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Myxopapillary Ependymoma Revealed by Dysphagia: A Case Report

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Abstract | **Case Report**

Myxopapillary Ependymomas are slow growing tumors of the cauda equina with a high incidence in young adult. We report the case of a 35-year-old male, who consulted our emergency for a difficulty swallowing food. Upon further investigation, the patient complained of a gradually worsening heaviness of the lower left limb which has since spread to all the other limbs, making it difficult to walk. In addition, the patient had reported notions of drip urination and chronic constipation. Physical examination revealed numbness of the 4 limbs with normal and symmetrical reflexes, as well as thermoalgesia and hypoesthesia on the left of the abdomen. During cranial nerve examination, a vertigo and dysphagia were noted. A T1-Weighted MRI showed a hyposignal intradural tumor from L2 to L4 with a strong enhancement after gadolinium injection. A T2-Weighted MRI showed a hypersignal tumor. Additionally, there was a syringomyelic cavity clearly visible which extended from the medulla oblongata to the conus medullaris. An L2 L3 laminectomy was performed on the patient with a gross total resection of the tumor that was attached to the cauda equina roots. Upon good clinical recovery, the patient was discharged after satisfactory clinical recovery. An MRI performed 6 months later showed a total resection of the tumor. Ependymoma of the filum terminale associated with holocord syringomyelia are a very rare entity with a still uncertain pathogenesis.

Keywords: Ependymoma, Syringomyelia, Dysphagia.

INTRODUCTION

Intramedullary spinal tumors are well known to be associated with secondary syringomyelia. Ependymomas and hemangioblastomas are the most common tumors associated with syringomyelia. However, holocord syringomyelia secondary to a cauda equina myxopapillary ependymoma is extremely rare. The worldwide incidence of myxopapillary ependymoma is approximately 0.05 to 0.08 per 100,000 people annually, or about 1 in a million and makes up about 13% of all spinal ependymomas. It is most more often diagnosed in males between the ages of 20 and 40[7]. We present one such rare case.

CASE REPORT

We present the case of a 35-year-old male patient whose medical history includes a birth related left brachial plexus lesion which required physiotherapy. The patient suffered a 6 year long gradually worsening of his already existing heaviness of the left upper limb. The heaviness started affecting his lower left limb before spreading to the opposite side. In addition, the patient had reported notions of drip urination and chronic constipation.

A week before his admission to our institution, the patient consulted our emergency team for a recent difficulty swallowing food.

Upon examination, the patient was conscious with a tetraparesis making it difficult to walk, as well as thermoalgesia and hypoesthesia on the left side of the thorax and the abdomen. During cranial nerve examination, a vertigo and dysphagia were noted.

A T1-Weighted MRI of the spine [figure 1] showed a hyposignal intradural tumor from L2 to L4 with a strong enhancement after gadolinium injection.

A T2-Weighted MRI [figure 2] showed a hypersignal tumor.

Additionally, there was a syringomyelic cavity clearly visible which extended from the medulla oblongata to the conus medullaris. [Figure 3]

Contrast-enhanced MRI revealed a well-defined intradural extramedullary mass at the level of L2/L4 vertebrae with inhomogeneous postcontrast enhancement with associated holocord syrinx.

An L2 L3 laminectomy was performed on the patient with a gross total resection of the tumor that was attached to the cauda equina roots. [Figure 4]

Intraoperatively, the tumor was greyish-white, moderately vascular, and encapsulated with a good plane of cleavage between the tumor and the cauda equina roots [Figure 5]. Histopathological examination confirmed the diagnosis of ependymoma. He was doing well at the last follow-up six months after surgery and his symptoms had improved significantly with no sign of dysphagia. MRI of the spine done 6 months later did not show any evidence of residual tumor [figure 6].

