architecture of this subtype of ependymoma.[7]

The effect of early surgery on final outcome remains unclear. In eight patients, the timing of surgery was reported. Five of eight cases and our present case underwent emergency surgical descompression and resection. Only three of these and the patient reported here showed improvement in their neurological condition. In three of eight cases, the timing of operation was delayed, and all had good motor and blader function recovery. Though series of surgical management of spinal ependymomas has previously been published, early diagnosis and treatment were associated with a more favorable outcome.[18] Furthermore, symptoms of sphincter dysfunction were associated with a poor outcome.[18] Moreover, in the presence of acute cord compression, prognosis is better if surgical descompression is performed promptly, as ocurred in our patient who made a good recovery.

It has been widely held that myxopapillary ependymoma has a better prognosis than other variants. Nevertheless, new series of cases show that prognosis after surgery for some myxopapillary ependymomas seems worse than generally believed.[3]

CONCLUSION

This report shows the importance of recognizing spontaneous bleeding of a spinal ependymoma as a possible cause of rapid neurological deterioration and acute cord compression, even in the absence of bleeding risk factors such as trauma or anticoagulation. It is also the first case description of synchronous bleeding from spinal ependymomas of different levels.

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