CASE REPORT

Myxopapillary ependymoma of the filum terminale in a pubescent patient: a case report

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ABSTRACT

Myxopapillary ependymoma is a slow-growing benign neoplasm that accounts for 13 % of all spinal ependymomas. Clinically, it often presents with long-developing radicular syndromes and motor, sensory, urinary, and gait abnormalities. This report describes the case of a 14-year-old female who experienced a six-month history of lumbosacral pain, which radiated to the lower limbs during the last month. She was admitted to the emergency department at Hospital Belén de Trujillo, Peru, with crural paraparesis, motor function graded as 3/5 on the Daniels Scale, and urinary retention. Magnetic resonance imaging with contrast revealed a lumbar lesion consistent with ependymoma. Consequently, a laminectomy and total resection of the lesion were performed to prevent local recurrence and cerebrospinal fluid dissemination. At the most recent postoperative follow-up, the patient demonstrated good functional recovery. In this case, complete en bloc resection of the tumor was achieved. The hope is that this clinical experience can be replicated in other pediatric cases of myxopapillary ependymoma to improve patient quality of life.

Keywords: Ependymoma; Cauda Equina; Adolescent (Source: MeSH)

Ependimoma mixopapilar del filum terminal en un paciente pubescente: un reporte de caso

RESUMEN

El ependimoma mixopapilar es una neoplasia benigna de crecimiento lento que representa el 13 % de todos los ependimomas espinales. Se presenta clínicamente con síndromes radiculares de larga evolución, y anomalías motoras, sensoriales, urinarias y de la marcha. En este reporte, se describe el caso de una mujer de 14 años de edad, quien presentó un tiempo de enfermedad de seis meses caracterizado por dolor lumbosacro. En el último mes, el dolor irradió a los miembros inferiores. La paciente ingresó al servicio de emergencia del Hospital Belén de Trujillo, Perú, por paraparesia crural, con función motora 3/5 en la Escala de Daniels y retención urinaria. En la resonancia magnética con contraste se observó una lesión compatible con ependimoma a nivel lumbar, por lo que se realizó una laminectomía más resección total de la lesión, a fin de evitar la recurrencia local y la diseminación del líquido cefalorraquídeo. Hasta el último seguimiento postoperatorio, la paciente presentó buena evolución funcional. En el presente caso, se logró la resección completa y en bloque del tumor. Se espera que la experiencia clínica presentada pueda ser replicada en otros casos pediátricos de ependimoma mixopapilar para mejorar la calidad de vida de los pacientes.

Palabras clave: Ependimoma; Cauda Equina, Adolescente (Fuente: DeCS)

INTRODUCTION

Myxopapillary ependymoma is a glial neoplasm characterized by the radial arrangement of spindle or epithelioid tumor cells around blood vessels, with perivascular myxoid changes and microcyst formation (1). According to the World Health Organization classification, myxopapillary ependymoma is classified as a grade II ependymal tumor (1). It is the most common tumor of the conus medullaris, accounting for nearly 13% of all ependymomas (1 ,2).

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These tumors arise almost exclusively from the conus medullaris and the filum terminale (1, 2). Although they are primarily located in the lumbar spine, just below the conus medullaris, they may extend to the thoracic spine and sacrum (3). Some of these tumors can behave aggressively despite their histopathological features, potentially causing metastases along the neural axis to distant cranial and spinal locations (4). In the United States and Europe, the incidence ranges from 0.6 to 1 case per million people per year (1), with a male-to-female ratio of 2:1.

This case report describes a pubescent patient diagnosed with myxopapillary ependymoma of the filum terminale and presents a literature review that addresses clinical presentation, required imaging studies for diagnosis, the preferred surgical technique, and prognosis.

Given the low incidence of myxopapillary ependymoma of the filum terminale in pediatric populations both nationally and globally, reporting this case is of significant value for the clinical community involved in the management of these tumors.

CLINICAL CASE

We report the case of a 14-year-old female with a six-month history of lumbosacral pain initially managed with over-the-counter analgesics, resulting in near-complete symptom relief. One month before the presentation, the pain persisted despite medication and was accompanied by pain in the lower limbs. In the week before admission, symptoms worsened, and the patient developed difficulty walking. On the day of admission to the emergency department of Hospital Belén de Trujillo, she presented with urinary retention. On physical examination, she was alert and oriented to time, place, and person. She had crural paraparesis with a muscle strength score of 3/5 on the Daniels scale and a palpable distended bladder.

Lumbosacral spinal magnetic resonance imaging (MRI) with contrast revealed an intradural extramedullary hypointense lesion at the L3–L4 vertebral level on the T2-weighted sequence, consistent with a filum terminale ependymoma (Figure 1). Subsequently, an L3–L4 laminectomy was planned and executed under general anesthesia, with the patient positioned prone and supported by thoracic and hip rolls. The midline surgical incision was delineated using fluoroscopic imaging for enhanced precision.