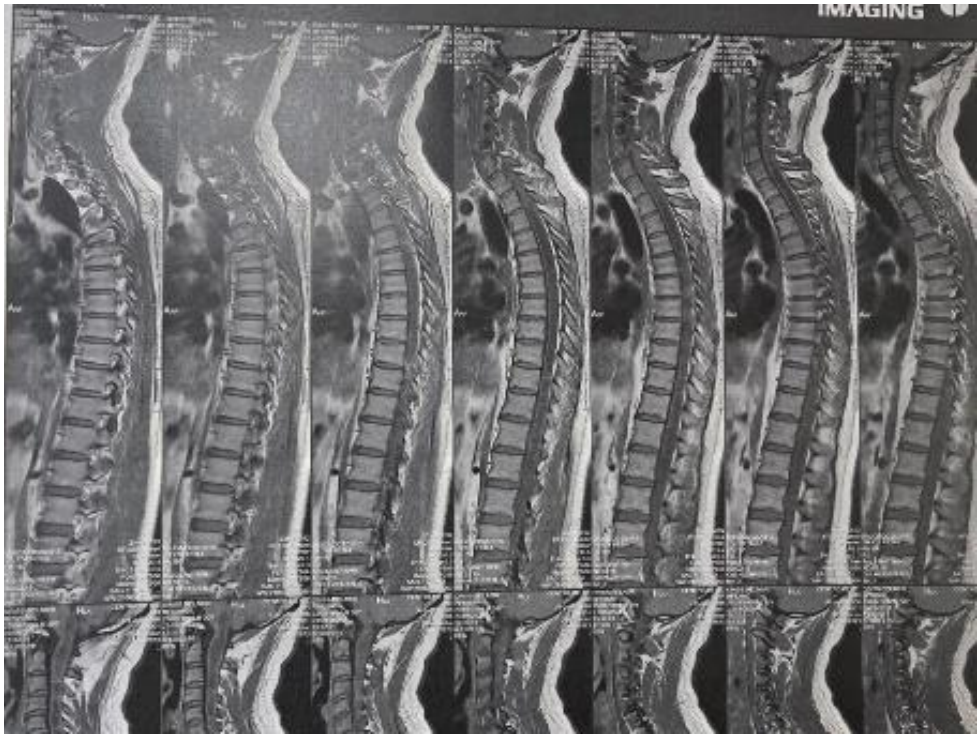

Figure 1: T1-Weighted sagittal MRI of the spine showing a hyposignal intradural tumor

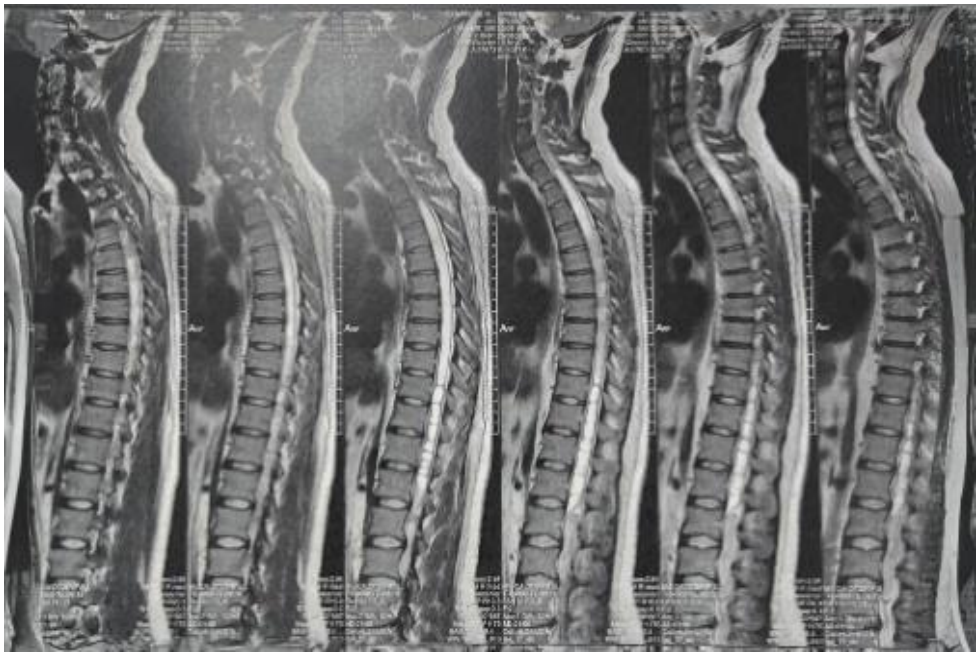

Figure 2: A T2-Weighted sagittal MRI showing a hypersignal intradural tumor with syringomyelia extending from medulla oblongata to conus medullaris

