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Cauda Equina Syndrome Caused by Spontaneous Bleeding in the Filum Terminale Myxopapillary Ependymoma: A Rare Pediatric Case

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Established Facts

- A pediatric case of acute cauda equina syndrome caused by filum terminale ependymoma has not been reported to date.

Novel Insights

- Treatment of this atypical case resulted in very good functional recovery.

Key Words
Cauda equina syndrome · Filum terminale · Myxopapillary ependymoma · Pediatric ependymoma

Abstract
The majority of the filum terminale ependymomas are of the myxopapillary type, which most commonly present as lumbago or sciatic pain, an insidious clinical condition, at times accompanied by paraparesis, bladder paresis and vesical alterations. We report the case of a 13-year-old patient who presented with acute cauda equina. He underwent total resection of the lesion, which resulted in progressive improvement. The clinical conditions, diagnoses and treatments of the medullary cone and cauda equina myxopapillary ependymomas are also discussed.

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Introduction

Ependymomas are neoplasms with a prevalence of 3–6% in all CNS tumors [1]. They correspond to approximately 35% of the intradural medullary primary neoplasms [2]. Myxopapillary ependymomas are more commonly encountered at the medullary cone, cauda equina and filum terminale [3]. They are neuroectodermal tumors which represent grade I tumors in the World Health Organization classification, being observed mainly in the 3rd and 4th decades of life [2–4]. Cases in the pediatric population are rare [3, 5]. Approximately up to 20% of the myxopapillary ependymomas afflict pediatric patients [4].

Case Report

A 13-year-old male was taken to the Children's First Aid with a history of lumbago mostly located in the coccygeal region, which had begun 3 months earlier. He reported that initially the pain was weak, in pangs, but occasionally radiated posteriorly to both the left (LLE) and right lower extremity (RLE). He added that the pain had worsened 6 days before, and 1 day before presentation he had unexpectedly experienced weakness and numbness of both legs, followed by urinary retention. He had no history of trauma or fever.

The neurological examination on admission demonstrated LLE plegia, and grade I muscular strength in the RLE, abolished patellar and ankle reflexes, and absence of the bulbocavernosus reflex and anal sphincter tonus. Sensory deficits were noted in both legs.

The patient was submitted to thoracic column and lumbar magnetic resonance imaging (MRI) (fig. 1), which gave evidence of an expansive process (heterogeneous and intradural) located between L1 and L3. He underwent emergency surgery: microsurgical excision of the medullary expansive process via laminectomy at L1–L3 and laminoplasty. Once the dura mater had been opened, an extensive hematoma attached to the expansive process was noted (fig. 2a).

The tumor lesion presented a cleavage plane of the surrounding structures; however, it was closely attached to the filum terminale, from which it was completely dissected (fig. 2b). In a control MRI performed 24 h after surgery, no residual lesion was detected (fig. 1c, d). Tomography of the lumbar column performed postoperatively demonstrated preservation of the thoracic-lumbar curvature and patency of the laminoplasty. Histopathological and immunohistochemical results discovered a myxopapillary ependymoma of WHO grade I.

The patient has improved significantly as to motor deficits; clinical examination 2 months postoperatively revealed proximal and distal muscular strength of grade IV in the RLE, and grade III in the proximal LLE and grade I in the distal LLE. Sensitivity was preserved in the RLE but reduced in the LLE. Reflexes were still absent in both legs. He is able to walk with a walking aid.

