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# Rare Case of Dorsal Epidermoid Cyst

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**Abstract:** ***Spinal epidermoid cyst is a rare entity. It can be congenital with associated spinal dysraphism or acquired due to repeated lumbar puncture and trauma. Congenital spinal epidermoid cysts without spinal dysraphism are rare. Here we are reporting a rare case of thoracic extramedullary intradural epidermoid cyst with no associated features of spinal dysraphism or any history of iatrogenic trauma in a young female patient.***

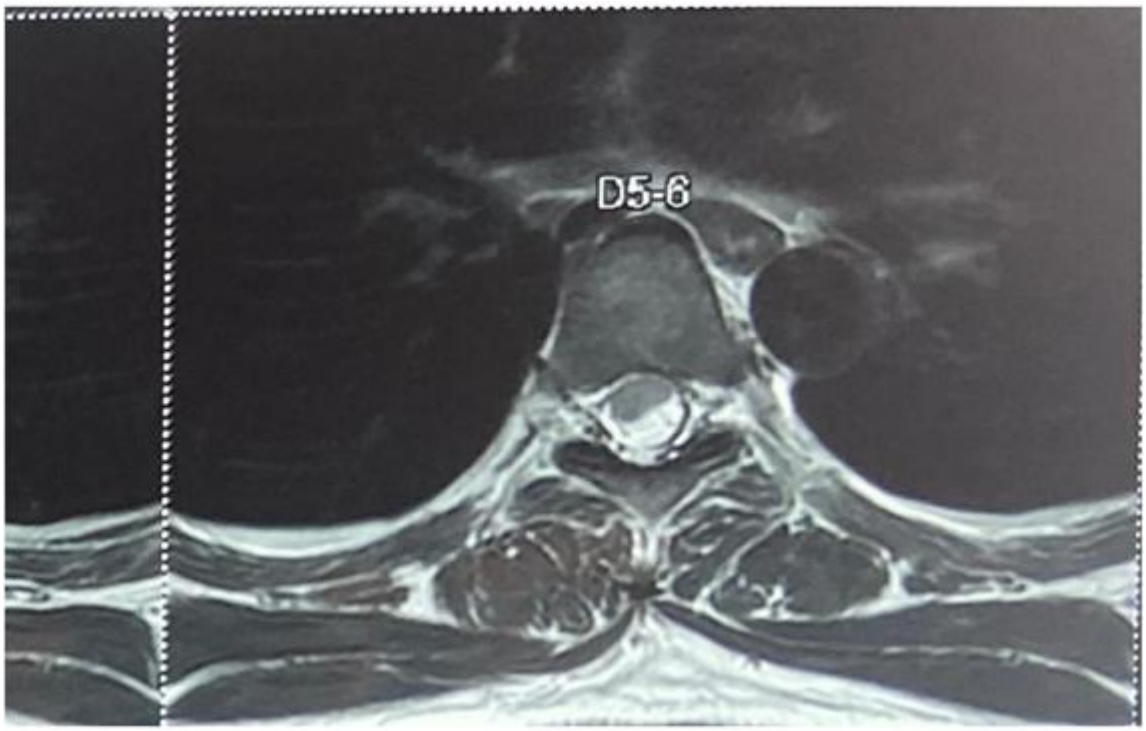
**Keywords:** Dorsal spine, epidermoid, lumbar spine

