

Drop foot and adult onset tethered cord syndrome caused by filum terminal ependymoma: A case report

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Drop foot and adult onset tethered cord syndrome caused by filum terminal ependymoma: A case report



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ABSTRACT

Introduction: Filum terminal is a fibrous tissue to stabilize the position of the spinal cord, located in both intradural and extradural sections anchored caudally to the coccyx. Tumors located at the filum terminal are a rare case. Filum terminal does not have any spinal cord component, but tumours in that area can directly cause a significant neurological disorder.

Case Presentation: Presenting 43 years old man with chief complaints of drop left foot and tethered cord syndromes. MR Image revealed 9,4x10mm intradural extramedullary mass at L1 with L1-L5 disc desiccation. The patient was untethered by laminectomy and underwent total mass excision. The sample was analyzed pathologically, and the results matched myxopapillary ependymoma. Pain sensation and sensory disturbance immediately improved after surgery. The urinary disorder also gradually improved. The patient was discharged without any urinary problems. After six months of follow-up, painless, mild left lower drop foot and mild gait disturbance was found without residual urinary and other neurological disorder.

Conclusion: Filum terminal ependymomas need to be considered for differential diagnoses in patients with drop foot and adult-onset tethered cord syndromes.

Keywords: Adult-onset Tethered Cord, Filum terminal ependymoma, Drop Foot.

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INTRODUCTION

Myxopapillary ependymoma (MPE), according to its pathology, was classified by World Health Organization (WHO) as Grade I Ependymoma, which is infrequently found.^{1,2} It grows from ependymal lining and embryonic nest cells, which have atypical clinical symptoms depending on the location and volume of the mass.³

The filum terminal is a fibrous tissue located both intradural and extradural of the spine to maintain and stabilize the spinal cord position, extending from the conus medullaris to the coccyx. The Filum terminal has no CNS component histologically. However,¹ a direct neurological disorder like drop foot and adult-onset tethered cord might occur since the filum terminal is anatomically located and serves the CNS.^{4,5} Serious neurological disorder caused by filum terminal mass has not been well reported

since its rarity.

CASE PRESENTATION

A 43-years-old man came with a chief complaint of 4 months history of progressively worsening back pain radiating to the left leg and left lower leg paralysis. In the last few days before admission, the complaint worsened with the frequency and urgency of urinating and further decreased sensory ability on the left leg. Another piece of information, there was no history of trauma or other chronic diseases and previous medical treatment.

Physical examination showed gait disturbance since the patient had leg weakness, radicular back and leg pain, and lasseque sign positive with slightly sensory loss. Abdominal examination found full bladder without any other abdomen complaint before. The neurological status was evaluated using American Spinal

Injury Association (ASIA) Impairment Scale at Grade A. MR Image revealed hyperintense T-2 weight images of 9,4x10 mm centrally localized intradural extramedullary mass lesion, which is well defined regular edge at L1 and intervertebral disc desiccation at L1-L5 level.

The patient underwent surgery with midline incision L1-L2 laminectomy and tumour excision. It revealed a white well-demarcated mass at the filum terminal without significant nerve root adhesion. Nerve root separation went well, and total excision was achieved with minimal bleeding and minimal damage to surrounding tissue.

The patient felt pain significantly improved after surgery, accompanied by sensory ability. After physiotherapy and six months of follow-ups after surgery, leg paralysis and pain disappeared, and he was not complaining of urinary or

sensory problems. Mild gait disturbance and drop foot remain. Pathology analysis found a proliferation of cuboidal to elongated tumor cells radially arranged in a papillary manner around vascularized stromal cores. Myxoid matrix material's foci accumulate between tumour cells and blood vessels, collecting in microcysts. These morphological appearances are consistent with MPE WHO grade I.

