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Atypical Imaging of Hemorrhagic Lumbosacral Myxopapillary Ependymoma with Histopathological Correlation: A Case Report

Authors' Contribution:

Study Design A
Data Collection B
Statistical Analysis C
Data Interpretation D
Manuscript Preparation E
Literature Search F
Funds Collection G

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Conflict of interest: None declared

Patient: Male, 16-year-old
Final Diagnosis: Hemorrhagic myxopapillary ependymoma • myxopapillary ependymoma
Symptoms: Gait abnormality • pain in lumbar region
Medication: —
Clinical Procedure: Resection of intraspinal mass
Specialty: Neurosurgery • Radiology

Objective: Unusual clinical course

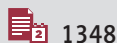
Background: Spinal myxopapillary ependymoma (MPE) is a slow-growing tumor arising from ependymal cells of the central nervous system. MPE rarely presents with acute neurological compromise and most commonly occur in the filum terminale or conus medullaris region. To date, only a few cases have been reported of patients presenting acutely because of hemorrhagic MPE.

Case Report: A 16-year-old boy without previous medical problems presented with a sudden onset of severe pain in the low back radiating to the thighs. He could not walk owing to the severity of the pain. Neurological examination revealed an unsteady gait, but the rest of the motor and sensory examination was normal. Lumbosacral spine magnetic resonance imaging revealed an intradural hemorrhagic mass extending from L5 to S2. The encapsulated hemorrhagic tumor was resected, and the pathology was consistent with MPE grade I. The patient made a significant recovery postoperatively. It is extremely rare for MPE to present with spontaneous hemorrhage in the lumbosacral region. Prompt diagnosis and management led to a favorable outcome. This case report is intended to highlight the atypical presentation and imaging features of hemorrhagic MPE.

Conclusions: We described a rare case of MPE in the lumbosacral region of a patient who presented with acute neurological compromise and atypical imaging features.

MeSH Keywords: Ependymoma • Lumbosacral Region

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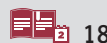
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Background

Spinal myxopapillary ependymoma (MPE) is a slow-growing tumor arising from ependymal cells of the central nervous system. It was first described by James Watson Kernohan in 1932 [1]. Patients are usually middle age and average 36 years old, and there is a slight male predominance. Common clinical manifestations may include back pain and progressive indolent course of nerve root-related impingement symptomatology, such as lower extremity stiffness, gait ataxia, sensory loss, and paresthesia [2]. The World Health Organization (WHO) classifies the histologic subtypes of ependymoma into 3 grades, and myxopapillary ependymoma and subependymoma are considered grade I tumors [3]. MPE rarely presents with acute neurological compromise [4], and to date, only a few cases have been reported of patients presenting acutely with hemorrhagic MPE [5,6].

Herein, we present a case report of a 16-year-old boy who presented acutely with spontaneous hemorrhagic MPE at the lumbosacral region.

Case Report

A healthy 16-year-old boy without previous medical problems presented with a 3-day history of sudden onset of pain in the low back radiating to both lower limbs. The pain extended from the buttocks to the back of the thighs, reaching to the back of knees, and it was associated with an inability to walk. He also reported constipation for over 2 days. No history of sphincter incontinence, trauma, fever, recent flu symptoms, or previous similar problems. Individual muscle examination while the patient was lying down did not reveal any motor deficit. However, he could not walk with a steady gait, which seemed to be related to the challenge of carrying his bodyweight when standing and walking. Sensory examination was unremarkable.

Magnetic resonance imaging (MRI) of the lumbosacral spine revealed a T2 hypointense intradural mass with corresponding heterogeneous diffusion restriction. The diffusion restriction was thought to be due to a hemorrhagic component, as illustrated by gradient-weighted images. Postcontrast images showed a lack of enhancement in most of the mass, except for an enhancing focus at the posterior aspect of the mass (Figure 1). MRI showed a slight vertebral bone scalloping. No bone erosions or foraminal extension was seen. Computed tomography scan of the lumbosacral spine was performed (not shown), but it did not extend the MRI findings.

