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CASE REPORT

Surgical treatment for posttraumatic hemorrhage inside a filum terminale myxopapillary ependymoma: a case report and literature review

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Abstract
Purpose Symptoms of cauda equina syndrome due to ependymoma in the conus medullaris or filum terminale develop slowly. However, hemorrhagic change inside spinal tumors can induce acute neurologic decline. Here, we report a case of posttraumatic hemorrhage inside a filum terminale myxopapillary ependymoma presenting as acute neurologic decline, which had a positive prognosis after surgical resection.
Methods A 28-year-old man presented with buttock pain, sensory disturbance, and motor weakness of bilateral lower extremities after falling on ice during smelt fishing. Magnetic resonance imaging demonstrated a mixed-intensity hemorrhagic intradural mass extending from L1 to L2.
Results The patient underwent emergent surgical decompression and resection. Pathologic examination revealed a myxopapillary ependymoma with intratumoral hemorrhage. After surgery, the patient demonstrated gradual improvement in neurologic deficits and no tumor recurrence.
Conclusions This is the first case of a filum terminale myxopapillary ependymoma with an acute neurologic decline after injury. Early diagnosis and treatment are associated with favorable outcomes.

Keywords Hemorrhage · Cauda equina · Ependymoma · Injury

Introduction

Myxopapillary ependymomas are not uncommon, accounting for approximately 1–5 % of all spinal neoplasms, and have a prevalence of 0.0–0.08 cases per 100,000 persons annually [1, 2]. Over half of these ependymomas are located in the conus medullaris or filum terminale [3, 4]. Generally, symptoms of ependymoma develop slowly. However, intratumoral hemorrhage can induce an acute neurologic decline. Here, we report a case of posttraumatic hemorrhage inside a filum terminale myxopapillary ependymoma, which had a positive prognosis after surgical resection.

Case report

A 28-year-old man presented with buttock pain, sensory disturbance, and motor weakness of bilateral lower extremities, which occurred several seconds after falling on ice during smelt fishing. The patient had no history or current usage of anticoagulation or antiplatelet agents. Motor weakness of the lower extremities progressed, and he was transferred and admitted to our hospital via another emergency hospital. On admission, neurologic examination revealed hypoesthesia of the anterior aspect of bilateral

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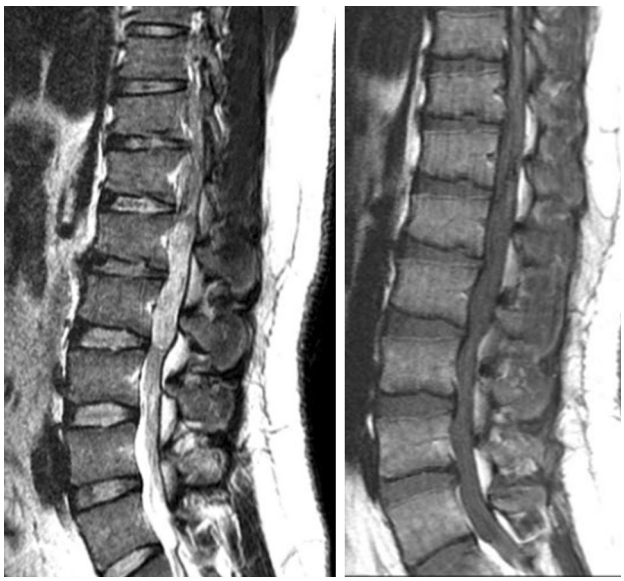

Fig. 1 Magnetic resonance images, showing a hemorrhagic intradural mass extending from L1 to L2, with iso signal intensity on T1-weighted imaging (*right*) and mixed signal intensity on T2-weighted imaging (*left*). This hemorrhagic intradural mass occupied the whole spinal canal and compressed the cauda equina strongly

thighs, anesthesia of the entire lower thigh, complete paresis of bilateral lower extremities, disappearance of the anal reflex, and urinary retention. All laboratory test results, including coagulative parameters, were within normal range. Magnetic resonance imaging (MRI) demonstrated a mixed-intensity hemorrhagic intradural mass extending from L1 to L2 (Fig. 1). During MRI, the patient could not maintain a resting supine position due to severe buttock pain; therefore, only minimal sequences without gadolinium enhancement were performed.

