



## Case Report

### Corresponding Author

J.K.B.C. Parthiban

<https://orcid.org/0000-0002-6142-3424>

Department of Neurosurgery, Kovai  
Medical Center and Hospital, Coimbatore  
641014, India

Tel: +91-9842223500

Fax: +91-4222627782

E-mail: [juttyparthiban@gmail.com](mailto:juttyparthiban@gmail.com)

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# Lumbar Intramedullary Epidermoid Following Repair of Sacral Myelomeningocele and Tethered Cord: A Case Report With a Review of the Relevant Literature and Operative Nuances

S.J. Balaparameswara Rao, Hitesh Bhaliya, J.K.B.C. Parthiban

Department of Neurosurgery, Kovai Medical Center and Hospital, Coimbatore, India

Epidermoid cysts of the spine are rare tumors. While the majority of them occur spontaneously, in very few cases, they can occur following previous surgery for spinal dysraphism. Such tumors tend to occur at the site of previous surgery. The occurrence of an epidermoid cyst at a level higher than the previous surgical site is a rare entity. We present a rare case of a lumbar intramedullary and extramedullary epidermoid occurring at a level higher than the previous surgical site, along with a discussion of the causes of such an occurrence and operative nuances regarding the management of an intramedullary epidermoid in a pediatric patient.

**Keywords:** Epidermoid cyst, Intramedullary epidermoid, Spinal dysraphism

## INTRODUCTION

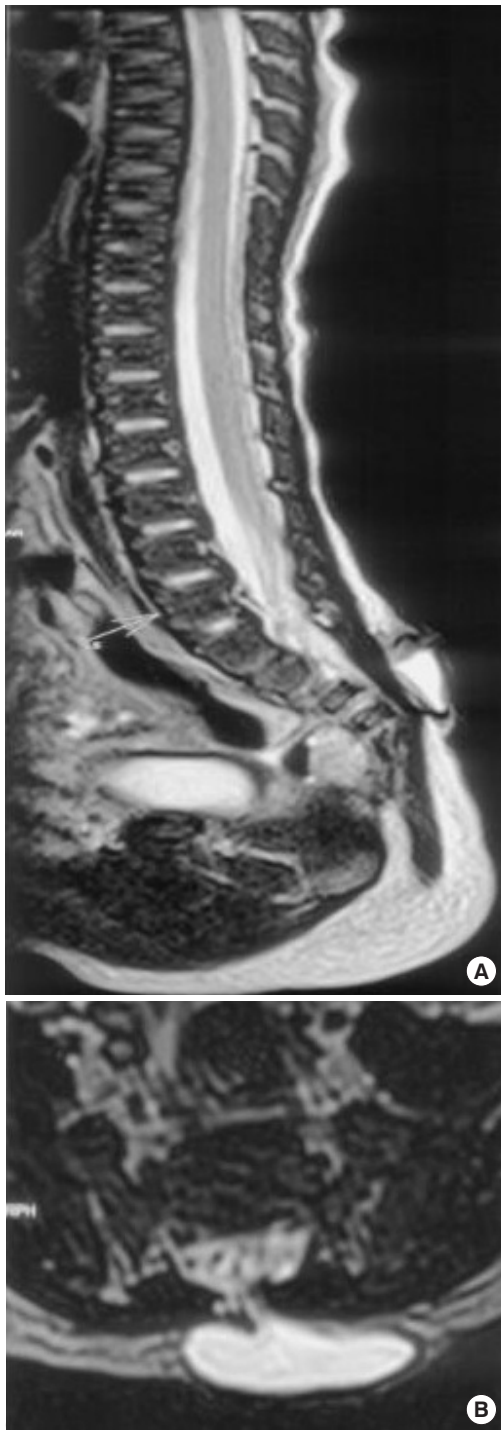
Intraspinous epidermoid cysts are rare tumors. While most of them occur spontaneously, very few develop following surgery for spinal dysraphism. Such tumors tend to develop and present in a delayed manner, as long as 38 years after surgery.<sup>1</sup> However most of these tumors occur at the level of previous surgery. We present a case of intramedullary and extramedullary septated cystic epidermoid tumor presenting 4 years following repair of sacral myelomeningocele with tethered cord at a level higher than the first surgery along with a review of literature highlighting the possible causes of such occurrence.

