

CASE REPORT

Spontaneous Rupture and Hemorrhage of Myxopapillary Ependymoma of the Filum Terminale : A Case Report and Literature Review

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Abstract : We present a rare case of acute onset cauda equina syndrome caused by a ruptured myxopapillary ependymoma with accompanying hemorrhage. A 26-year-old healthy woman developed muscle weakness and sensory disturbances in her bilateral lower extremities. Magnetic resonance imaging showed a huge mass from the L1 body to the L2-3 disc level. She was able to ambulate with crutches after the tumor was successfully removed. To prevent recurrence, she received whole brain and spinal cord radiation. No sign of recurrence were detected at the 8 month follow up. *J. Med. Invest.* 61 : 430-435, August, 2014

Keywords : *spinal myxopapillary ependymoma, hemorrhage, rupture, acute neurological deterioration, magnetic resonance imaging*

INTRODUCTION

Myxopapillary ependymoma is defined as a slow-growing, glial-derived tumor that arises from ependymal cells within the central nervous system. Its preferential manifestation is in young adults with an annual incidence of less than 0.01 per 100,000 (1-4). The majority of myxopapillary ependymomas are found in the conus medullaris or filum terminale (5, 6) and tend to have an insidious presentation with a gradual onset of symptoms and prolonged history (7, 8). Therefore, it is rare for myxopapillary ependymoma to present with acute neurological deterioration secondary to rupture and hemorrhage. To the best of our knowledge, only 12 reports of acute neurological deterioration caused by ruptured

myxopapillary ependymoma with accompanying hemorrhage have been published in the English literature.

In this report, we present a case of acute onset cauda equina syndrome caused by a ruptured myxopapillary ependymoma with accompanying hemorrhage.

CASE REPORT

A 26-year-old healthy woman with no history of major trauma or coagulation disorders presented with acute low back pain. During the following 3 days, she developed muscle weakness and sensory disturbances in her bilateral lower extremities. A diagnosis of paraparesis secondary to cauda equina tumor was made by a local physician prior to her referral to our department for surgical treatment. Muscle strength of the right iliopsoas, quadriceps femoris, tibialis anterior (TA), extensor hallucis longus (EHL), and triceps surae (TS) was reduced

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to 0/5 on manual muscle testing (MMT), and that of the left was reduced to 1/5. Loss of pin-prick and vibration sensation, severe reduction of light touch sensation, and urinary dysfunction were found. Anal tone was lax.

Magnetic resonance imaging (MRI) revealed an intradural tumorous mass extending from the L1 body to the L2-3 disc level, severely compressing the epiconus and conus level of the spinal cord (Fig. 1A-D). These findings were suggestive of bleeding from the tumorous mass secondary to capsule rupture. Notably, the hemorrhage extended from the cranial end of the tumor to the T10-11 disc level, and a finding similar to drop coagulation in the bottom of the dural sac was detected.

Surgery was performed via the dorsal midline

approach. After L1-2 laminectomy with durotomy, xanthochromic fluid was observed in the subarachnoid space. Blood clots and hematoma protruded from the ruptured capsule (Fig. 2A). After removing the blood clots and hematoma, a reddish-gray, soft, pliable tumor with attachment to the filum terminale was found among the cauda equina (Fig. 2B). The conus and cauda equina were compressed by the tumor to the ventral side. The granular tumor arising from the filum terminale was excised (Fig. 2C).

Histological examination of the tumor revealed typical features of a myxopapillary ependymoma with fresh blood cells : marked mucous changes of the stroma and papillary arrangement of tumor cells (Fig. 3A-C). The tumor cells were also seen within the hematoma (Fig. 3D). The glial cells were

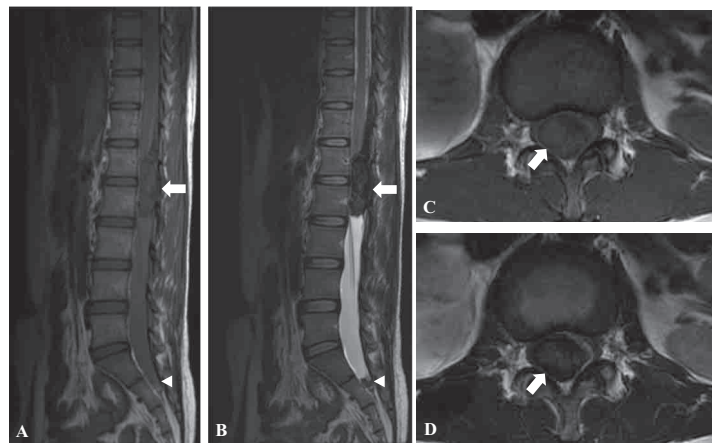


Fig. 1. Preoperative magnetic resonance imaging (MRI) (A, B) Sagittal images showing a huge mass from the L1 body to the L2-3 disc level (arrows). The lesion was heterogeneous isointense on T1-weighted images and had low intensity on T2-weighted images. The hemorrhage extends from the cranial end of the tumor to the T10-11 level, and drop coagulation in the bottom of the dural sac is visible (arrowheads). (C, D) Axial images showing a huge mass at L1-2 displacing the spinal cord (arrows).