A linear 10 cm incision was made through the skin and subcutaneous tissue, followed by subperiosteal dissection to expose the spinous processes, laminae, and articular facets. After separating the tumor from the nerve roots and coagulating and cutting the filum terminale, a laminectomy was extended to the medial border of the articular facets at L3 and L4, followed by a dural opening. The tumor was resected en bloc, achieving gross total resection. The mass had a cylindrical shape measuring 2.00 cm in length and 1.30 cm in width (Figure 2). The entire specimen was sent for pathological analysis (Figure 3).

Following surgery, the patient demonstrated a remarkably favorable recovery, with muscle strength improving from 3/5 to 4/5 on the Daniels scale. The Foley catheter was removed on day 7 after the procedure, and the pathology report confirmed a diagnosis of myxopapillary ependymoma. An MRI conducted after the surgery revealed no residual tumor, leading to the patient's discharge on day 8. Subsequent MRI studies with contrast of the cervical and thoracic spine, as well as the brain, showed no abnormalities. At the most recent follow-up, the patient had achieved good functional recovery under the care of the neurosurgery and clinical oncology teams.

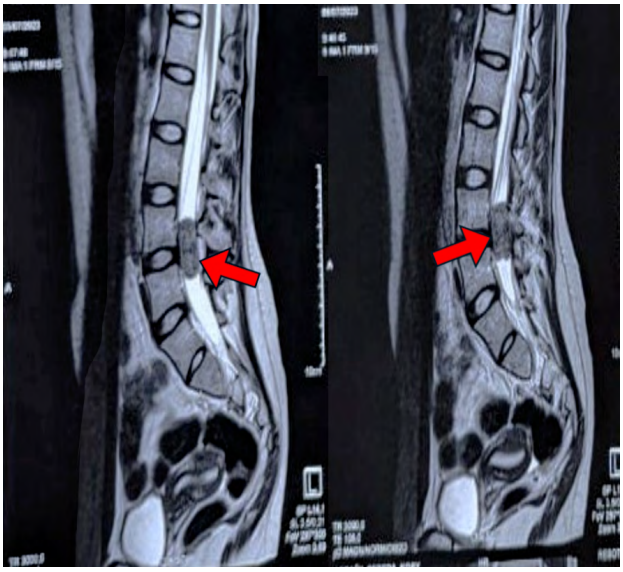

Figure 1. Preoperative lumbar spine magnetic resonance imaging.
The red arrow indicates the intradural extramedullary lesion at the L3–L4 level.

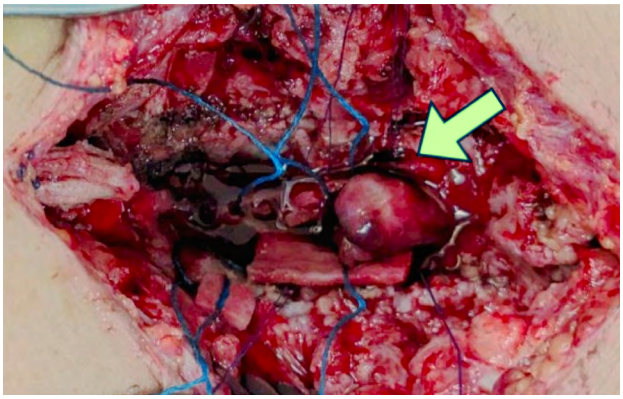

Figure 2. Dural opening and tumor exposure.
The green arrow indicates the location of the tumor.

DISCUSSION

Myxopapillary ependymoma is a tumor that commonly affects the filum terminale and the cauda equina. It was first described as a subtype of ependymoma by Kernohan in 1926 (5). The tumor originates from the ependymal glia of the filum terminale, and to date, no specific risk factors have been

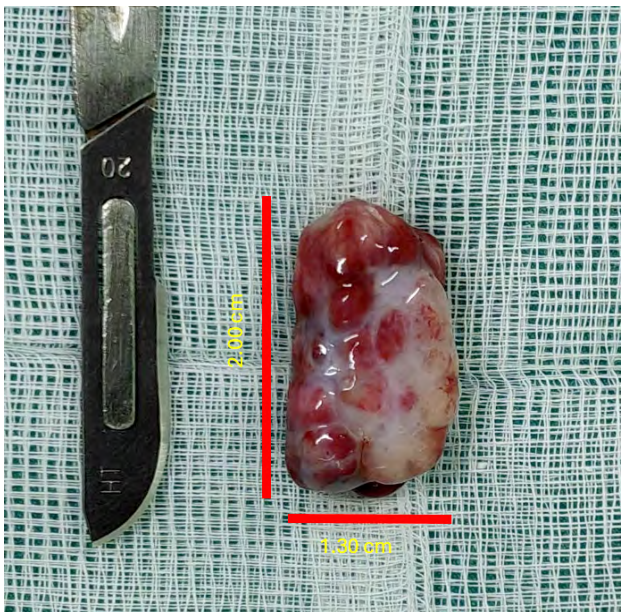

Figure 3. Tumor removed during the surgical procedure. A No. 20 scalpel blade is shown on the left for size comparison.