DISCUSSION

The filum terminal is a strand of intradural and extradural fibrous tissue extending from the conus medullaris to the caudal that stabilizes and supports the distal spinal cord from cephalic and caudal traction.⁶

There is no anatomically spinal cord component in the filum terminal, but neurological defects due to disruption of the filum terminal are very likely to occur. Spinal ependymoma most commonly occurs in the cervical region. However, MPE, a typical subtype of glioma, can occur exclusively in the filum terminal and conus medullaris due to the direct apposition of ependymal cells to the connective tissue in the filum terminal.^{7,8} Symptoms are atypical depending on the location and mass effect; the most common is low back pain. In this case, the mass grows slowly and progressively in the caudal spinal cord. The patient was unaware until the mass was large enough to tense the spinal cord causing tethered cord syndromes (TCS).^{9,10} TCS is a stretch-induced functional disorder of the spinal cord anchored by the filum terminal to the coccyx, usually present at birth or during childhood. Adult-onset TCS is a rare entity that usually presents with various symptoms. Diagnosing and treating adult TCS is challenging because of its rarity and mimics wide pathologies.¹¹ Radiological studies often help diagnose TCS based on the position of conus medullaris below L2, fatty and thickened filum terminal (63%).¹²

According to this case, symptoms contributing to the diagnosis were low back and leg pain, loss of sense of the entire left leg, and urinary disorder. As Sofuoglu reports, low back pain (65,2%) and leg pain (56,5%) were the common

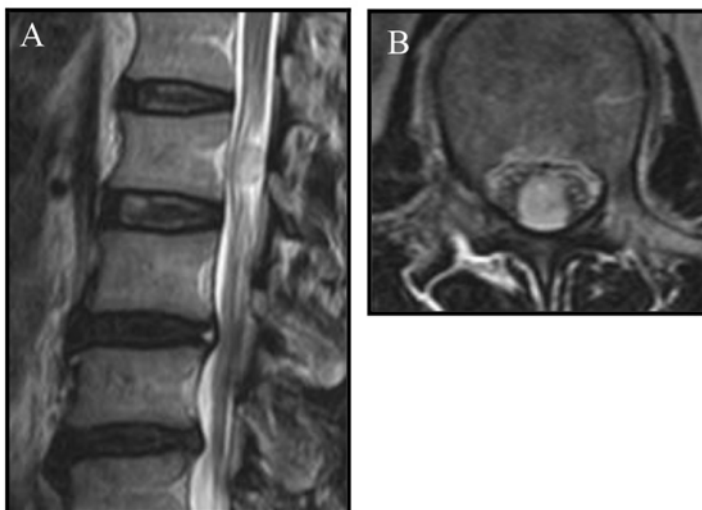


Figure 1. Magnetic Resonance Imaging (MRI) Findings. [A] Sagittal view T2 Weighted MR Image showing well-demarcated intradural low to intermediate signal mass at L1. [B] Axial view image showing intradural mass at filum terminal overlying with cauda equine.

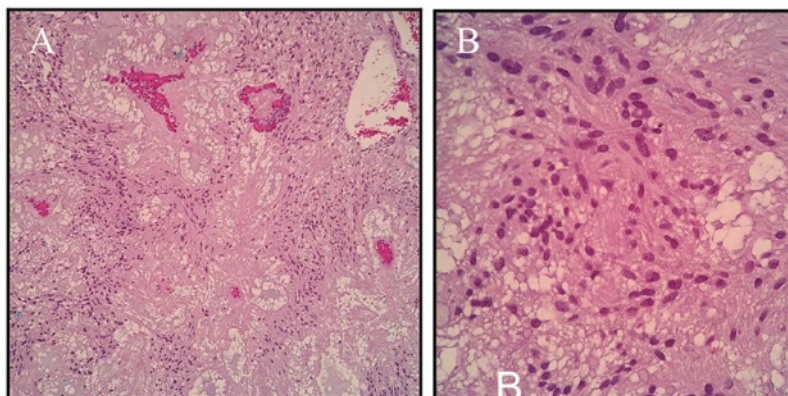


Figure 2. Pathology findings. [A] Layers of tumour cells with a radial perivascular arrangement and mucoid degeneration (H&E, 200x). [B] Cuboidal to elongated tumour cells with uniform round-ovoid nuclei (H&E, 400x).

