



A Commoner at an Extremely Rare Site: A Case of Dorsal Intramedullary Spinal Dermoid Cyst

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AJNS 2023;18:219–222.

Abstract

Keywords

- ▶ intramedullary dorsal dermoid cyst
- ▶ histopathological diagnosis
- ▶ asymptomatic
- ▶ nonspecific imaging
- ▶ India

Intramedullary dorsal dermoid cysts are rare benign tumors, arising from the nests of embryonic ectoderm, which get buried or trapped under the lines of fusion of the ectodermal folds in the developing embryo. We present a rare case of intramedullary dermoid cyst in a 30-year-old asymptomatic female, who presented with paraplegia and ataxia. Magnetic resonance imaging was suggestive of neoplastic intramedullary mass. Intraoperatively, findings were suspicious of tuberculosis, but on final histopathological evaluation, the final and definitive diagnosis of intramedullary dermoid cyst was rendered. The postoperative phase was uneventful.

Introduction

Intramedullary dermoid cysts within the spine are rare, benign, and slow growing tumors. Dermoid cyst usually occurs on the head and scalp in the pediatric population. The adult spinal tumors can be extradural, extramedullary, or intramedullary, with intramedullary involvement, being extraordinarily rare site, and comprising of 0.3% of all adult spinal tumors. Only a few cases have been reported in the literature till date.

Dermoid cysts can be congenital or acquired. Congenital dermoid cysts are considered to be a developmental anomaly, associated with the entrapment of ectodermal elements along the lines of embryonic closure and are considered hamartomas.¹ Acquired cysts can be due to trauma or surgery. Dermoid cysts show mature skin appendages and usually contain keratinous debris and hair follicles.

In this patient, no congenital spinal abnormalities were reported, and the patient was absolutely asymptomatic before the fourth decade of life, illustrating the atypical presentation of this dermoid cyst.

We report a case of dorsal spine space-occupying intramedullary lesion in a 30-year-old female having an atypical presentation in conjunction with imaging and histopathological findings, and relevant scientific review, so as to create an awareness among the neurosurgeons, radiologists, oncologists, and pathologists. Histopathology always remains the cornerstone of definitive diagnosis in such cases.

Case Presentation

A 30-year-old female presented with a history of right lower limb weakness in the first trimester of pregnancy, which improved eventually. Now she presented with bilateral lower

article published online
March 31, 2023

DOI <https://doi.org/10.1055/s-0043-1764326>.
ISSN 2248-9614.

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limb paraplegia and ataxia in the third trimester. There were no associated systemic comorbidities. General physical examination revealed Glasgow coma scale-E4V5M6, extra-ocular movements were normal, and respiration was spontaneous, stable, and adequate. Systematic examination showed 5/5 power with mildly increased tone in bilateral lower limbs. Superficial sensation and pain were normal. Magnetic resonance imaging (MRI) was performed.

Imaging Findings

Contrast-enhanced MRI revealed a well-demarcated central intramedullary abnormal signal intensity mass that was isointense on T1-weighted imaging (T1WI) and heterogeneously hyperintense on T2WI and showed intense contrast enhancement in the dorsal spinal cord at D7-D8 vertebral level. Findings were suggestive of neoplastic lesion, ependymoma, or schwannoma (→Fig. 1A,B).

Management

Neurosurgical evaluation was done and the patient was planned for D8-D9-D10 laminectomy and excision of intramedullary tumor.

Intraoperatively, the lesion contained what looked like purulent material to the surgeon, and it was sent for frozen and further culture studies.

Frozen sections showed a cystic cavity lined by squamous epithelial lining. The culture report showed no growth after 48 hours of incubation at 37 °C.

Postoperatively, she developed paraparesis with foraminal power of 2/5 and distal 3/5, and hypoesthesia bilaterally in the infraumbilical region. She was not aware of passing stools.