## CASE REPORT

A 1-month-old male child was brought with a translucent swelling at the back since birth. On magnetic resonance imag-

ing (MRI), there was evidence of a sacral spinal dysraphism with cerebrospinal fluid outpouching suggestive of a sacral myelomeningocele with tethering noted at the lower end of the cord (Fig. 1). The child underwent repair of the sacral myelomeningocele with sac excision and detethering of cord at the age of 1 month. Closure of the defect was done with advancement flaps using multiple Z plasty technique. Postoperatively, child developed pyogenic meningitis and urinary tract infection which was treated with intravenous antibiotics. The child recovered completely and had normal developmental milestones and could walk normally and was on regular follow-up for 1 year.

The child now presented at the age of 4 years with complaints of fever since 7 days, weakness of both lower limbs with difficulty in walking and urinary disturbance for 4 days. On examination, tone was decreased in both lower limbs with decreased deep tendon reflexes. On evaluation for fever, the child was di-



**Fig. 1.** Magnetic resonance imaging showing a sacral myelomeningocele in sagittal (A) and axial (B) planes. Arrow indicates level of tethered cord at first presentation.

agnosed to have urinary tract infection and was started with appropriate antibiotics. On ultrasonogram, child had a significant postvoid residual urine volume of 73 mL. On further evaluation with an MRI, bulbous dilatation of the lower end of spi-

nal cord was present with 3 well defined peripherally enhancing septated cystic lesions at L3–5 levels. Lower end of the cord could not be identified separately from the upper 2 cystic lesions, which thus appeared to be intramedullary. The intramedullary component showed greater peripheral enhancement indicating a thicker capsule with greater adhesions as per the senior author. There was a syrinx from D6–9 levels (Fig. 2). In view of fever and short duration of symptoms, we considered the possibilities of an abscess or an infected dermoid/epidermoid cyst.

The patient underwent L3–5 suspension laminoplasty and excision of intradural lesion. The cystic lesions had a capsule with pultaceous, sebum like material emanating from the capsule. The nerve roots of cauda equina were pushed laterally to the left by the lesion. The intramedullary component within the conus was also decompressed. However, attempts to dissect the capsule within the conus caused hyperemic changes due to adhesions as was assessed based on the preoperative MRI. Hence the capsule within the conus was left behind. The capsule of the extramedullary lesion could be dissected and excised (Fig. 3). Some of the cystic contents were sent for culture in view of suspicion of an infected lesion. Dura was closed primarily.

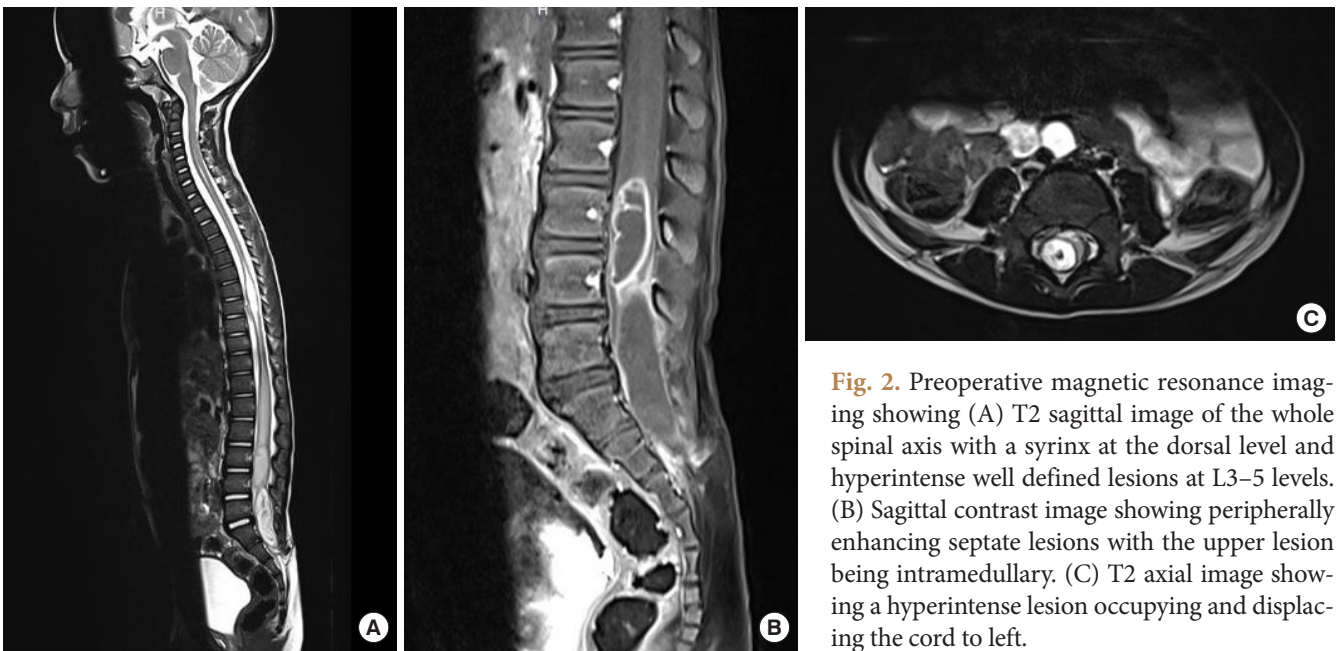
The patient recovered uneventfully in the postoperative period. The child could walk comfortably from the first postoperative day and could void adequately. Lower limb weakness had improved. Urinary tract infection was treated with antibiotics. Cystic contents were sterile. On histopathological examination, specimen showed keratin flakes and strips of stratified squamous epithelium with focal areas of macrophage collections and granulation tissue (Fig. 4). No adnexal structures were seen. These findings were consistent with an epidermoid cyst.