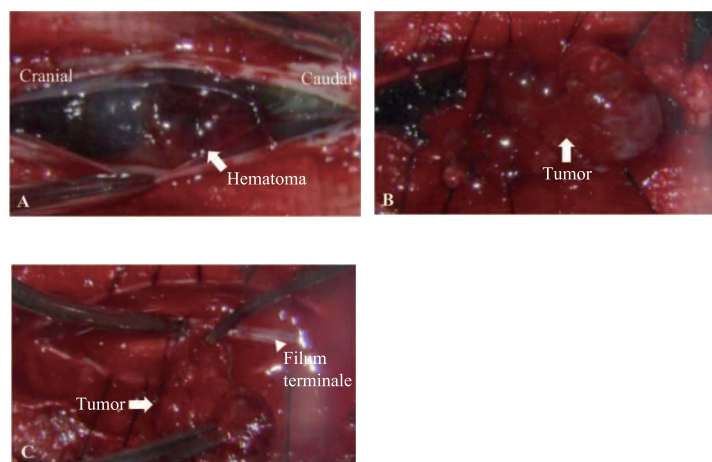


Fig. 2. Intraoperative findings (A) Opening of the dura reveals an extensive blood clot intermingled with cauda equine (arrow : hematoma). (B) The mass consists of solid blood clots and reddish-gray tumor (arrow : tumor). (C) Tumor originating from the filum terminale (arrow : tumor, arrowhead : filum terminale).

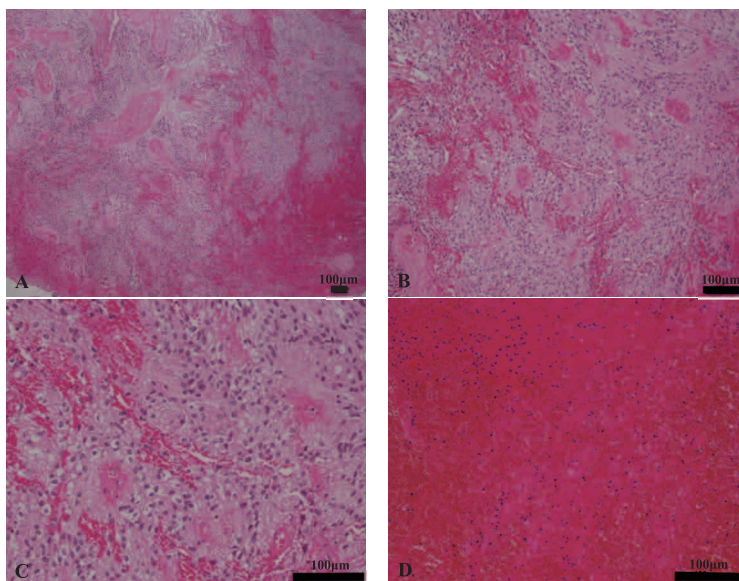


Fig. 3. Pathological analysis

(A-C) Pathological evaluation after resection of the mass resulted in a diagnosis of hemorrhage from a myxopapillary ependymoma. Cuboidal to elongated tumor cells are radially arranged in a papillary manner or like pseudorosettes around vascularized stromal cores with no mitotic activity. Hematoxylin and eosin staining ; magnification for A, B, and C : $\times 4$, $\times 10$, and $\times 20$, respectively. (D) Tumor cells are apparent within the hematoma. Hematoxylin and eosin staining ; Magnification : $\times 20$.

strongly positive for glial fibrillary acidic protein, which was consistent with a diagnosis of myxopapillary ependymoma. The Ki67 labeling index was around 1%.

Immediately after the surgery, the patient's low back pain, bladder dysfunction, and sensory disturbance of the bilateral lower extremities resolved. Her bilateral lower extremity strength on the MMT returned to 5/5, except for the right TA (1/5) and

right EHL (1/5), and she was able to ambulate with crutches.

To prevent recurrence, she received whole brain and spinal cord radiation (45 Gy). Postoperative MRI at the 8 month follow up revealed complete resection of the tumor and hematoma, as well as the disappearance of the drop coagulation in the bottom of the dural sac (Fig. 4A, B).