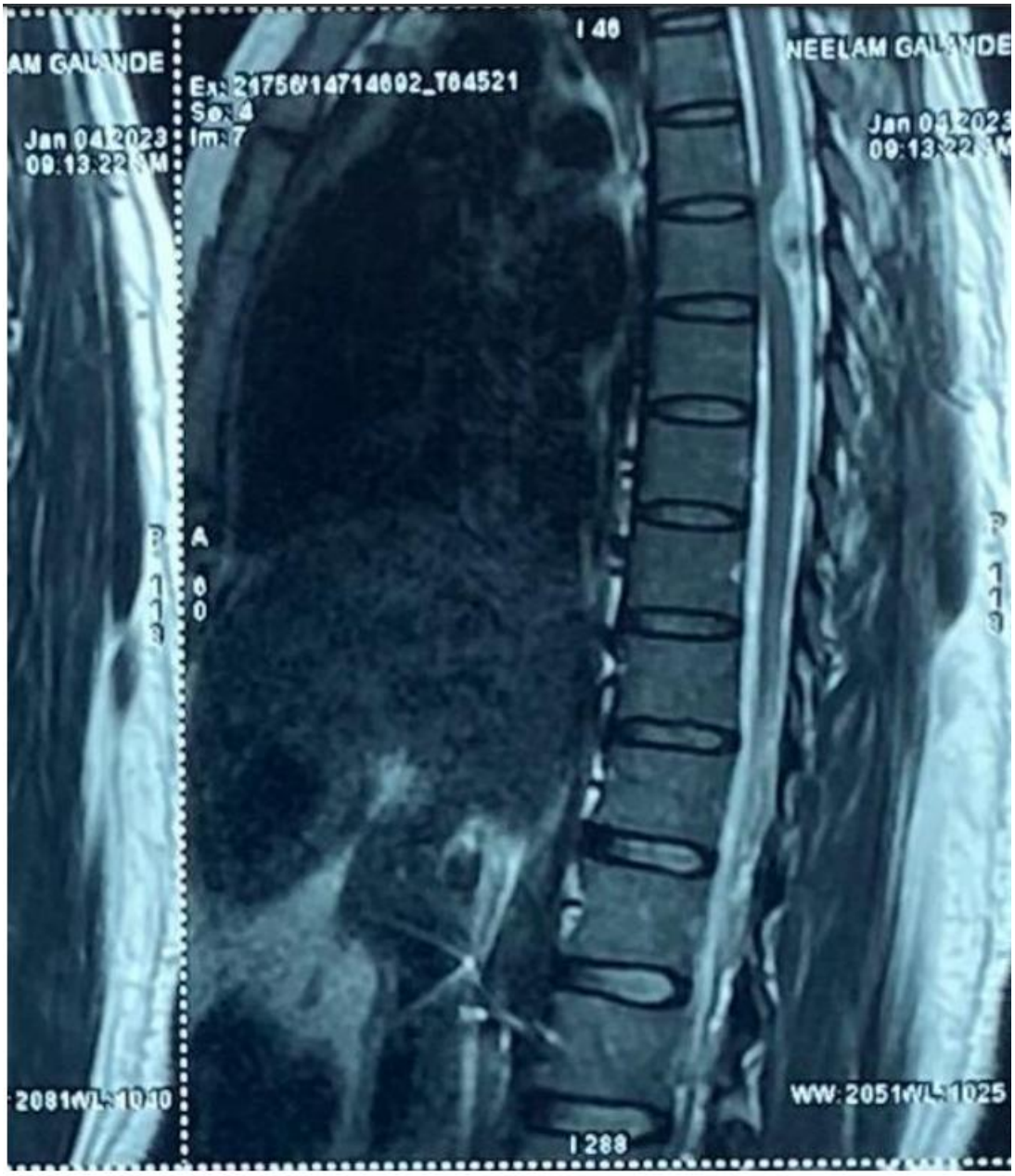
## 1. Introduction

Spinal epidermoid tumors are rare, accounting less than 1% of tumors involving the spine.[1] These tumors arise from pathological displacement of epidermal cells into the spinal canal. Therefore, these tumors can be congenital if there is defective closure of the neural tube; or acquired, in patients who have history of lumbar punctures, trauma, or surgery.[2, 3] Thoracic region is the commonest site of spinal epidermoid cyts, followed by the lumbar region and least commonly, the sacral and cervical regions.4 They appear, intraoperatively, as white masses that are encapsulated and are commonly referred to as "pearly tumors. "[5]. Patients usually present with pain as well as neurologic dysfunction that may include: muscle weakness, any sensory disturbances or loss of sphincter control[3, 5]

## 2. Case Summary

A 30 year old female presented in casualty with complaints of pain in both lower limb and difficulty in walking since 2 months with no co - morbidities. On neurological examination, spastic weakness was present in both lower limb with no involvement of the bowel and bladder. No features suggestive of spinal dysraphism were observed. There was no evidence of spinal dysraphism, the congenital nature of the lesion could not be established. Also, there was no his tory of trauma and lumbar puncture. Contrast enhanced Magnetic resonance Imaging (MRI) of spine revealed an intradural extra medullary nodular enhancing lesion at D5 - D6 level on T2w /STIR images. Lesion was seen along left dorsal aspect of spinal cord and causing significant compression and displacement of cord ventrally. Lesion is hypointense on T1. Compromised cord shows hyperintense signal suggestive of cord edema. Figure 1, 2






On surgical intervention intradural extramedullary epidermoid cyst was found and total excision was achieved. On decompression of cyst, there was cheesy material corresponding to keratin.

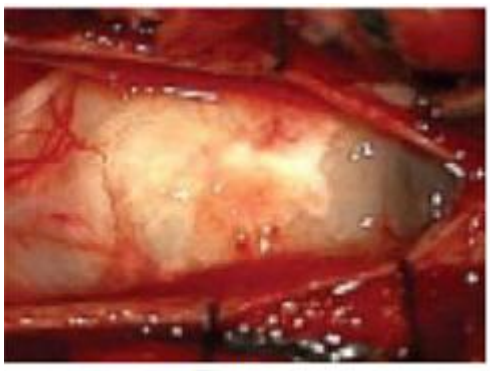


Figure3

Histological examination of the specimen with extensive sampling revealed afibrocollagenous cyst wall lined with keratinizing stratified squamous epithelium with preserved granular layer. Cyst contents were composed of abundant keratin flakes. Post operative period was uneventful with a progressive recovery and patient was discharged on post operative day 7 and is asymptomatic on follow up. Discussion Epidermoid cyst was first studied by Cruveilhier in 1835, who termed them pearly tumors.[6] Intramedullary epidermoid cyst was first described by Hans Chiari in 1883.[7] Gros was the first to undertake successful surgical resection of epidermoid cyst in 1934. The time interval between the procedure and the incidence of intraspinal lesion varies between 1 and 20 years.[8] 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, [9, 10]. 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. Epidermoid 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.[11] 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.[12] 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. 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.[13]

## 3. Conclusion

Spinal epidermoid tumors are rare, benign tumors that are more common in women and younger patients. As seen in other intradural, extramedullary tumors, patients typically present with low back and leg pain, bladder and bowel dysfunction, muscle atrophy, and sensory disturbances. Many cases are acquired from trauma, surgery, or lumbar puncture (46%). MRI with DWI is particularly useful to confirm the diagnosis with the lesions appearing avidly hyperintense. The most common approach to resection involved laminectomies and intradural tumor resection. GTR provided better outcomes in the majority of cases even when the tumor was adherent to the surrounding spinal cord or nerve roots.

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