Follow-up MRI done after 4 weeks, showed residual enhancing cyst wall of the intramedullary component within the conus with complete excision of the extramedullary component. As compared to the preoperative MRI, the residual intramedullary cyst wall and cavity were smaller in size (Fig. 5). Child was doing well with no neurological deficits.

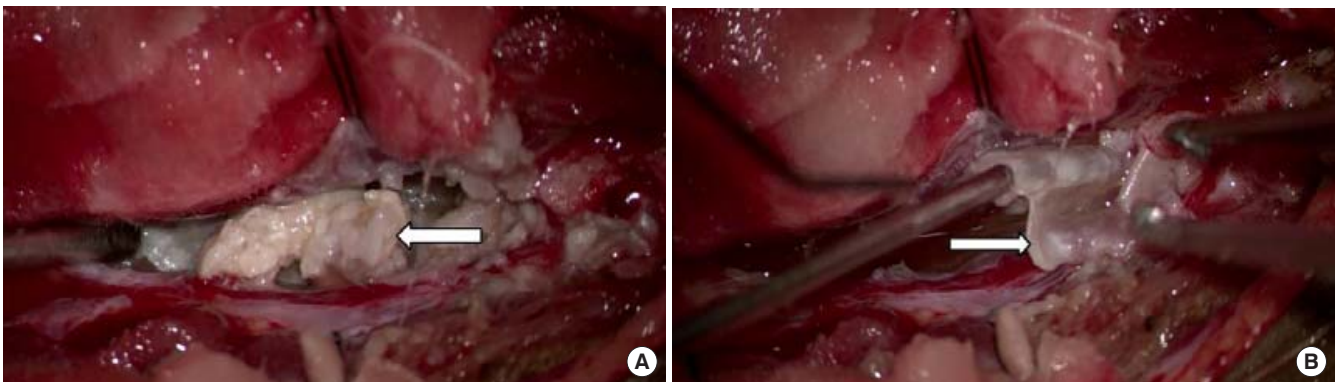
Informed consent to publish the details of the case has been obtained from the parents.