The patient was taken to the operating room 10 h after injury. After laminectomy from T12 to L3 and opening of the dura mater, a wine-colored tumor was visualized (Fig. 2a). The cauda equina was compressed ventrally by the tumor, and the conus medullaris could not be observed because of the tumor. A blood clot was localized in the thin tumor capsule without perforation into the subarachnoid space. Initially, internal decompression of the tumor was performed using an ultrasonic surgical aspirator. During a subsequent tumor removal, meticulous care was taken to preserve the tumor capsule without the tumor substance falling into the subarachnoid space in order to avoid postoperative dissemination. There was no adhesion between the tumor wall and cauda equina. The filum terminale attached to the tumor was resected at the rostral and caudal portion, resulting in gross total tumor resection (Figs. 2b, 3). No attachment between the tumor and the conus medullaris was confirmed during and after the tumor removal.

The day after surgery, the patient's motor weakness and dysesthesia of bilateral lower extremities showed slight improvement. Histopathologically, the tumor was consistent with myxopapillary ependymoma (Fig. 4a–d). Histopathologic examination revealed an encapsulated tumor composed of numerous papillary villous structures of varying size and shape, with prominent intervillous fresh hemorrhage. Each villous structure was characterized by an abundant perivascular myxoid stroma that was lined by glial tumor cells with oval-to-spindle-shaped nuclei and fibrillary cytoplasmic processes. Nuclear pleomorphism was observed in the tumor cells, but no mitotic figures were seen. Immunohistochemically, the tumor cells were positive for glial fibrillary acidic protein (GFAP) and negative for CAM5.2; furthermore, Ki-67 labeling index was less than 1 %. These histologic features and immunohistochemical profile are pathognomonic of myxopapillary ependymoma.

Postoperative adjuvant chemotherapy and radiation therapy were not performed because complete tumor

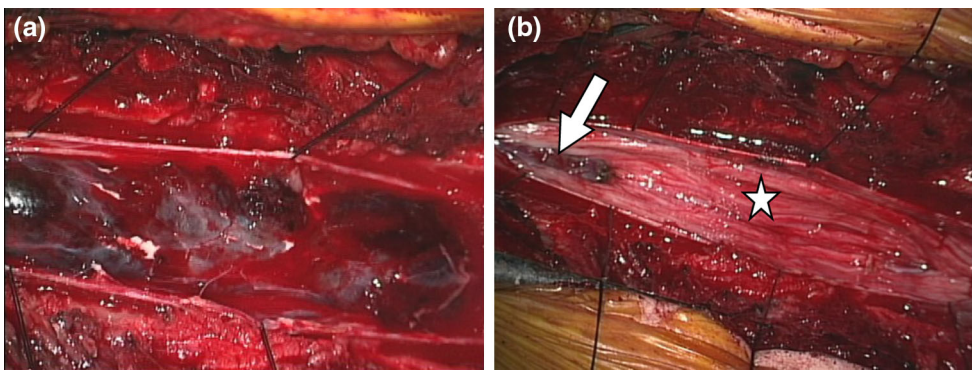

Fig. 2 **a** Intraoperative microscopic photograph, showing an expansive tumor with intratumoral hemorrhage (*arrowheads*), and a blood clot localized in the thin tumor capsule without perforation into the subarachnoid space. **b** Intraoperative microscopic photograph, showing the cauda equina after total tumor resection. The *arrow* indicates the proximal stump of the filum terminale resected from the tumor, while the *star* indicates the cauda equina

