**Received:** 25.03.2010 / **Accepted:** 27.10.2010
**DOI:** 10.5137/1019-5149.JTN.3096-10.2

Case Report

# Giant Intramedullary Epidermoid Extending from the Brain Stem to the Upper Thoracic Spinal Cord

## *Beyin Kökünden Üst Torasik Spinal Korda Uzanan Dev İntramedüller Epidermoid*

Ritesh KANSAL, Amit MAHORE, Nitin DANGE

*Seth G.S. Medical College @ K.E.M. Hospital, Department of Neurosurgery, Mumbai, India*

**Correspondence address: Ritesh KANSAL** / E-mail: drkansal@yahoo.co.in

**ABSTRACT**

Epidermoid cysts are benign tumors originating from ectoderm remnants. Most epidermoid cyst cases are intracranial. Spinal epidermoid cysts are uncommon and most of the reported cases are in the thoracic and lumbar spine. Occurrence of intramedullary epidermoid cysts in the cervical spine is extremely rare. An 18-year-old male with a giant intramedullary tumor extending from the brain stem to the upper thoracic spinal cord presented at our outpatient department. The patient underwent total excision of a silvery white lesion through a midline myelotomy. Histopathological examination was suggestive of an epidermoid cyst. We present a brief report of the case and discuss the relevant literature.

**KEYWORDS:** Epidermoid, Intramedullary, Spinal cord

**ÖZ**

Epidermoid kistler ektoderm kalıntılarından gelişen benign tümörlerdir. Çoğu epidermoid kist vakası intrakraniyaldir. Spinal epidermoid kistler nadirdir ve bu vakaların çoğu torasik ve lumber omurgada bildirilmiştir. Servikal omurgada intramedüller epidermoid kistlerin oluşması çok nadirdir. Beyin kökünden üst torasik spinal korda kadar uzanan dev bir intramedüller tümörü bulunan 18 yaşında bir erkek hasta polikliniğimize başvurdu. Hastada bir orta hat myelotomisi yoluyla gümüş renginde beyaz lezyonun total eksizyonu yapıldı. Histopatolojik inceleme epidermoid kist düşündürdü. Vakanın kısa bir sunumunu yapıyor ve ilgili literatürü gözden geçiriyoruz.

**ANAHTAR SÖZCÜKLER:** Epidermoid, İntramedüller, Spinal kord

### INTRODUCTION

Epidermoid cyst occurring in the spinal cord is rare, comprising 0.6–1.1% of spinal tumors. Spinal intramedullary epidermoids are even rarer. Intramedullary epidermoids occur in the thoracic and lumbar area regions and their occurrence in the cervical spine is extremely rare. We present a patient with a giant intramedullary epidermoid cyst extending from the brainstem to the upper thoracic spinal cord.

### CASE REPORT

An 18-year-old male presented to us with weakness in the upper limbs for 2 years, neck pain for 1 year and stiffness in both lower limbs for 10 months. He also had a change in voice but no difficulty in swallowing, with intermittent retention of urine over the past 5 months. The weakness was more in the proximal than the distal group of muscles. He had to strain at micturition and defecation. On examination he had wasting of upper limb muscles. There was generalized hypertonia with exaggerated reflexes. The power in all four limbs was grade 4 (MRC grading). Magnetic Resonance Imaging (MRI) showed an intramedullary lesion extending from the medulla to the D2 vertebra. The lesion had heterogeneous intensity on T1W and T2W images (Figures 1A,B, 2). After suboccipital craniectomy with C1 to D3 laminectomies, the dura was opened. Midline myelotomy was performed to identify the pearly white tumor which was gradually debulked and total excision of tumor was achieved. The patient was unable to breathe well following surgery and had to be kept intubated on ventilatory support. His motor power remained the same post operatively. His respiratory condition did not improve and he died of pneumonia one month later. Histopathology was suggestive of epidermoid.

### DISCUSSION

Epidermoid cysts are benign tumors originating from ectoderm remnants from the 3$^{rd}$ to 5$^{th}$ week of gestation. They result from incomplete cleavage of the neural ectoderm from cutaneous ectoderm. The resultant of epiblasts in the neural tube may slowly grow with accumulation of normally dividing cells (1,2). They may reach a large size gradually before onset of symptoms (4,5).

The incidence of intracranial epidermoid is estimated to be around 0.2%-1%. Spinal epidermoid cysts are much rarer. Intramedullary epidermoid tumors remain limited to

