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*SNI: Spine*

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*Case Report*

# Spinal epidermoid cyst associated with limited dorsal myeloschisis

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## ABSTRACT

**Background:** Epidermoid cysts (ECs) are rare benign tumors arising from epidermal cells, associated with congenital abnormalities or acquired through trauma, surgery, or lumbar punctures. They represent <1% of all intraspinal tumors and may be associated with limited dorsal myeloschisis (LDM).

**Case Description:** A 7-year-old neurologically intact male had a dorsal skin mass since birth located posteriorly in the midline of the inferior thoracic spine. The mass was palpable, painless, mobile, vascularized, and could be transilluminated. Thoracic magnetic resonance imaging showed an extensive intradural extramedullary cystic lesion extending from D6 to D8 that did not enhance with contrast, accompanied by a subcutaneous fluid collection at D8–D9 communicating with the subarachnoid space. The patient underwent gross total resection of the lesion, pathologically confirmed as an EC. The postoperative course was uneventful, with no recurrence 1 year postoperatively.

**Conclusion:** LDM may be associated with ECs. Early diagnosis and surgical resection of these lesions are essential for favorable outcomes.

**Keywords:** Congenital epidermoid cyst, Limited dorsal myeloschisis, Spine

## INTRODUCTION

Limited dorsal myeloschisis (LDM) is a rare form of spinal dysraphism characterized by a focal midline skin lesion and a fibroneural pedicle or stalk connecting the spinal cord to the overlying skin lesion.[4] Spinal epidermoid cysts (ECs) are benign tumors arising from epidermal cells and occur in two forms: congenital malformations (bone and skin) or acquired lesions.[5,8] Here, we present a rare case of a 7-year-old male with an EC from T8–T9 accompanied by LDM.

## CASE DESCRIPTION

A 7-year-old male presented with a progressively enlarging lower thoracic dorsal T8–T9 skin mass since birth. On examination, it was palpable, painless, mobile, vascularized, and readily transilluminated [Figure 1]. Thoracic magnetic resonance imaging (MRI) showed a T1 hypointense/T2 hyperintense intradural extramedullary cystic lesion at the T8 level with an exophytic component that did not enhance with contrast, and a subcutaneous fluid collection [Figure 2].