The patient was placed in a prone position for operation. L4–L5 laminectomy and durotomy were performed. An intradural

encapsulated hemorrhagic mass was visualized (Figure 2). No bone erosion or foraminal extension was demonstrated. Gross total resection (GTR) of the tumor was achieved with the aid of intraoperative neuromonitoring. The neuromonitoring modalities used were somatosensory evoked potentials, motor evoked potentials, and free-running electromyography with direct nerve stimulation. Resection was achieved by microscopic technique and was done piecemeal and intralaminar. The tumor was dissected off the nerve roots. After all visible tumor was resected, the last structure appeared to be filum terminale that had firmer tumor attachment; a positive electromyography stimulation was suggestive of a nerve root. Therefore, this structure was not resected with the tumor that was removed from its surface. Primary closure of the dura was then done, and it was covered with fibrin glue medical sealant. No cerebrospinal fluid leak occurred during the postoperative period.

Histopathological examination showed an organized hematoma with an ependymal tumor that had a rich myxoid background. The tumor cells were well differentiated, cuboidal to elongated in shape, and radially oriented around vascularized myxoid cores (Figure 3). Immunohistochemical studies showed that the tumor cells were strongly positive for glial fibrillary acidic protein, pan-cytokeratin, and occasionally for S-100 protein. The Ki-67 proliferative index was low (Figure 4). The findings were consistent with WHO grade I ependymoma of the myxopapillary type.

Postoperatively, the patient was fully ambulatory with no deficits, and the postoperative course was unremarkable. No radiotherapy was given postoperatively because GTR was achieved. Postoperative MRI was not performed.

Discussion

MPE represents 13% to 27% of all spinal ependymomas [1] and 30% to 80% of ependymomas arising from the filum terminale and conus medullaris. MPE may present as an intradural or extradural lesion. Most MPEs present as intradural extramedullary masses, reflecting the usual origin of these tumors and the typical exophytic growth that often fills the distal spinal canal. MPE is typically isointense in relation to the spinal cord on T1-weighted images and hyperintense on T2-weighted images. The tumor typically demonstrates homogeneous postcontrast enhancement [1,7]. In our case, the tumor was T2 hypointense with corresponding heterogeneous diffusion restriction. In addition, postcontrast images showed a lack of typical enhancement in most of the mass, except for an enhancing focus at its posterior aspect. These features were attributed to the hemorrhagic content, which was illustrated by blooming in gradient-weighted images.