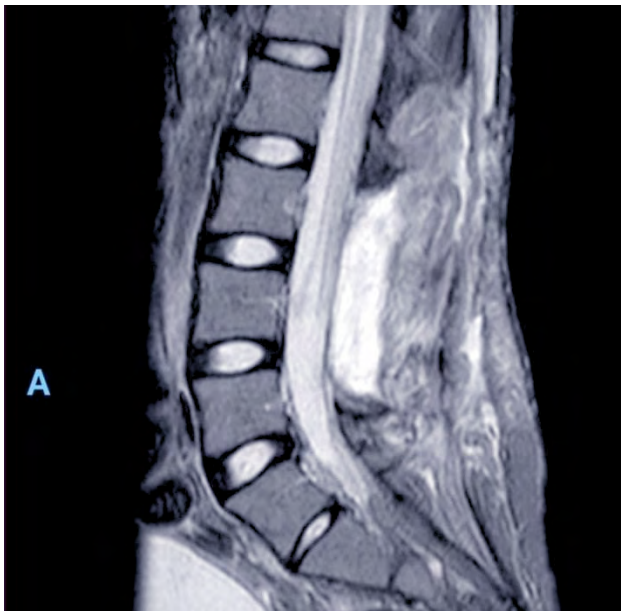

Figure 4. Postoperative magnetic resonance imaging

identified (6). MRI is the most effective imaging modality for diagnosing myxopapillary ependymoma due to its superior soft tissue contrast, which helps determine the tumor's extent and its relationship with intraspinal structures. MRI also enables visualization of the cauda equina above and below the tumor and can detect drop metastases in the subarachnoid space, providing essential information for preoperative surgical planning. Although MRI findings in myxopapillary ependymoma are nonspecific, the technique is crucial for guiding diagnosis (6,7).

The classification of myxopapillary ependymomas is based on tumor location and correlation with resection extent. Type IA includes intradural extramedullary tumors involving only the filum terminale, which are typically amenable to gross total resection. Type IB tumors involve the extramedullary conus with lumbar nerve roots and the filum terminale, making surgical management more challenging. Type II includes intramedullary tumors involving the conus and filum terminale that can also be grossly resected. Type III tumors involve the lower spinal cord, conus medullaris, and filum terminale, causing enlargement of the upper lumbar cord. Type IV A includes solid or cystic tumors of the lower spinal cord; type IV B involves the entire conus medullaris and typically presents with cystic components and signs of hydromyelia/syringomyelia at the tumor site. Gross total or subtotal resection may be achieved. Type V A includes tumors located outside the lumbar cord but within the spinal canal in the thoracic or cervical spine. Type V B includes tumors located outside the spinal canal, such as in the intracranial or sacrococcygeal regions (8).

The recommended treatment for patients with myxopapillary ependymoma is gross total resection. Patients undergoing subtotal resection generally receive adjuvant radiotherapy (9). Microsurgical techniques and spinal cord monitoring are fundamental for achieving complete tumor removal and optimizing neurological recovery. En bloc marginal resection without capsular rupture can be curative and is often achieved by resecting the filum above and below the tumor (8,9). However, depending on the tumor's size, shape, and relationship with cauda equina nerves or the spinal cord, this approach can be technically challenging. Nevertheless, in the hands of experienced surgeons, surgical morbidity is low, and postoperative clinical outcomes are generally favorable. Ten-year survival rates range from 92% to 100% (8,9).

Despite being classified as benign tumors, myxopapillary ependymomas pose a clinical challenge due to the potential for dissemination and local recurrence. Literature reports indicate distant dissemination and treatment failure in approximately 30% of cases. Recurrence rates are estimated at around 15% following gross total resection and up to 30% following subtotal resection (6,8,9). Up to one-third of adult patients may present with metastases at the time of diagnosis, so initial evaluation should include an MRI of the entire spine and brain. Adjuvant radiotherapy is recommended following partial resection or capsular rupture during surgery to prevent local recurrence and cerebrospinal fluid dissemination (8,9). In the present case, gross total en bloc resection was successfully performed. We hope this clinical experience can be replicated in future pediatric cases of myxopapillary ependymoma to improve patient outcomes.

Author contributions

Conceptualization: JM, CM; data collection, management, and curation: JM, CM; data analysis: JM, CM; visualization: JM, CM; original draft writing: JM, CM; writing and reviewing the final version: JM, CM.

Conflicts of interest

The authors declare no conflicts of interest related to the content of this manuscript.

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Ethical considerations

The study adhered to fundamental ethical principles in accordance with the Declaration of Helsinki, including non-maleficence and confidentiality. All collected data were strictly confidential and used as outlined in the study. Authorization for medical record review and informed consent were obtained prior to data collection.

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