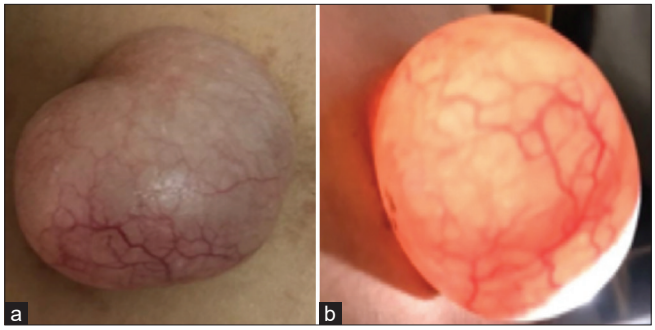

**Figure 1:** (a) Posterior view of the thoracic meningocele, (b) positive transillumination test.

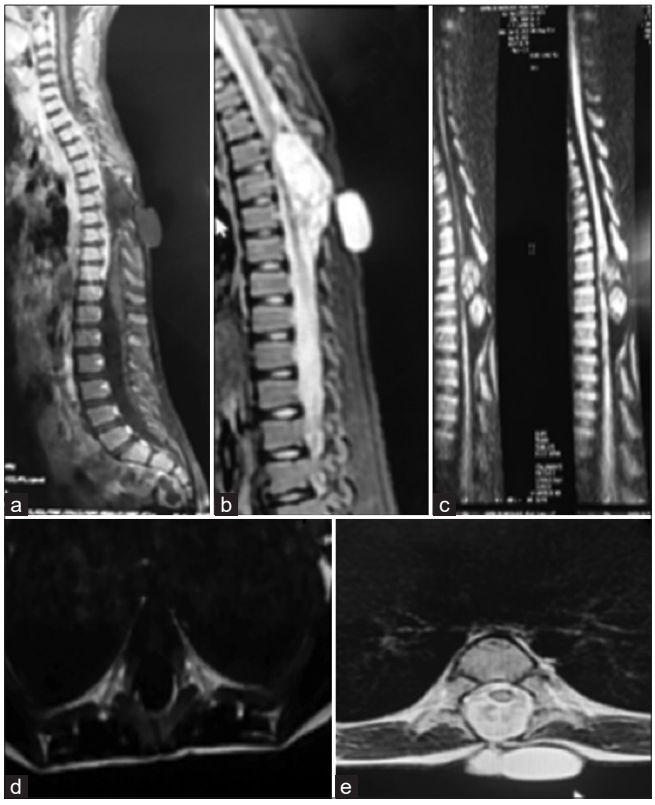

**Figure 2:** (a) Medullary magnetic resonance imaging (MRI) on a sagittal section in T1-weighted sequence ,(b) T2 sequence, (c) and short-tau inversion-recovery showing an extramedullary intradural cyst from D6 to D8 in T1 hyposignal and T2 hypersignal. (d) Spinal cord MRI on axial section in T1-weighted sequence objecting the extramedullary intradural cystic lesion in T1 hyposignal, (e) T2 sequence showing the subcutaneous fluid formation in T2 hypersignal at the level of D8-D9 communicating with the subarachnoid spaces.

### Surgery

During surgery, the LDM was initially punctured, followed by a D6–D7 laminectomy. The LDM stalk extended deep through a midline defect into the ligamentum flavum and through the dura. Intradurally, the stalk was divided [Figure 3a]. The cutaneous lesion, a thin rim of normal skin, and the superficial part of the stalk were removed *en bloc*. Two other extramedullary oval lesions were also encountered. The lesions were pearly white with a transparent avascular capsule containing a white, waxy, and flaky material; there were no apparent skin appendages. The two lesions were found immediately on the posterior surface of the spinal cord, gently removed, and the cord decompressed [Figures 3b and c].

### Postoperative course

The postoperative course was uneventful, and the patient was discharged 10 days following surgery. The patient remained neurologically intact, and no postoperative cerebrospinal fluid fistula developed. Histopathology confirmed the diagnosis of an EC [Figures 4 and 5]. At 1-year follow-up, control MRI showed no recurrence [Figure 5].

## DISCUSSION

### Frequency and location of ECs

The term "tumor perlée" (pearly tumors) was coined by Croveilhier in 1835 to describe the first EC.[9] Spinal ECs account for <1% of intraspinal tumors. They predominantly affect the lumbosacral region (60%), followed by the upper thoracic region (10%) and cervical areas (5%).[8] These cysts can manifest as either congenital or acquired, with our patient's case presumed to be congenital.[1]

### LDM

LDM represents a rare form of closed spinal dysraphism often associated with various congenital anomalies such as tethered cord, lipoma, lipomyelomeningocele, split cord malformation, and teratoma, among others.[3]

### MRI diagnosis of EC and LDM

On MRI, ECs exhibit variable characteristics. They typically appear isointense/hypointense on T1-weighted images and hyperintense on T2-weighted images, with limited enhancement under contrast. However, the presence of a thin rim of enhancement on gadolinium-enhanced MRI images has been inconsistently described in the literature.[6,7,9]

### Surgery for EC

Surgical resection remains the cornerstone of EC treatment. Aimed at achieving gross total resection whenever feasible, as subtotal resections are associated with a higher risk of recurrence (reported at 11% by Sîrbu *et al*.[8]). The removal of the cyst capsule is crucial in