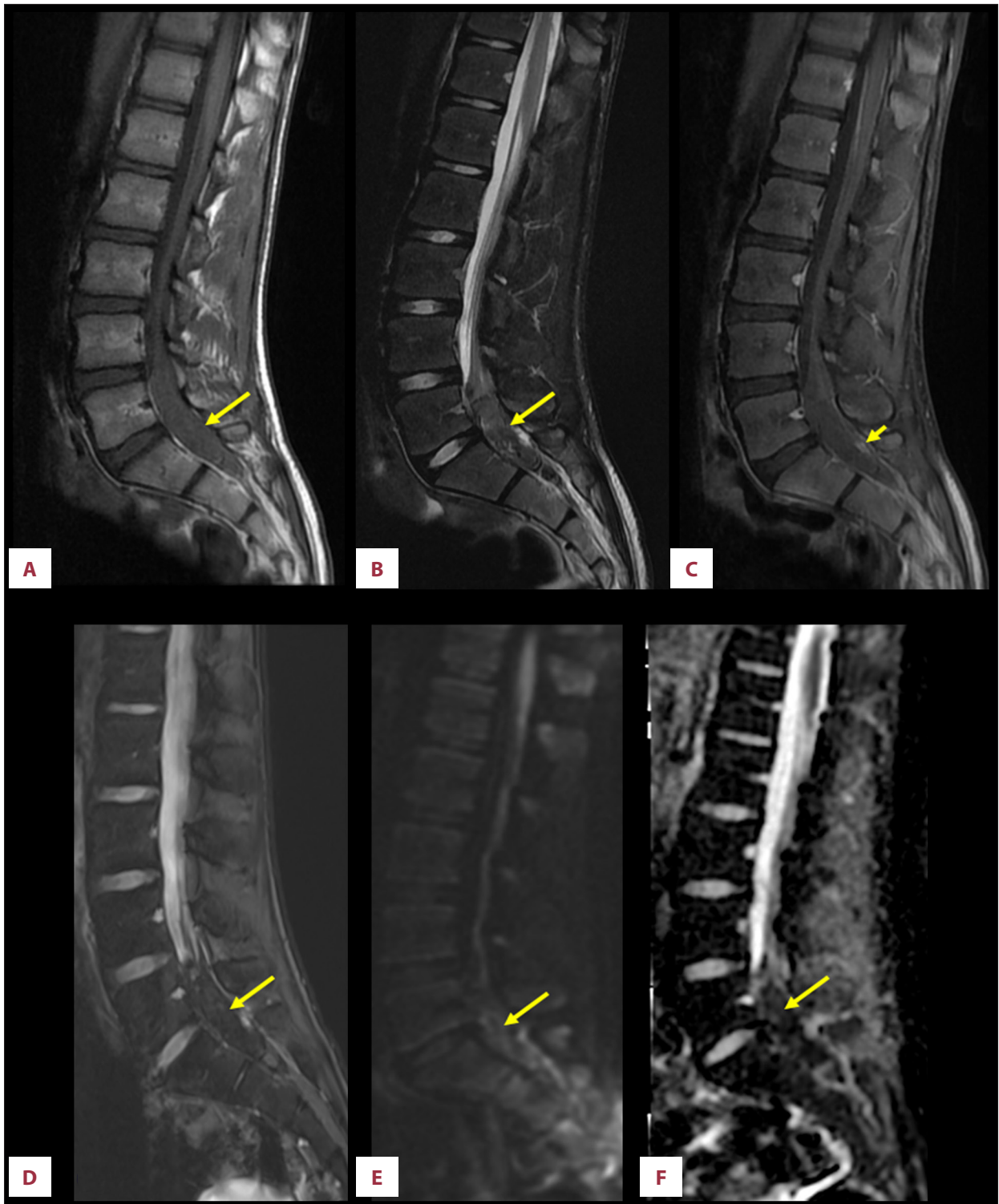


Figure 1. Lumbar spine magnetic resonance imaging. (A) Precontrast sagittal T1WI. (B) Fat-saturated mid-sagittal T2WI. (C) Postcontrast midsagittal T1WI. An intradural lesion can be seen at the levels of L5, S1, and S2. The lesion is isointense on T1WI and heterogeneously hypointense on T2WI. Postcontrast images showed a lack of enhancement except for an enhancing focus at the posterior aspect of the lesion. (D) T2* sequence showed blooming artifacts indicating hemorrhagic content within the lesion. (E, F) Diffusion-weighted and apparent diffusion coefficient (ADC) sequences showed the corresponding heterogeneous diffusion restriction.