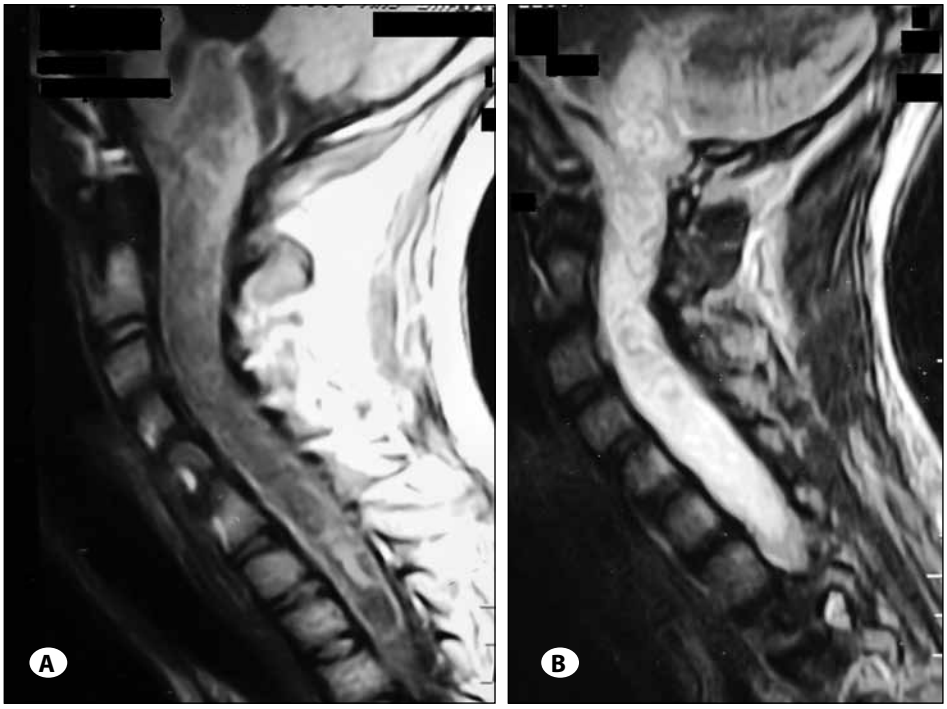


**Figure 1: A)** Post-contrast T1-weighted sagittal MRI of the craniovertebral junction and cervicothoracic spine showing heterogenous intramedullary lesion extending from the medulla to the D2 level with contrast enhancement at the periphery and widening of the spinal canal.
**B)** T2-weighted sagittal MRI image of the craniovertebral junction and cervicothoracic spine showing a heterogenous intramedullary lesion with predominant hyperintensity extending from the medulla to the D2 level.

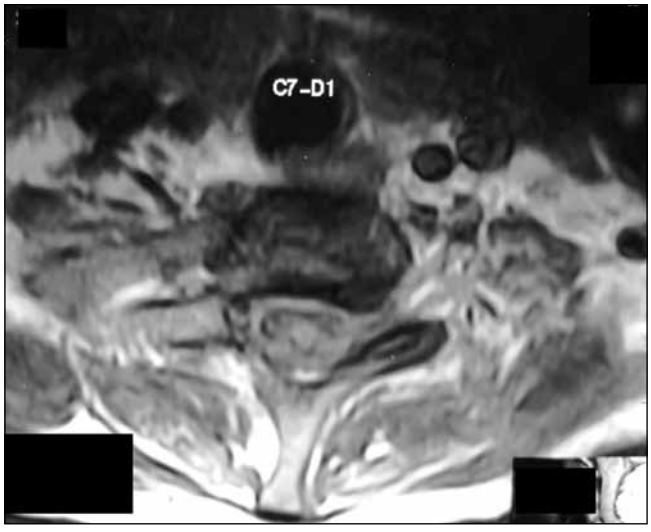


**Figure 2:** T1-weighted axial MRI at the C7-D1 level showing the heterogenous intramedullary lesion with contrast enhancement at the periphery.

case reports. Reported incidence within the spinal cord is extremely low, accounting for less than 1% of intramedullary spinal cord tumors.

Most cases of intramedullary epidermoid occur distally with rostral lesions being rare. The thoracic cord is the most common site for intramedullary epidermoid followed by the lumbar spine. Only two cases of cervical intramedullary epidermoids have been reported in the literature (3). This may be the first case of an intramedullary epidermoid extending from the brain stem to the thoracic spinal cord.

Radiological findings of these lesions may help to differentiate them from other intramedullary lesions. On magnetic resonance imaging, epidermoids show heterogeneity on T1- and T2-weighted images. Disparity in tissue density may be due to the variable lipid and protein component in these lesions. No contrast enhancement may be seen. Subtle abnormalities in bony spine due to slow progressive growth of lesion and due to imperfect midline developmental closure should be looked for (3).

Total excision of intramedullary epidermoids should normally be attempted. However, the capsule of the tumor when densely attached may have to be left behind to prevent neurological damage. Total excision of the lesion was achieved in our case but the patient developed respiratory difficulty in the postoperative period.

## ACKNOWLEDGEMENTS

The authors acknowledge with gratitude the encouragement and support of Dr. Atul Goel.

## REFERENCES

1. Berger MS, Wilson CB: Epidermoid cysts of the posterior fossa. J Neurosurg 62:214–219, 1985
2. Love JG, Kernohan JW: Dermoid and epidermoid tumors (cholesteatomas) of the central nervous system. JAMA 107:1876–1883, 1936
3. Ogden AT, Khandji AG, McCormick PC, Kaiser MG: Intramedullary inclusion cysts of the cervicothoracic junction. Report of two cases in adults and review of the literature. J Neurosurg Spine 7(2):236-242, 2007
4. Pennybacker J, Tytus JS: Pearly tumours in relation to the central nervous system. J Neurol Neurosurg Psychiatry 19:241–259, 1956
5. Rosenbluth PR, Lichtenstein BW: Pearly tumor (epidermoid cholesteatoma) of the brain. Clinicopathologic study of two cases. J Neurosurg 17:35–42, 1960