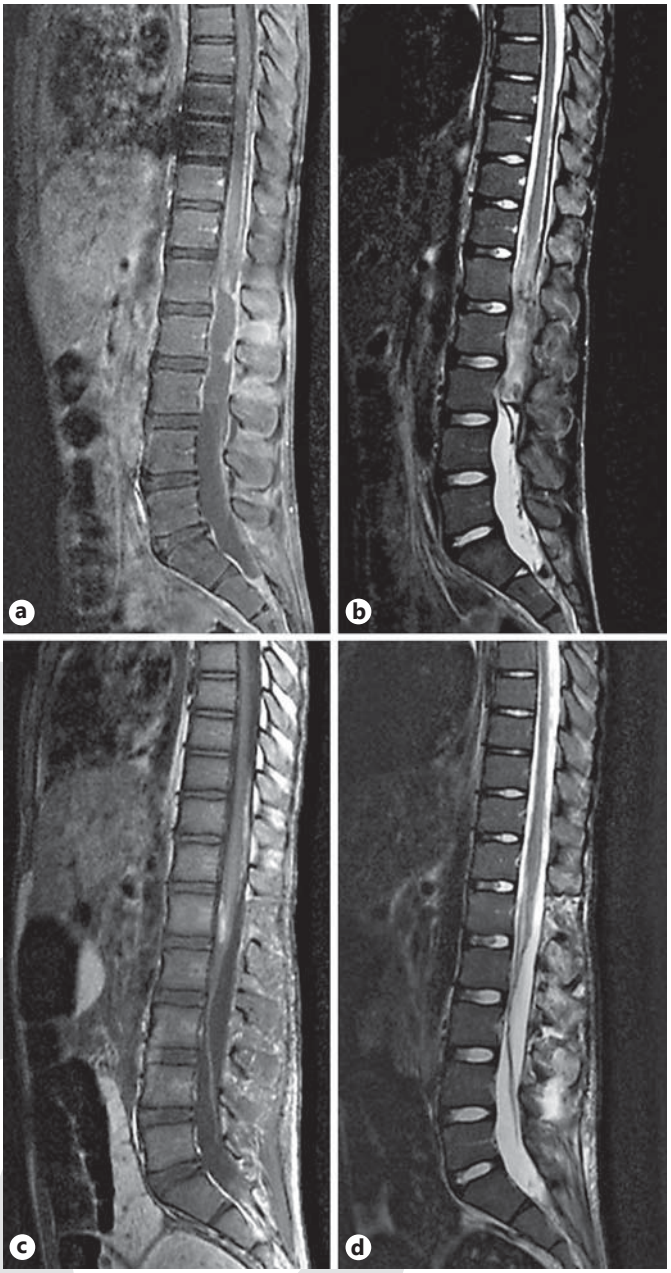

Fig. 1. Preoperative MRI of the dorsal and lumbosacral spine showing expansive intradural lesion of heterogeneous signal intensity adjacent to the medullary cone and filum terminale. Peripheral contrast enhancement (**a**) and concomitant hemorrhage at lesion margins are observable. **c**, **d** An MRI performed 1 day postoperatively depicted no residual tumor. **a**, **c** T1-weighted imaging. **b**, **d** T2-weighted imaging.

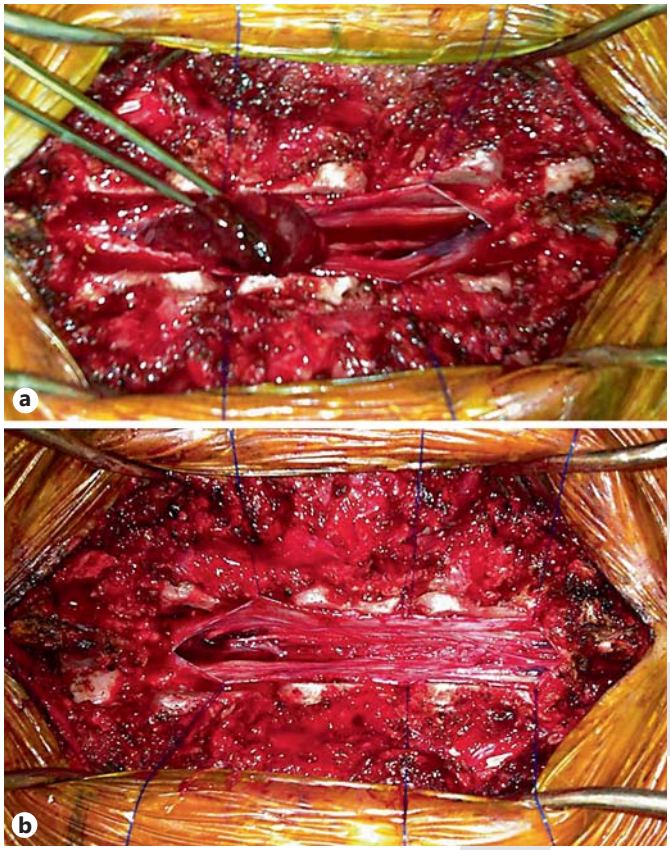

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Fig. 1. Perioperative microsurgical photographs showing an open dural sac from L1 (left) to L3 (right) with expansive process surrounded by hematoma (**a**) and the surgical bed following microsurgical resection of the expansive process (**b**).

Discussion

In the majority of cases, filum terminale and medullary cone ependymoma presents as insidious and prolonged progressive disease. It has been reported that 20 [2] to 90% [6] of the medullary cone or cauda equina tumors are myxopapillary ependymomas, which almost exclusively affect this region [2, 7]. Clinically, they are nonspecific [1, 2, 6], and lumbago or sciatic pain are the most common symptoms presenting [2, 6], but numbness, weakness and vesical alteration also occur [2]. Sinus pilonidalis has also been reported [8]. As the cauda equina syndrome induced by spontaneous bleeding of the ependymoma is rare [1, 2], there have been only 9 cases reported up to 2010 [1]. To our knowledge, there is no report on a child whose filum terminale myxopapillary ependymoma has caused an acute cauda equina syndrome following tumor bleeding.

Different theories, including mechanical and histopathological ones, have been put forward to explain the medullary cone ependymoma bleeding. The mechanical theory refers to peritumoral traction forces with rupture of tumor surface vessels [7], while the histopathological theory relates the bleeding to the myxopapillary variant, which presents numerous small blood vessels and loss of connective tissue [7], which determines the hemorrhaging.

The best diagnostic examination for expansive medullary lesions is MRI, either with or without contrast enhancement. Myxopapillary ependymomas are characterized by well-circumscribed lesions originating in the medullary cone, cauda equina or filum terminale.

In contrast to conventional ependymomas, the majority of these lesions are hypertense in T1-weighted MRI, as well as T2-weighted imaging; following administration of contrast medium, they become bright, and cystic or hemorrhagic alterations may become visible. The differential diagnosis of myxopapillary ependymoma includes drop metastases, meningioma and schwannoma [3]. They appear as fusiform masses along the filum terminale, as well as adjacent nodes [3].

Primary treatment consists of gross total resection [2, 3, 5]. Adjuvant radiotherapy does not have a clearly defined role [2]; however, in pediatric cases it may increase local control of the lesion and progression-free survival by 5–10 years [5]. However, in cases of grade I and II medullary ependymomas subjected to total resection, the watch-and-wait management appears to be the best strategy [9]. Prognosis is good, and overall and progression-free survival rates at 5 years are 93 and 68%, respectively [9]. Patients with neurological improvement have a greater chance of having a good neurological outcome after treatment [2].

Conclusion

We reported on a rare case of cauda equina syndrome due to spontaneous myxopapillary ependymoma bleeding in a 13-year-old boy. Although almost 48 h had elapsed between paraparesis and surgery, treatment resulted in progressive improvement in muscular strength, which enabled the patient to walk with support, making him independent in his daily activities.

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