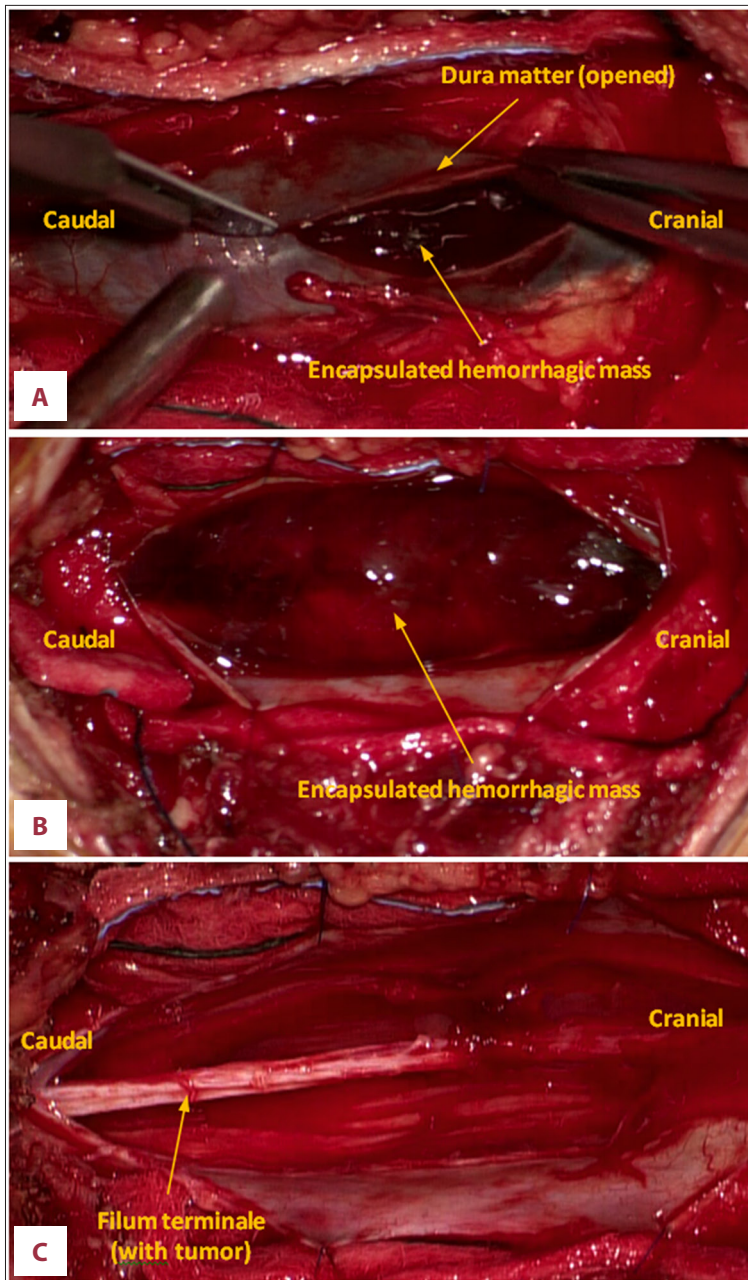


Figure 2. Intraoperative view of lumbosacral hemorrhagic myxopapillary ependymoma (A) during dural opening, (B) after full exposure revealing the encapsulated hemorrhagic mass, and (C) after removal of the tumor.

Spontaneous hemorrhagic MPE is rare. To date, only 14 case reports describing acute neurological compromise after hemorrhage of spinal MPEs have been published in the English medical literature [8,9]. In our case, the MPE was hemorrhagic and located at the L5–S2 level. A lumbosacral location of hemorrhagic MPE is rare. Among the previously published case reports, only 1 case report described a hemorrhagic MPE at the lumbosacral level [4,10]. MPE is a highly vascular tumor, and it has the highest risk of intratumoral bleeding relative to the other subtypes of ependymal tumors [9,11]. Multiple theories have been proposed to explain the tendency of an MPE to bleed [8,9]. High mobility, physical tension, and muscular

mechanical compression to the vessel walls in the lower lumbosacral region may predispose disruption and bleeding of tumoral blood vessels. Additionally, activity associated with elevated intrathoracic pressure, such as heavy lifting, has an effect similar to the Valsalva maneuver on the cardiovascular system. The pressure is transmitted to spinal veins, which can lead to stasis, rupture of tumoral blood vessels, necrosis, and tumoral hemorrhage [8]. Furthermore, vascular insult has been suggested to be predisposed by abuse of anabolic steroids [8]. The average age of presentation has been reported as 34.9 years (range, 15–65 years), and the main presenting symptom is predominantly severe back pain [11]. Our