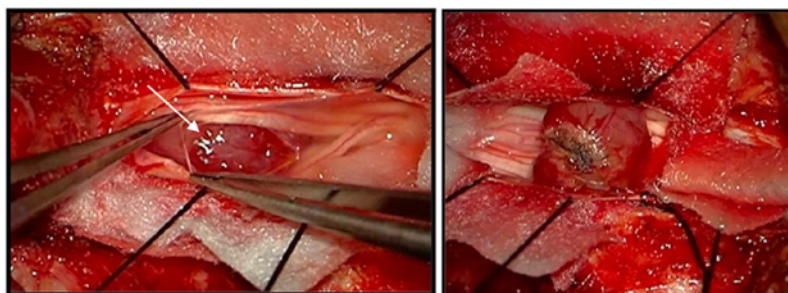


Figure 3. Intraoperative findings. [A] Well-demarcated tumour surrounded by nerve root while opening the dura mater. [B] Mass and nerve root was separated, and total mass was achieved.

presenting symptom of TCS in adults, including urinary disorder found in 52% of TCS patients.^{13,14} Bulent's study on 22 adults with TCS revealed the most common symptom was low back pain (68,1%), urinary disturbance (36,3%), fecal incontinence (9,09%), and hydrocephalus (4,5%).¹⁵ There are no significant changes yet in conus medullaris, and filum terminal as the patient suffered from some symptoms for less than a year.

Lower extremities symptoms typically consist of drop foot, weakness in left anterior tibialis, extensor hallucis longus, and digitorum longus. This condition is usually generated by herniated nucleus pulposus or foraminal stenosis that disrupts the peroneal nerve and radiculopathy.^{4,16} patient had worsened the case since he suffered by mass effect at the filum terminal and L1-L5 disc desiccation. Franklin D et al. reported that L4-L5 radiculopathy is the most commonly recognized cause of foot drop.⁴

Laminectomy and total excision of the mass with 1,5cm x 0,8cm dimension at filum terminal overlying unadhered to cauda equine were achieved in this case. As many as 84%-93% of MPE possible to do total excision since it rarely infiltrates the spinal cord; as Celano et al. reported, 79% of the patient who performed total excision have experienced significant improvement in a neurological deficit.^{7,17} A six-month follow-up visit in the outpatient department performed a detailed neurological evaluation. He came with painless residual motor weakness with a slight disability and underwent rehabilitation. Re-evaluate neurology status was improved as ASIA Impairment Scale Grade D. Urinary and sensory problem remains normal without any residual symptoms. The pain significantly decreased post-surgery, but other neurological symptoms take time to recover. Data from Toyoda et al., dividing 420 patients who underwent spinal decompression surgery into several clusters related to the outcome of 5 years of postoperative follow-up, revealed that 57% experienced a significant reduction in pain and disability starting at two years postoperatively, 17.6% experienced

mild residual pain, and 14% experienced severe leg numbness despite a significant reduction in pain during five years of follow-up.¹⁸

CONCLUSION

Filum terminal ependymoma should be considered for differential diagnoses. Early diagnosis is important as it could manifest in progressive symptoms, leading to drop foot and adult-onset tethered cord syndrome.

AUTHORS CONTRIBUTION

All authors contribute in all areas to prepare this study.

CONSENT FOR PUBLICATION

The patient approves verbally for publication of this study and the patient's identity remains confidential. Ethics approval from the International Committee of Medical Journal Editors (ICMJE) is fulfilled.

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CONFLICT OF INTEREST

The authors report no conflicts of interest in this work.

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