She was discharged 2 days after surgery, and was conscious and oriented with stable vitals. At the time of discharge, paraparesis hip extension 2/5, knee extension-2/5, hypoesthesia below umbilicus. Joint position was impaired. Regular follow-up and clinical monitoring were advised.

Pathology

Macroscopically, multiple gray white to gray brown soft tissue pieces were received. Microscopically, hematoxylin and eosin-stained sections showed cyst wall with focally keratinizing squamous epithelial lining, focal stratified columnar, and mucinous lining and ducts and glands in a collagenous stroma with smooth muscle, focal calcification, bone, and neuroparenchyma. Findings were suggestive of intramedullary spinal dermoid cyst, with no evidence of malignancy (→Fig 1C-E).

Discussion

Dermoid cysts rarely occur in the spine, especially at the intramedullary site that was first described in 1745 by Verratus.² Intramedullary dermoid tumor also called as

spinal cutaneous inclusion tumor arises due to improper separation of neuroectoderm from surface ectoderm. The incidence ranges from 60% in the lumbosacral spine to 10% in the thoracic spine and 5% in the cervical spine.²

The symptoms usually depend upon the location and extent of the tumor and are the result of compression of adjacent structures by the lesion.

Symptoms include pain, motor regression, weakness, and urinary incontinence.

The differential, which can be entertained in this case, is an epidermoid cyst, which differs from dermoid cyst histologically. Dermoid cyst is lined by epidermis that contains skin appendages such as hair and sebaceous glands and other embryological layer derivatives such as bone, glands, and neuroparenchyma, while epidermoid cyst is lined by multi-level squamous epithelium and contains only declaimed keratin.³

These cysts are often diagnosed when they present clinically with neurological manifestations, from paresthesia to paralysis and sphincter complications.⁴

Gercek et al also reported a case of a patient with a spinal dermoid cyst with a dyssynergic bladder. The case series of epidermoid and dermoid tumors of the spinal cord presented by Bradford, most of the cases presented with urinary symptoms, either retention or incontinence.⁵

In our case, the patient had paraparesis and ataxia during pregnancy, which improved later on with no bladder and bowel symptoms prior to surgery. Our case report also calls attention to the diagnosis of dorsal intramedullary dermoid cyst in spinal surgical patients with the help of MRI and histopathological evaluations. Our patient was initially diagnosed as possible tuberculosis of the spine due to conventional conjecture and overlapping symptoms. In the case of misdiagnosis, rupture of the cyst can result in arachnoid, meningeal, or cerebral irritation as a result of dissemination through cerebrospinal fluid pathways.

The mainstay of treatment is total excision at an early stage. Excision of extramedullary dermoid cysts is possible, while in intramedullary dermoid cysts, the capsule is attached to the spinal cord and often causes difficulty in complete resection through surgery. Intramedullary dermoid cysts have the propensity to rupture during surgery, after trauma or spontaneously, causing the spread of contents through the arachnoid space and leading to meningitis. In our case, the postoperative phase was uneventful.⁴

Conclusion

Spinal intramedullary dermoid cysts are rare tumors without specific clinical presentation. Early diagnosis can be facilitated with the help of MRI. Clinical presentation is usually neurological. Surgical excision is the treatment of choice, which not only results in cessation of clinical progression but also in the remission of symptoms. Histopathological examination is the gold standard for diagnosis and an intraoperative consultation may aid in excluding an infective etiology in such scenarios.

