Egyptian Journal of Radiology and Nuclear Medicine

CASE REPORT Open Access

Unusual cause of incomplete cauda equina syndrome: dermoid cyst with a split cord malformation case report

Hamza Retal[1*], Soumya EL Graini[1], Hafsa EL Ouazzani[2], Nadia Cherradi[2], Meriem Fikri[1], Najwa Echcherif El Kettani[1], Mohamed Jiddane[1] and Firdaous Touarsa[1]

Abstract

Cauda equina syndrome is a surgical emergency caused by the compression of the lumbosacral nerves. The most frequent cause is discal herniation, while tumoral pathology in this region is rare, and congenital processes are less common. We report the case of a young 37-year-old male patient, admitted to the emergency room with an incomplete cauda equina syndrome. As medical history, a lumbar surgery 15 years ago was reported with no documentation. An MRI was performed, revealing a mass within the filum terminal exhibiting three components suggestive of a dermoid cyst accompanying a split cord malformation, which was confirmed by pathology. Various disorders may be associated to intradural dermoid cyst, such as lipoma, low-lying cord, hydromyelia or thick filum. Surgery is the treatment of choice, and MRI plays a crucial role in detecting associated abnormalities and determining surgery priorities.

Keywords Cauda equina syndrome, Dermoid cyst, Split cord malformation, Spinal cord MRI, Spine surgery, Intramedullary lesion

Introduction

Dermoid cyst is a congenital benign condition that manifests in various scenarios. The intraspinal location accounts for 0.8–1.1% of all intraspinal benign tumors being reported with a greater prevalence observed in the pediatric population comprising 5%–17% of intradural lesions in some series. It is often situated in the sacrolumbar region and represents rare causes of cauda equina syndrome, as the symptoms are generally progressive, manifesting with pain and paresthesia [1]. Furthermore, the mass may coexist with other congenital disorders such as split cord malformation. A higher incidence of dermoid cyst has been described in patients with spina bifida, dermal sinuses, myelomeningocele and syringomyelia [2, 3].

Surgical management can be challenging due to the risk of recurrence and the frequent associations, underscoring the necessity of MRI. This imaging modality is indispensable for evaluating its extent and identifying other accompanying congenital disorders before any treatment [4].

Case presentation

We report the case of a 37-year-old male patient with a history of urethral stenosis and lumbar surgery performed 15 years ago for undetermined cause, lacking corresponding documentation. Subsequent to the procedure, the patient was lost to follow-up. Over the following 10 years, he endured chronic low back pain coupled with progressive weakness in the lower extremities. This condition steadily worsened, prompting the patient to

*Correspondence:
Hamza Retal
Retal.hamza95@gmail.com
[1] Neuroradiology department, Specialties Hospital of Rabat, Mohammed V University, Rabat, Morocco
[2] Anatomic Pathology Laboratory Department, of Specialties Hospital of Rabat, Mohammed V University, Rabat, Morocco

seek consultation at the emergency room due to further deterioration of his condition.

The clinical examination revealed a conscious patient with preserved walking ability but unable to stand on tiptoes, exhibiting an incomplete plantar flexion deficit. Bilateral and symmetrical power decrease to 4/5 was observed in the lower limbs, accompanied by a slight reduction in patellar reflexes and saddle anesthesia. No sphincter disorders were detected upon examination.

The patient underwent an immediate spinal cord MRI due to the urgent compressive situation. A 1.5 Tesla MRI machine equipped with dedicated phase-array coil antenna was utilized. The MRI findings unveiled an intradural ceriths-like process straddling the terminal conus of the spinal cord and cauda equina, at L1/L2 level, measuring 19×28 mm in axial plane and 78 m in height. The lesion appeared oval-shaped with lobular contours, displaying three components. It predominantly shows a cystic nature, appearing hyperintense on T2-weighted sequences and hypointense on T1-weighted images. This lesion contained an upper heterogeneous peripheral fatty area that appeared hyperintense on both T1- and T2-weighted images, suppressed after fat saturation (Fig. 1).

Additionally, a ring-like enhancement of the wall and septa was observed after an IV injection of 7, 5 ml of Gadobutrol (Gadovist), equivalent to a dose of 0, 1 mmol/kg (Fig. 2).

The process is leading to a slight scalloping and enlargement of vertebral bodies in front of the lesion, delineating a segmentary bifurcation of the medullary cone sharing the same dural sac with no spur between the two hemicords, related to an adjacent split cord malformation (Fig. 3).

Signs of previous L2–L3 laminectomy were evident, disclosing the site of the prior surgical intervention.

A complete resection was carried out through a posterior medial incision at the L1/L2 level involving dissection and laminectomy, which revealed a yellowish, soft mass containing strands of hair. Subsequent anatomical pathology study confirmed the diagnosis of a dermoid cyst by revealing a foreign body reaction in association to sebaceous appendage (Fig. 4).