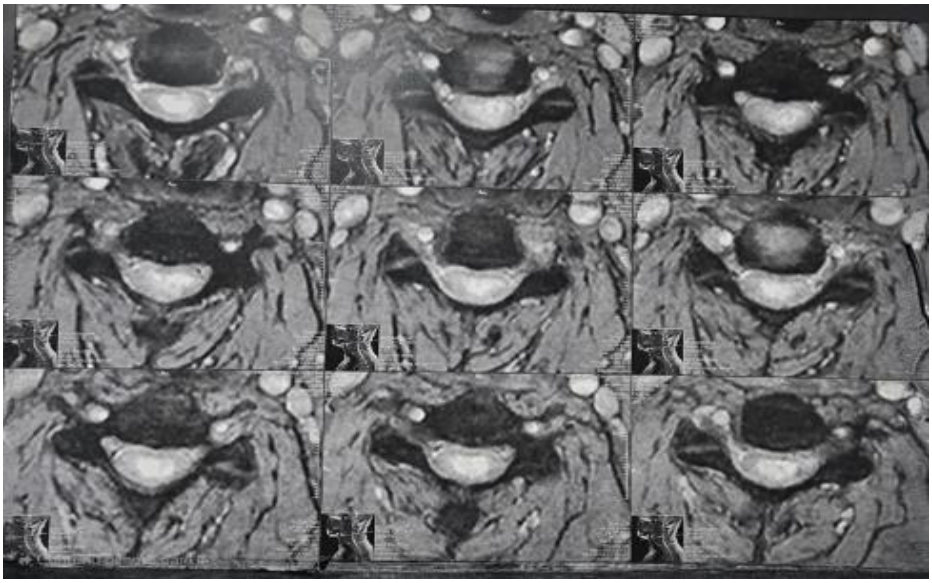

Figure 3: Axial MRI of the cervical Spine showing the syringomyelic cavity

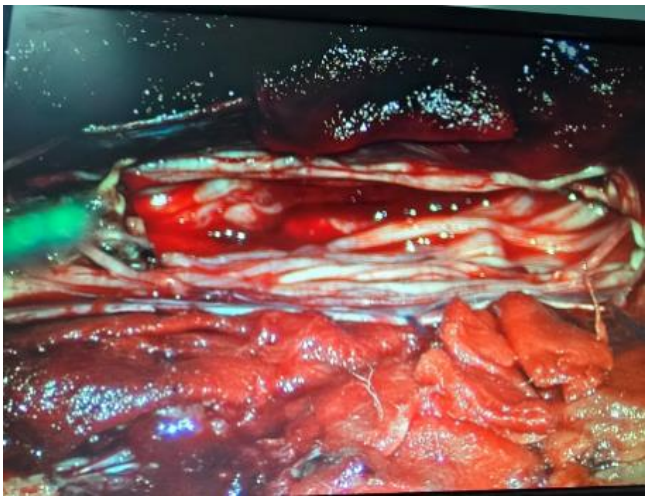

Figure 4: Intraoperative picture showing Gross total removal of the tumor after Laminectomy L2 L3