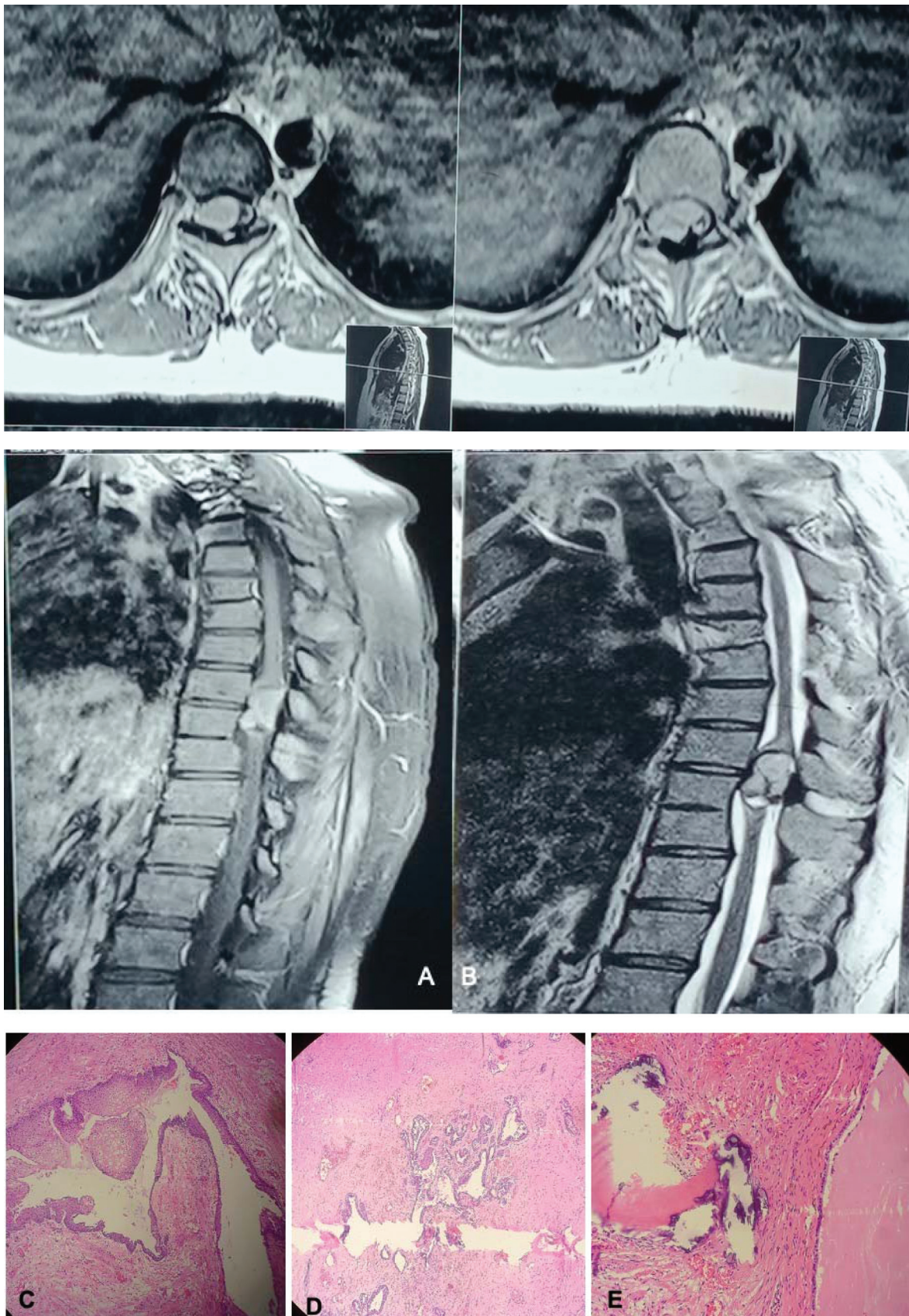


Fig. 1 (A, B) Contrast-enhanced computed tomography spine showing central intramedullary dorsal spine mass at D7-D8. (C-E) Microscopic photograph showing cyst lined by squamous lining, mucinous glands, neuroparenchyma, and bone formation, suggestive of dermoid cyst (hematoxylin and eosin $\times 100$).

Ethical Approval

Legal entity of this case report: Dharamshila Narayana Superspeciality Hospital, New Delhi, 110092, India

Informed Consent

(i) All participants provided written informed consent for the participation in the study.

(ii) Patient consent was obtained for purpose of the study with due care to maintain his/her privacy.

Conflict of Interest

None declared.

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