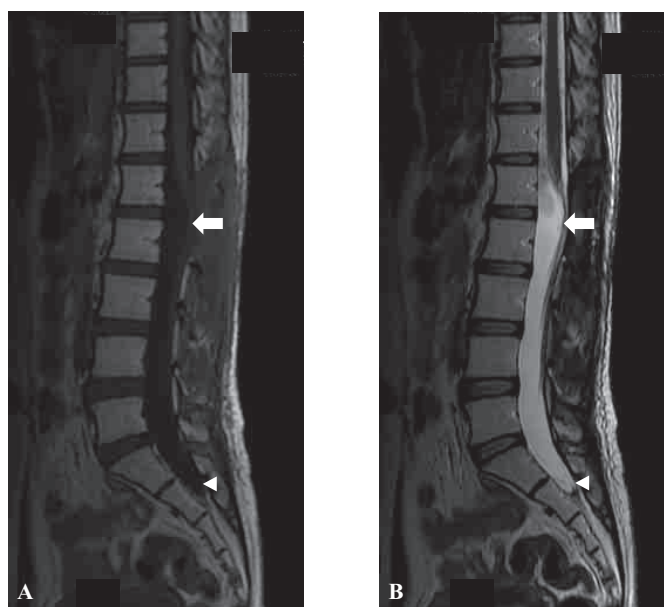


Fig. 4. Postoperative MRI

(A, B) Sagittal images showing disappearance of drop coagulation in the bottom of the dural sac (arrowheads), and no signs of tumor recurrence 8 months after the operation (arrows).

DISCUSSION

Myxopapillary ependymoma is defined as a slow-growing glioma and is considered a distinct variant of ependymomas classified as World Health Organization grade 1 (1-4). More than 50% of myxopapillary ependymomas are located in the conus medullaris or filum terminale (5, 6), and the majority of myxopapillary ependymomas exhibit an insidious presentation with a gradual onset of symptoms and a prolonged subjective history, although clinical manifestations are frequently nonspecific (7, 8). Therefore, it is rare for a patient with myxopapillary ependymoma to present with acute neurological decline after spontaneous hemorrhage, even if the myxopapillary ependymoma has a more vascular architecture than the other types of ependymomas (9-11).

We presented here a rare case of a 26-year-old woman with a ruptured myxopapillary ependymoma and accompanying hemorrhage that expanded from the L1 body to the L2-3 disc level. A review of the English literature revealed only 12 reports describing 15 cases of acute neurological decline caused by hemorrhage from myxopapillary ependymoma of the filum terminale or conus medullaris (Table 1) (7, 8, 12-21). Clear descriptions of the tumors or MRI findings were documented in only 8 of these cases. All 8 and the present case showed heterogeneous

lesions, although the presentation may have been affected by the phase of the hemorrhage from the tumor. Furthermore, drop coagulation in the bottom of the dural sac indicating dissemination of the tumor cells was detected in 4 patients.

Myxopapillary ependymoma tends to disseminate and recur despite its benign histologic character (22). The role of radiotherapy is controversial, but some authors proposed that radiation to the whole brain and spinal cord (including local radiation ; total dose, 30-50 Gy) prevented further tumor progression (23, 24). Nakamura *et al.* suggested that the surgical margins obtained in the initial surgery and the extent and amount of postoperative radiation could be crucial factors in determining the prognosis of patients with myxopapillary ependymoma because cerebrospinal dissemination can occur once the tumor capsule is violated before or during surgery (24). In the present case, radiotherapy was performed because tumor cells were detected within the hemorrhage. Although the long-term outcome is unclear as yet, postoperative radiotherapy should be considered in such cases.

CONFLICT OF INTEREST

There are no conflicts of interest to disclose.

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

Authors and year	Age (years)	Sex	Level of tumor localization	T1 intensity	T2 intensity	Tumor homogeneity	Drop coagulation in the dural sac end
Djindjian M <i>et al.</i> , 1978	18	M	L2	N/A	N/A		
	34	F	L3	N/A	N/A		
Destee A <i>et al.</i> , 1984	47	F	L5-S1	N/A	N/A		
Gomori JM <i>et al.</i> , 1985	32	M	N/A	N/A	N/A		
Rappaport B <i>et al.</i> , 1990	21	M	L1	N/A	N/A		
Admiraal P <i>et al.</i> , 1992	23	M	L1-2	N/A	iso	N/A	N/A
Malbrain ML <i>et al.</i> , 1994	65	F	L2-3	N/A	N/A		
Argyropoulou PI <i>et al.</i> , 2001	15	F	L1-L4	iso	iso	heterogeneous	+
Saadah M <i>et al.</i> , 2004	33	M	L1-3	N/A	N/A	N/A	N/A
	48	M	T12-L4	N/A	N/A	heterogeneous	N/A
Heuer GC <i>et al.</i> , 2007	31	F	L1-L2	low	low	heterogeneous	+
	31	M	T11-L2	N/A	N/A	heterogeneous	N/A
Becco de Souza R <i>et al.</i> , 2012	13	F	L1-3	iso-low	iso-high	heterogeneous	+
Martinez-Perez R <i>et al.</i> , 2012	32	M	L2-L3	iso	low	heterogeneous	N/A
Khalatbari MR <i>et al.</i> , 2014	15	M	L1	high	low-iso	heterogeneous	-
Present case	26	F	L1-2	iso	low	heterogeneous	+

M : male ; F : female ; N/A : not applicable

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