At 6 months following the surgery, the patient reported persistent paresthesia of the lower limbs. However, he demonstrated clinical improvements, with regained mobility and a reduction in lumbar pain.

Discussion

Cauda equina syndrome (CES) is an emergency condition with devastating consequences. It is due to dysfunction of the sacral and lumbar nerve roots in the spinal

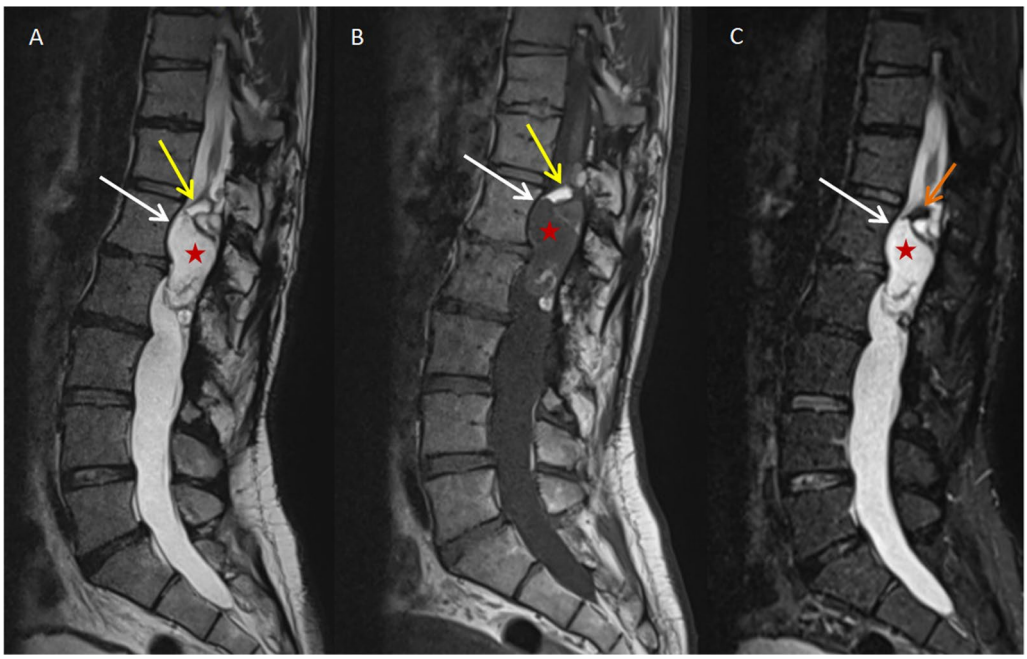

Fig. 1 Sagittal MR images showing a mass within the cauda equina at the level of L1–L2. **A** Sagittal T2-weighted MRI sequence showing a mass (white arrow) within the cauda equina at the level of L1–L2, containing a cystic component (red star) and a fatty portion (yellow arrow), both displaying hyperintense signals. **B** Sagittal T1-weighted MRI sequence of the same mass (white arrow) within the cauda equina confirming the cystic nature (red star) as hypointense, with the fatty portion (yellow arrow) remaining hyperintense. **C** Short tau inversion recovery (STIR) sequence confirming the fatty nature of the upper component of the mass (orange arrow)