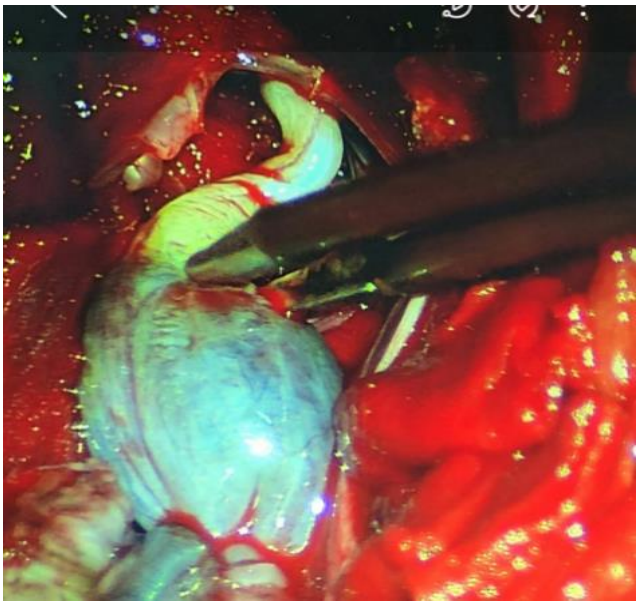

Figure 5: The tumor was greyish-white, moderately vascular

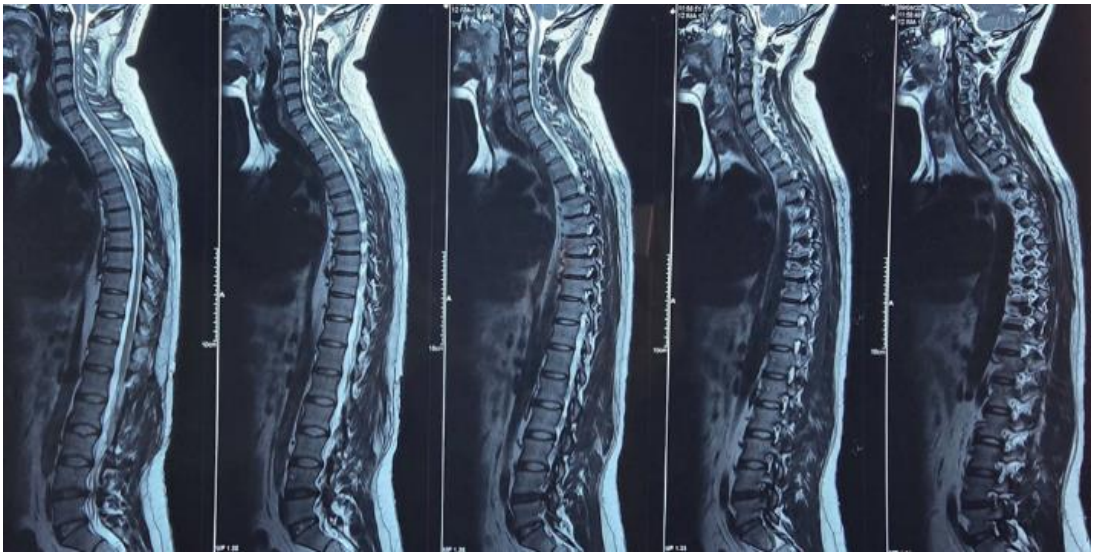

Figure 6: Post op sagittal MRI of the spine done 6 months later [T2W image]

DISCUSSION

Syringomyelia associated with spinal tumors is commonly reported with hemangioblastomas and ependymomas [1]. In most cases, tumors are intramedullary located in the cervicothoracic region. However, tumors which are located in the distal end of the spinal cord rarely cause syringomyelia [2]. In our case, the tumor was purely extramedullary and aroused from the cauda equina with extensive holocord syringomyelia from the medulla oblongata to the conus medullaris.

The presentation of syringomyelia depends on the extent of spinal involvement, going from motor and sensory weakness to cranial nerves involvement in case of syringobulbia [3], such as with our case.

The pathogenesis of syringomyelia associated with spinal cord tumors is still not well understood. It appears to be most likely due to transudation of fluid from the pathological tumor vessels [4]. Strong pressure force leads CSF from the subarachnoid space into the spinal cord's perivascular space, a process that occurs with every heartbeat. Over time, this fluid accumulation creates a cavity within the spinal cord that progresses further compressing the spinal cord and thus creating a space to further grow [8].

The other possible mechanisms include obstruction to CSF flow in the central canal and extracellular perimedullary fluid flow [5]. However, the exact reason is still uncertain.

Our case was unique as to the presentation of the syringomyelia with initial weakness of the upper limbs associated with dysphagia that led the patient to consult. As in our case, the initial presentation of ependymoma may be a complication of syringomyelia. Initial presentation with only bulbar palsy is also possible and was prior reported in a 16-year-old female patient [6]. Likewise, in all other similar cases that we came across, the removal of the tumor was sufficient for the resolution of syringomyelia without any additional procedure [2].

CONCLUSION

A unique patient with extramedullary myxopapillary ependymoma associated with extensive syringomyelia has been reported. Serial MRI demonstrated the total removal of the lesion and clinical recovery has been promising. However, the reason for the occurrence of syringomyelia associated with the tumor is still uncertain. Furthermore, the removal of the tumor was sufficient for the resolution of symptoms. This case should serve as a reminder that cranial nerve symptoms must not discard a thoracolumbar origin.

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