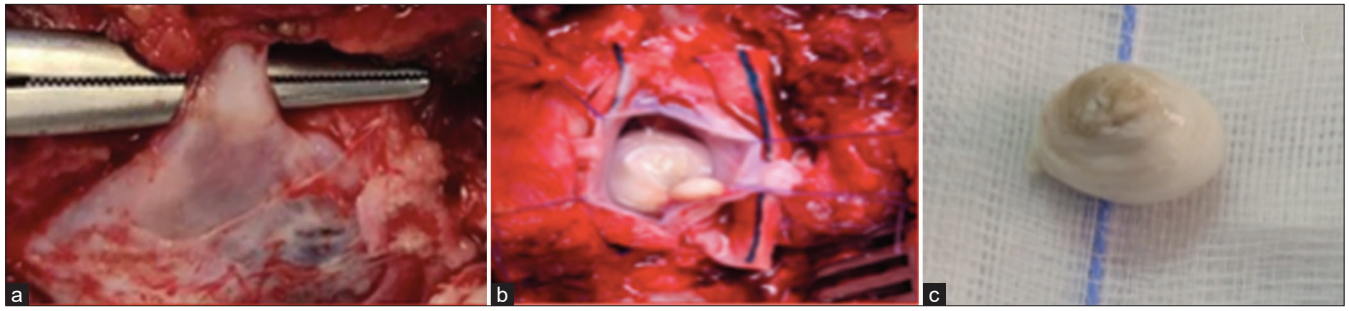


**Figure 3:** (a) Band of communication of the meningocele with the mass subcutaneously,(b) epidermoid cyst in extramedullary intradural after opening of the dura, (c) epidermoid cyst in candle wax appearance after resection.

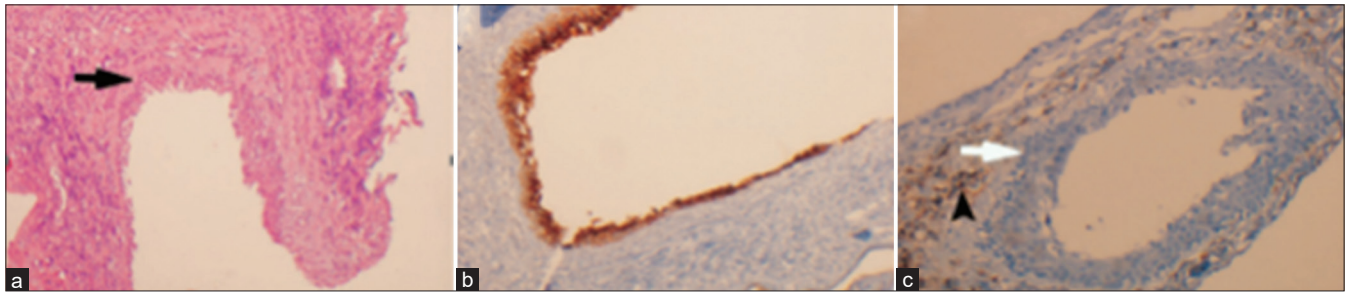


**Figure 4:** In the histopathological examination, (a) cystic lesion with stratified squamous epithelium (black arrow) (hematoxylin-eosin, ×100). (b) Stained by leukocyte common antigen (LCA), around the cystic lesion with stratified squamous epithelium (LCA, ×100). (c) Stratified squamous epithelium stained with pancytokeratin. Desquamation of keratin from the epithelial lining can be observed (pancytokeratin, ×100) (White arrow), inflammatory cells (black arrowhead).

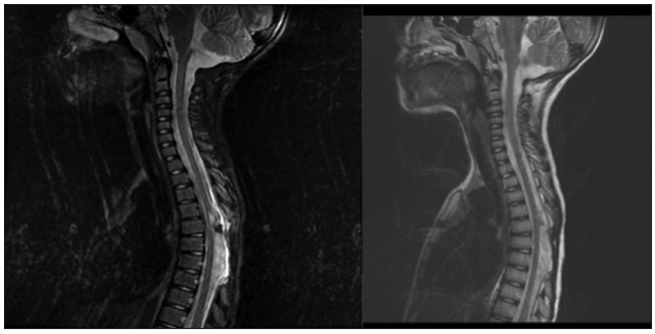

**Figure 5:** At 1-year follow-up, control magnetic resonance imaging of the spine showed no recurrence.

reducing recurrence rates.[10] However, in cases where the capsule is thin and tightly adherent to or within the spinal cord, surgeons often opt to leave remnants to prevent cord damage. Radiotherapy has also shown efficacy, as reported by Bretz *et al*., who successfully treated cervical ECs with this modality.[2]

### Prognosis

Benign ECs generally have a favorable prognosis, particularly in younger patients at the time of diagnosis.[10] Early and appropriate surgical intervention, focusing on total cyst removal and dysraphism repair, leads to optimal outcomes.

## CONCLUSION

Despite EC being benign and slow-growing lesions, early diagnosis and gross total excision achieves the best results with/without LDM.

### Ethical approval

The Institutional Review Board approval is not required.

### Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent.

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### Conflicts of interest

There are no conflicts of interest.

### Use of artificial intelligence (AI)-assisted technology for manuscript preparation

The authors confirm that there was no use of artificial intelligence (AI)-assisted technology for assisting in the

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