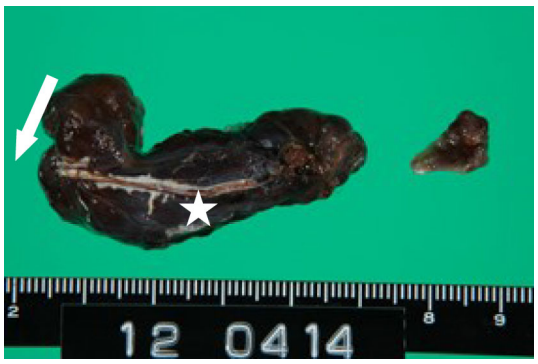

Fig. 3 Photograph, showing the resected tumor with intratumoral hemorrhagic change

resection was confirmed on intraoperative observation and postoperative MRI. At the 4-week follow-up, the patient-â€™s motor weakness showed gradual improvement, and he was ambulating with a crutch. Additionally, his bladder dysfunction had recovered completely. After 4 years, there was no tumor recurrence and symptom exacerbation did not persist.

Discussion

Of all the spinal neoplasms, ependymomas might bleed during their natural clinical course; however, most cases involve subclinical hemorrhage [5, 6]. There have been only 14 cases of acute deterioration of conus medullaris or cauda equina ependymoma with intratumoral hemorrhage, including the present case (Table 1) [5, 7–17]. In these cases, the predisposing factors were spontaneous (8 cases), anticoagulation (1 case), heavy lifting (3 cases), and trauma (2 cases). The tumor subtypes were myxopapillary (7 cases), cellular variant (3 cases), World Health Organization (WHO) grade 2 (2 cases), WHO grade 3 (1 case), and in 1 case, the subtype was not reported. In a previous trauma case, a WHO grade 3 tumor was located in the filum terminale [15]. To the best of our knowledge, this is the first case of a myxopapillary ependymoma in the filum terminale presenting with posttraumatic intratumoral hemorrhage.

The mechanical and histopathologic factors promoting hemorrhage inside a spinal ependymoma have been described [15, 18]. Mechanically, conus medullaris and filum terminale lesions are located at a highly mobile segment of the spine, and the traction forces might cause disruption of blood vessels on the surface of the tumor. Histopathologic factors relate to the presence of numerous small blood vessels and loss of connective tissues in the tumor. Additionally, in our case, trauma might induce significant venous hypertension following elevation of intrathoracic pressure, as in the Valsalva maneuver. These various factors might lead to rupture of small, thin-walled blood vessels in the tumor, and induce intratumoral hemorrhage.

