Epidermoid Cyst of the Thoracic Spine: A Rare Case

Nilesh Jain, Sharadendu Narayan, Harshad Patil, Abhishek Songara

ABSTRACT

Intraspinal epidermoid cyst is a rare tumor. The incidence in adults is lesser than 1% and in children lesser than 3%. Epidermoid cyst is predominantly seen at the dorsal spinal level. A large percentage of epidermoid cysts are intradural extramedullary. Intradural epidermoid cysts are rarer, with about 70 cases reported in the literature. These may be congenital or acquired with known association with spinal dysraphism. We hereby report a case of thoracic epidermoid cyst in a 22-year-old male with an extramedullary exophytic component and intramedullary cyst with accompanying split cord malformation at the level of lesion.

Keywords: Epidermoid cyst, Intramedullary, Intraspinal, Split cord, Thoracic.

INTRODUCTION

Intraspinal epidermoid cyst is a rare entity with an incidence of less than 1% in adults. Most epidermoid cysts are intradural extramedullary in location; the purely intramedullary variety is still rarer. Approximately 62 cases have been reported in the literature since the entity was first reported by Chiari in 1883. Epidermoid cysts are commonly located at the thoracic level, usually in D4–6 and D11–12 regions. Lumbar spine is the next site of predilection. Rarely epidermoid cyst is known to involve the cervical cord. Even though it is a rare lesion, timely diagnosis and surgical intervention have an important role in patient prognosis. We report the case of a 22-year-old male with an intradural extramedullary dorsal epidermoid cyst. This particular lesion appeared to be intradural extramedullary on imaging; however, intraoperatively the tumor was found to be intramedullary with a large exophytic component and split cord at the level of the tumor.

CASE REPORT

A 22-year-old male presented with mid-truncal paresthesia with band of hyperesthesia at the D4 level for the past 8 months associated with insidious onset progressive weakness of bilateral lower limbs for 1.5 months. The patient also had an associated history of urinary and bowel dysfunction since 2 months. Neurological examination revealed spastic paraparesis with a power of 4+/5 in the lower right limb and 3/5 in the lower left limb. The deep tendon reflexes were brisk at knee and ankle bilaterally, with the presence of left-sided patellar and ankle clonus and extensor plantar. The patient had suspended sensory loss to all sensory modalities below the D4 dermatome. He had no cutaneous manifestations of dysraphism with no history of any spine procedures or surgeries done in the past.

Magnetic resonance imaging (MRI) showed evidence of a 2.4 cm in cranio-caudal (CC) × 1.1 cm in right-left (RL) × 1 cm in anteroposterior (AP) intradural extramedullary lesion along the dorsal aspect of cord opposite the D2 vertebra and compressing the cord. The lesion was hypointense on T1W sequence (Figs 1A and 2A) and hyperintense on T2W sequence (Figs 1B and 2B) with an irregular or shaggy appearance of margins. There was no enhancement on contrast sequences (Figs 1C and 2C). The DWI image showed restriction of diffusion with corresponding low ADC (Fig. 2D). Complete neural axis imaging demonstrated no other lesions.
Fig. 2A: Magnetic resonance imaging DWI sagittal image: the mass lesion shows restriction of diffusion with corresponding low ADC

Fig. 2B: Axial MRI of the cervical thoracic spine, hypointense on T1W image

Fig. 2C: Axial MRI of the cervical thoracic spine, hyperintense on T2W image

Fig. 2D: Axial MRI of the cervical thoracic spine, no enhancement on contrast intraoperative images

Fig. 3A: Intramedullary pearly white, cheesy material, being removed in piecemeal fashion
The patient underwent a D2 laminectomy with microsurgical gross total excision of tumor in piecemeal fashion (Fig. 3A). Intraoperatively, a pearly white lesion with thin translucent cyst wall was noted (Fig. 3B). The lesion had a large exophytic component; the split cord was noted at the level of the lesion (Fig. 3C). Precaution was taken to prevent leakage of cyst contents into the subdural cavity, to prevent possible chemical meningitis. Histopathological analysis confirmed the lesion to be epidermoid cyst with characteristic compressed stratified squamous epithelium and fibrous tissue with small foci of calcification and cholesterol clefts (Fig. 3D). The patient had an uneventful postoperative period and was discharged on the 5th postoperative day. Postoperatively, the patient had improvement in the paresthesia symptoms and had improvement of lower limb power to grade 4+/5 bilaterally over 2 months. The symptoms of urinary and bowel dysfunction also showed improvement over a follow-up period of 4 months.