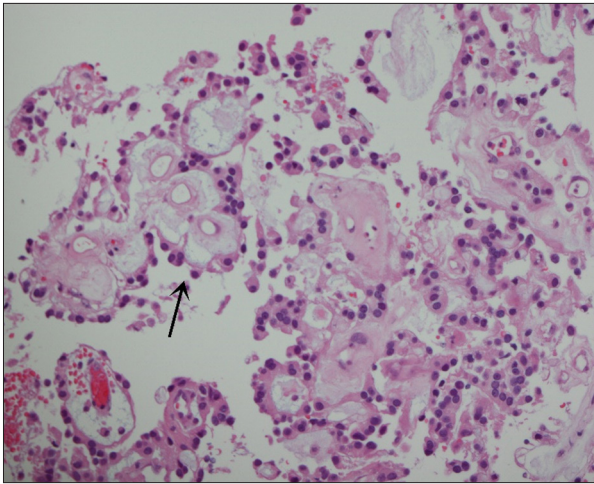


Figure 3. The tumor shows well-differentiated cuboidal to elongated tumor cells radially oriented around vascularized myxoid cores (arrow, hematoxylin and eosin staining, $\times 200$ magnification).

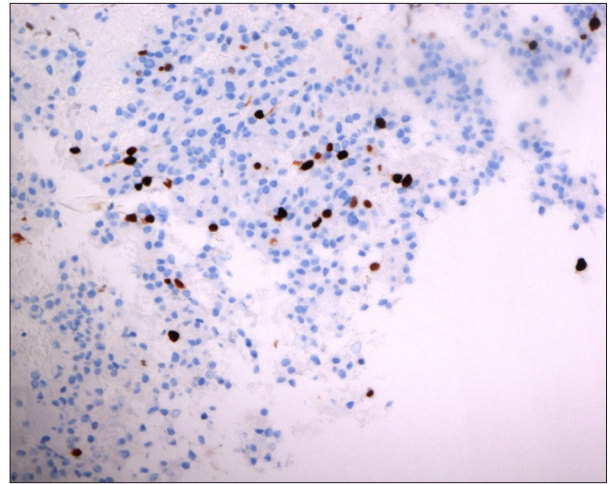


Figure 4. The Ki-67 proliferative index was low but focally increased (up to 8%, Ki-67 staining, $\times 200$ magnification).

patient was relatively young and physically active, with no history of any proposed risk factors, such as trauma, heavy lifting, or medication use.

The timing of surgical intervention relative to the timing of presenting symptoms is critical. Multiple studies reported a favorable prognosis for early surgical decompression [12]. The mainstay of treatment of spinal MPE is GTR, avoiding violation of the tumor capsule as much as possible. MPE has a high risk of central nervous system dissemination and may recur locally or distally even after a prolonged follow-up period [1]. The risk of recurrence is higher with subtotal removal of the tumor or tumoral capsule disruption. Marginal en bloc resection (i.e., GTR without tumoral capsule disruption) is the strongest predictor of overall survival, long-term control, and progression-free survival [1,13,14]. No consensus has been developed regarding adjuvant radiation therapy (RT) for MPEs [12]. In the majority of cases, RT is used for recurrence or subtotal tumor removal [1]. In a study conducted between 1972 and 2005 by Nakamura and his group, no tumor recurrence was found during a mean postoperative follow-up period of 10.4 years among 14 of 15 patients who underwent GTR followed by either whole brain and spinal cord radiation or local irradiation [11,15]. Feldman et al. [16] found that the

recurrence rate of MPE after GTR alone was 15.9%, which was similar to the recurrence rate for patients who underwent GTR with adjuvant RT. However, the recurrence rate for patients who underwent subtotal resection was 35.1% versus 29.3% for patients who underwent subtotal resection with adjuvant RT [11,16]. A similar conclusion was found by Tsai and his group [17]. Moreover, Abdulaziz et al. [14], found a positive trend for increased progression-free survival for patients who underwent adjuvant radiotherapy after either GTR or subtotal resection. Also, they found no recurrence among patients who underwent en bloc resection. In contrast, multiple other studies demonstrated that RT improves local control and progression-free survival regardless of the extent of resection [12,18].

Conclusions

We described a rare case of hemorrhagic MPE in the lumbosacral region in a patient who presented with acute neurological compromise and atypical imaging features.

Conflict of interest

None.

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