## DISCUSSION

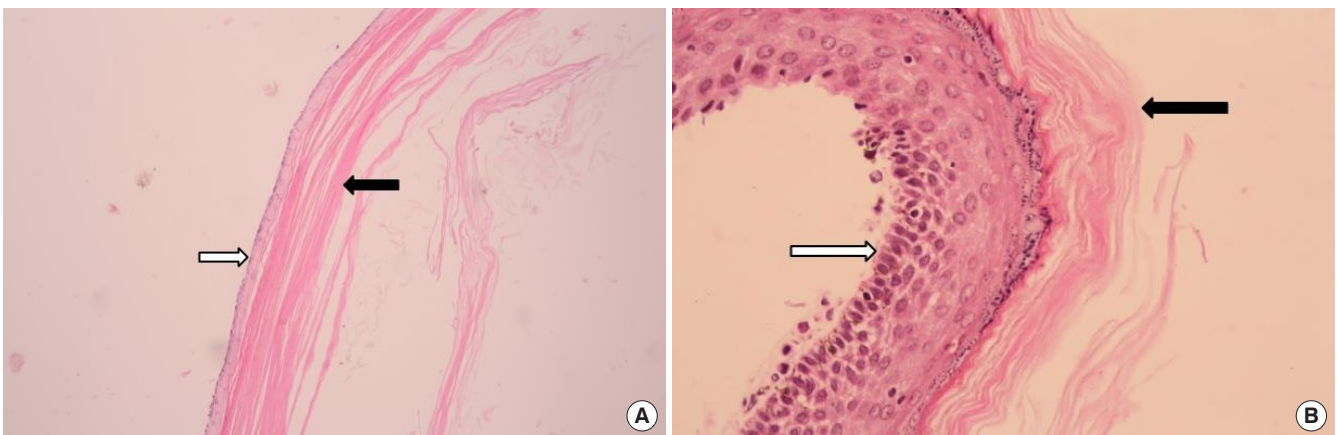
Spinal epidermoids are rare and constitute only 0.5%–1% of all spinal tumors and 1.8% of all intramedullary tumors.<sup>2</sup> These tumors can be congenital or may be due to remnants or implantation of cutaneous elements during lumbar puncture<sup>3-5</sup> or surgery for spinal dysraphism including repair of myelomeningocele.<sup>6</sup>



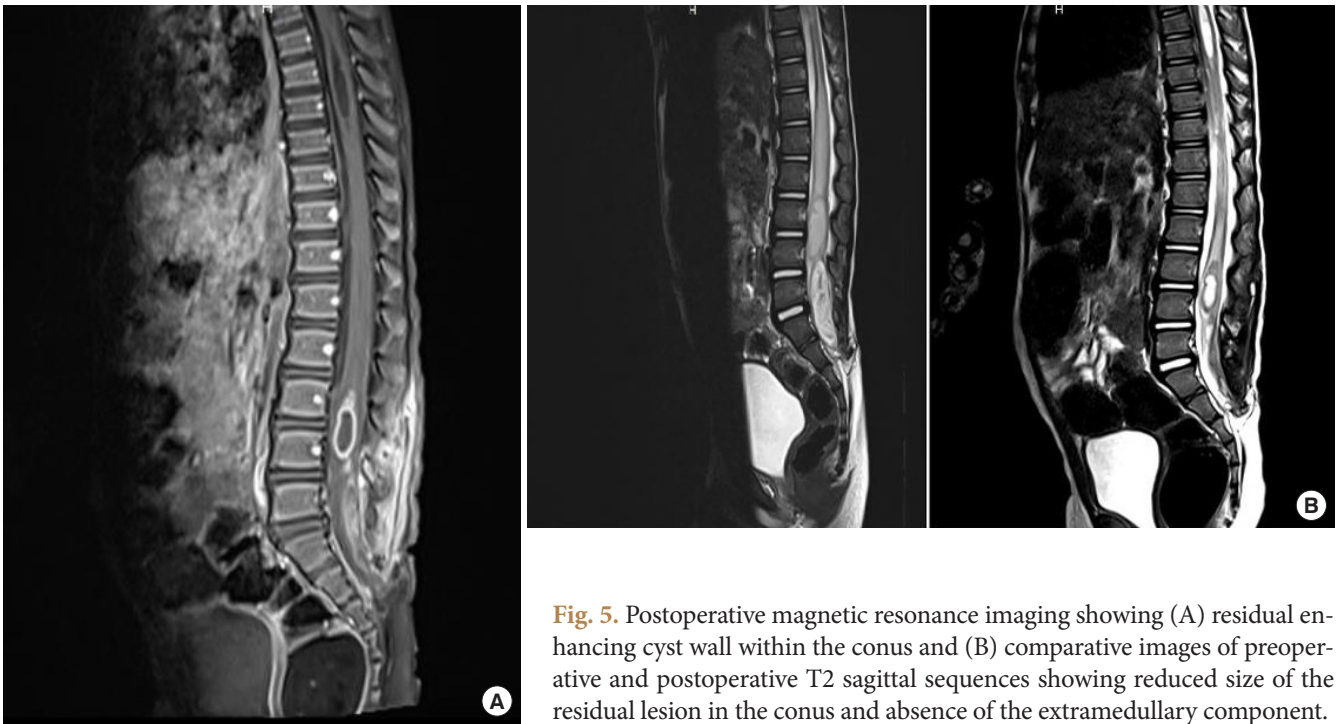
**Fig. 2.** Preoperative magnetic resonance imaging showing (A) T2 sagittal image of the whole spinal axis with a syrinx at the dorsal level and hyperintense well defined lesions at L3-5 levels. (B) Sagittal contrast image showing peripherally enhancing septate lesions with the upper lesion being intramedullary. (C) T2 axial image showing a hyperintense lesion occupying and displacing the cord to left.