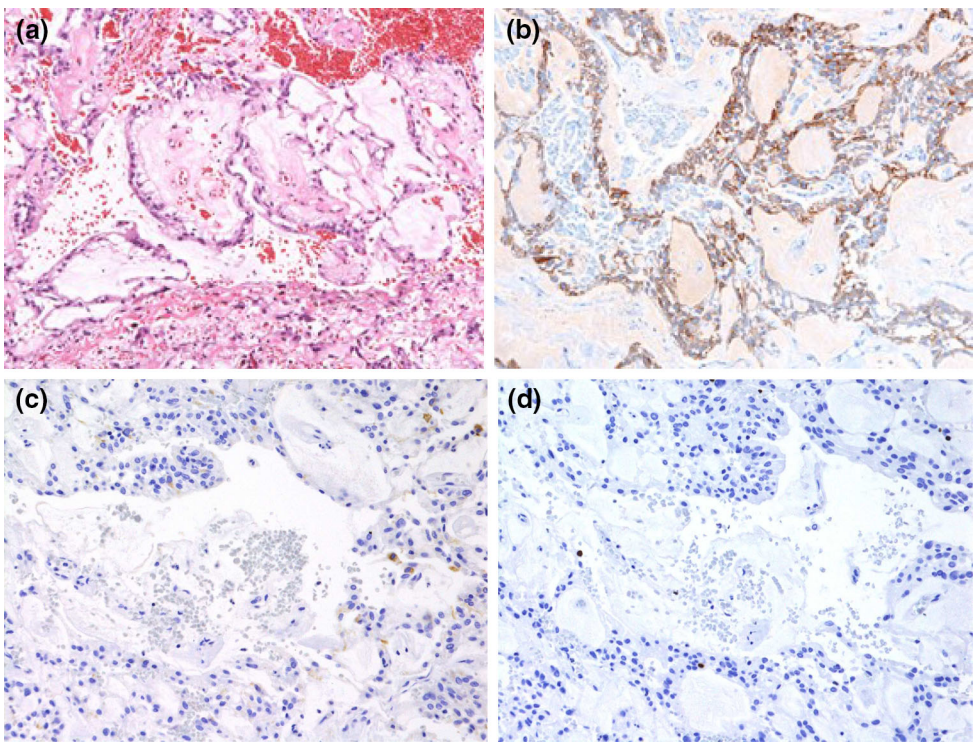

Fig. 4 Histopathologic images, demonstrating hemorrhagic change and papillary-like tumor cell lining around a myxoid connective tissue stroma with small vessels. **a**, **b** Glial fibrillary acidic protein expression is positive (hematoxylin and eosin stain, magnification × 20). **c** CAM5.2 expression is negative (CAM5.2 stain, magnification × 20). **d** Ki-67 labeling index is less than 1 % (Ki-67 stain, magnification ×20)

Table 1 Cases of conus medullaris or cauda equina ependymoma with hemorrhage

Case report	Age/sex	Predisposing factors	Symptoms	Clinical course	Timing of surgery	Location	Subtype	Removal	Motor recovery Bladder function
Herb [9]	63/M	Spontaneous	Paraparesis, BD, pain	Acute	Emergency	L3	NR	NR	No improvement No improvement
Lagares [12]	24/M	Spontaneous	Paraplegia	Acute	Emergency	Lower lumbar	Grade 2	Total	Ambulating w/crutch NR
Oertel [14]	35/M	Spontaneous	Paraplegia, SD, pain	Acute	Emergency	Th9–11	Grade 2	Total	Ambulating w/o aid Normal
Turgut [5]	51/M	Spontaneous	Paraparesis, BD, SD, pain	Acute	Emergency	L4–5	Cellular	Total	Improvement Improvement
Heuer [10]	31/F	Spontaneous	Paraparesis, BD, pain	Acute	Delay 1 month	L1–3	MP	Total	Ambulating w/o aid Normal
Martinez-Perez [13]	32/M	Spontaneous	Paraparesis, BD, pain	Acute	Emergency	Th9/L2–3	MP	Total	Normal Normal
Souza [8]	13/M	Spontaneous	Paraparesis, BD, pain	6 days	Emergency	L1–3	MP	Total	Ambulating w/aid NR
Tonogai [17]	26/F	Spontaneous	Paraplegia, BD, SD, pain	Acute	Emergency	L1–2	MP	Total	Ambulating w/crutch
Admiraal [7]	23/M	Heavy lifting	Pain	2 weeks	NR	L1–2	Cellular	Total	Normal Normal
Heuer [10]	31/M	Heavy lifting	Paraparesis, BD, pain	Acute	Delay 1 week	Th11–L2	MP	Total	Ambulating w/o aid Normal
Khalatbari [11]	15/M	Heavy lifting	Paraparesis, BD, pain	Acute	Emergency	Th12–L1	MP	Total	Normal Normal
Tait [16]	57/M	Anticoagulation	Paraparesis, BD, pain	Acute	Emergency	L3	Cellular	Total	Ambulating w/o aid Abnormal
Ozdemir [15]	62/M	Trauma	Paraparesis, SD, pain	Acute	Emergency	L1	Grade 3	Total	Normal Normal
Present case	28/M	Trauma	Paraplegia, BD, SD, pain	Acute	Emergency	L1–2	MP	Total	Ambulating w/crutch Normal