DISCUSSION

Epidermoid cyst was first described by Cruveilhier in 1835, who called them pearly tumors. Intramedullary epidermoid cyst was first described by Hans Chiari in 1883. Gros was the first to undertake successful surgical resection of epidermoid cyst in 1934. The epidermoid cyst is a rare intraspinal entity and has an incidence of 0.2 to 1% in most large series. Guidetti and Gagliardi reported a series of 452 intraspinal tumors in patients of all ages and found an incidence of 0.7% for spinal epidermoid cysts. Manno et al have reported a series of 90 intraspinal epidermoid cysts, out of which 39 were acquired and 51 congenital. Between 1883 and 1992, there were only 47 reported cases of pure intramedullary epidermoid cysts. Since then, in our literature survey, we have found approximately 22 cases.

The epidermoid cyst can be congenital or acquired in etiology. The acquired variety is more common with maximum incidence at the lumbar level. Acquired spinal epidermoid cyst was a known complication of lumbar puncture in the pre-stylet lumber puncture needle era. The time interval between the procedure and the incidence of intraspinal lesion varies between 1 and 20 years. Acquired epidermal cyst is usually extramedullary and is located at vertebral interspaces. Congenital epidermoid cyst arises from aberrant ectoderm inclusion during neural tube closure in 3rd to 5th week of embryonic life. This accounts for the midline location of most cysts, and their possible association with spinal dysraphisms, such as diastematomyelia, hemivertebra, dermal sinus tract, meningomyelocele, etc. Split cord malformation or diastematomyelia is a congenital spinal cord anomaly that is usually symptomatic. There are two varieties of diastematomyelia: Type 1 is the more severe variety with signs and symptoms of tethered cord; type 2 variant is more benign and is often asymptomatic. Epidermal cysts
are slow-growing lesions and the symptoms may often arise in 2nd to 3rd decade of life. The cyst behaves like any other space-occupying lesion, and signs and symptoms at presentation are usually progressive paraparesis, sensory loss, and rarely urological manifestations. The proliferation of magnetic resonance imaging has resulted in an early diagnosis in apparently normal patients. Patients with neurological findings should undergo surgery as soon as the diagnosis is made.

Epidermoid cysts are commonly hypo- or isointense on T1-weighted sequence and hyperintense on T2-weighted sequence; contrast enhancement is uncommon. There is restriction on diffusion weighted sequence with corresponding low intensity on apparent diffusion coefficient. The differentials of a spinal epidermoid cyst include hemangioblastoma, meningioma, metastasis, schwannoma, and dermoid cyst.

The treatment of epidermoid cysts is surgery with gross total excision as the goal. The capsule may be adherent to the surrounding cord tissue and any attempt to perform complete removal may result in neurological deficit. The prognosis and surgical outcome of epidermoid tumors of the spine is good, since they are histologically benign in nature. Even the long-standing lesions with neurological deficits may show some benefit from surgical intervention. Recurrence is rare after a complete removal of the tumor. Radiotherapy was offered as treatment in one case when the patient refused surgery. There has been one reported case in the literature of malignant transformation of an intramedullary epidermoid cyst in the thoracic region 3 years after its removal.

CONCLUSION

Spinal epidermoid cysts are rare benign tumors and have a long duration of symptoms with severe neurological deficits. Complete microsurgical excision is the treatment of choice; radiotherapy has a role in recurrent disease. Patients exhibit an improvement in symptoms following surgery even with severe neurological symptoms and urological dysfunction.

REFERENCES