**Fig. 3.** Intraoperative images: (A) pultaceous cystic contents being removed (arrow), (B) cyst wall being dissected (arrow).



**Fig. 4.** Histopathological examination of hematoxylin and eosin stained sections. (A) Microscopic image and (B) magnified view showing stratified squamous epithelium (white arrow) with underlying keratin flakes (black arrow) and absence of adnexal structures.



**Fig. 5.** Postoperative magnetic resonance imaging showing (A) residual enhancing cyst wall within the conus and (B) comparative images of preoperative and postoperative T2 sagittal sequences showing reduced size of the residual lesion in the conus and absence of the extramedullary component.

The etiology for occurrence of these tumors following surgery for myelomeningocele and tethered cord have been a matter of debate. Scott et al.<sup>6</sup> classified the etiology of these lesions according to their location into 2 groups. Those located posterior to the cord were considered to be due to implantation at the time of initial repair while those located anterior to the cord were considered to represent components of neural tube malformation. Storrs<sup>7</sup> noted that hamartomatous elements are relatively common in the myelodysplastic sequence. Microscopic dermal and epidermal rests were discovered in many myelomeningoceles at the periphery of the neural placode as well as in tethered cord specimens.

Our patients developed urinary tract infection along with features suggestive of meningitis following first surgery which was confirmed by a lumbar puncture. Though there is no correlation between postoperative meningitis and development of a secondary epidermoid, lumbar puncture is known to have a causal role. However, the occurrence of intramedullary component cannot be explained by lumbar puncture alone. Since microscopic examination of both the myelomeningocele sac and tethered cord segment did not show evidence of epidermal cell rests, we consider implantation of epidermal elements at the time of initial repair to be the likely cause of the epidermoid. Occurrence of the epidermoid at a higher level can be explained due to ascent of the cord following detethering.

Since these tumors grow slowly, they usually present in a delayed manner. Case reports have been published with presentations as late as 15 years and 38 years following the first surgery.<sup>1,8</sup> MRI is a useful tool for diagnosis. Early surgical excision is the treatment of choice. Literature regarding recurrence of spinal epidermoids after complete excision is sparse. Lunardi et al.<sup>9</sup> reported no recurrence in their series of 8 patients followed over a period of 5 to 30 years. However, it is difficult to achieve complete excision along with cyst capsule in intramedullary epidermoids due to dense adhesions. Hence subtotal excision is more commonly performed to avoid neural damage.<sup>10,11</sup> Radiotherapy can play a role to arrest growth in case of a recurrent epidermoid.<sup>12</sup>

Our patient presented with features of cauda equina syndrome 4 years following first surgery. The location of tumor was also slightly higher than the previous surgical site. The tumor was partly extramedullary and partly intramedullary within the conus. Though the extramedullary component could be excised completely, the cyst wall of the intramedullary component had to be left behind. We did not consider the option of radiotherapy to prevent neural regression in a growing child.

## CONCLUSION

Intraspinal epidermoids though rare, are known to occur fol-

lowing surgery for myelomeningocele or tethered cord. With the etiology being unclear, whether it is due to implantation or due to hamartomatous cell rests, approaches to prevent their occurrence is not possible. These cases require regular clinical as well as a radiological follow-up to detect an early occurrence. A safe surgical excision is the treatment of choice. Further treatment following a subtotal excision needs to be tailored according to the age of the patient.

## CONFLICT OF INTEREST

The authors have nothing to disclose.

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