BD bladder dysfunction, *F* female, *M* male, *MP* myxopapillary ependymoma, *NR* not reported, *SD* sphincter dysfunction, *w/*with, *w/o* without

Clinically, cauda equina syndrome can present as low back pain, sciatic pain, saddle anesthesia, decreased rectal tone, perineal reflex, bowel and bladder dysfunction, and lower extremity weakness. Ependymomas in the distal spine are almost entirely of the myxopapillary subtype. Generally, these clinical symptoms gradually worsen with a prolonged history, and acute neurologic decline is rare [19]. During an acute-onset cauda equina syndrome after trauma, as in the present case, MRI should be performed immediately to detect intraspinal hemorrhage. This is especially necessary if radiography and computed tomography do not reveal traumatic findings, such as thoracolumbar fracture, which could induce symptoms. Neurinomas in the conus medullaris or filum terminale, which are typically intradural extramedullary tumors, could cause intratumoral hemorrhage, facilitating an acute-onset cauda equina syndrome [20]. Ependymomas and neurinomas might be indistinguishable based on imaging features alone, especially when neurinomas present with bleeding. In this situation, diagnosis can be made based on the intraoperative and pathologic findings. In addition, apart from these spinal tumors, spinal

arteriovenous malformation or intramedullary tumor should be considered in the differential diagnosis of intraspinal hematoma. In fact, in the present case, we could not diagnose the patientâ€™s intratumoral hemorrhage based on MRI findings before the operation. However, diagnosis could be made based on the intraoperative findings. Therefore, during the surgical treatment for hematoma around the conus medullaris, we should consider several causes for the hematoma; myxopapillary ependymoma should be considered even if the patient has no history of this type of tumor and preoperative imaging does not reveal a tumor.

In cases of intratumoral hemorrhage in the conus medullaris or cauda equina, emergent surgery provides positive clinical outcomes [5, 7–17]. In fact, in 2 trauma cases, including the present case, emergent surgery facilitated recovery. The tumors did not adhere to the cauda equina, and thus, could be totally resected. Surgical indication for conus medullaris hemorrhage depends on neurologic findings because it is difficult to quickly diagnose intratumoral hemorrhage. Based on the present case and a review of previous cases, early diagnosis and surgery may be associated with more favorable outcomes regardless of predisposing factors or tumor subtype.

The main histologic differential diagnosis is metastatic mucin-producing (papillary) adenocarcinoma. However, no mitotic activity (and extremely low Ki-67 labeling index), lack of significant atypia, GFAP immunopositivity, and CAM5.2 immunonegativity virtually excludes adenocarcinoma. It should be noted that pancytokeratin, especially AE1/AE3, immunostaining is known to be positive in glial tumors, including myxopapillary ependymomas, as seen in this case. This finding is explained by cross immunoreactivity between GFAP and cytokeratin, both of which are intermediate filaments of cells.

Myxopapillary ependymomas are histologically classified as WHO grade 1. During surgery, tumor resection can be safely achieved if the tumor is encapsulated. However, as tumor grows, tumor encapsulation can be lost and adherence to cauda equina can inhibit complete resection and induce new deficits after surgery [21]. They can recur following incomplete resection of large unencapsulated tumors or complete resection with capsular violation, in which the tumor substance falls into the subarachnoid space, resulting in cerebral spinal fluid dissemination [22, 23]. In these situations, postoperative adjuvant radiotherapy may be considered [24, 25]. Therefore, during tumor removal, meticulous care should be taken to preserve the tumor capsule, and follow-up MRI is necessary to detect recurrence even if the tumor is completely resected.

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Compliance with ethical standards

Conflict of interest The authors report no conflicts of interest concerning the materials or methods used in this study or the findings specified in this paper.

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