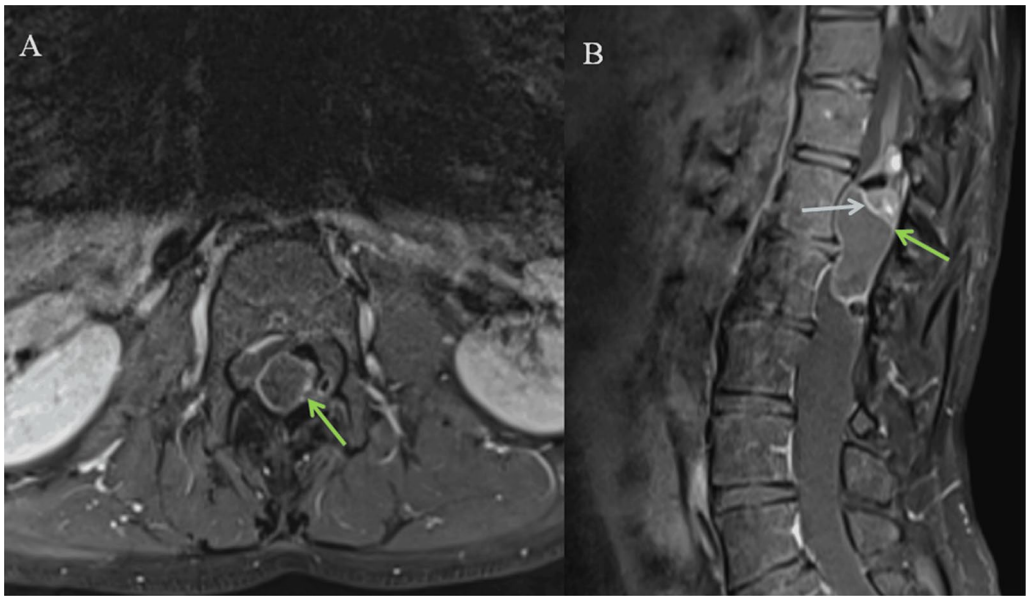

Fig. 2 Enhanced MRI sequences of the lumbar spine showing the cauda equina mass. **A** Axial enhanced T1-weighted sequence after Gadolinium injection, exhibiting enhancement of the contours (green arrow) and septa (gray arrow) of the mass. **B** Sagittal enhanced T1-weighted sequence after Gadolinium injection, showing the peripheral enhancement of the mass (green arrow)

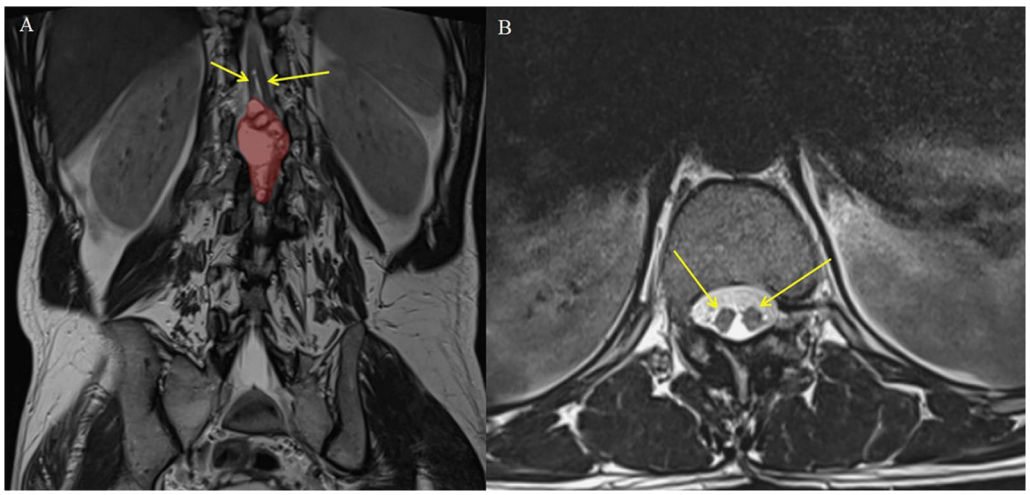

Fig. 3 T2-weighted sequences showing a dermoid cyst in the cauda equina associated with a split cord. **A** Coronal sequence demonstrating a ceriths-like mass of the cauda equina (red surface) located just below a split medullar cord (yellow arrow). **B** Axial sequence showing the diastematomyelic spinal cord (yellow arrow) within the same dural sac, without evidence of an osseous spur

canal, causing bladder, bowel or sexual dysfunction and perianal or saddle numbness that requires immediate intervention [5].

Other possible symptoms include back pain with or without radiculopathy, altered sensation or numbness in the lower extremities, weakness with decreased or absent reflexes in the lower extremities, depending on the underlying cause. Incomplete forms can show only saddle anesthesia with lack of bladder and bowel dysfunction [5].

The most common causes of cauda equina syndrome remain discal herniation, traumatic injury or tumoral disease. Malformative etiologies leading to this syndrome are rare. Spinal dermoid cysts represent one such rarity, accounting for 0.8%–1.1% of all primary spinal tumors. They typically manifest in the extra medullary or

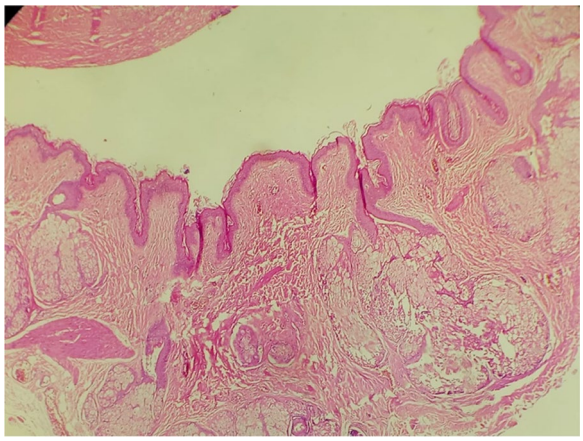

Fig. 4 Postoperative histopathology, microphotograph demonstrating a dermoid cyst lined with squamous epithelium, associated with sebaceous structures. Hematoxylin and eosin stain, magnification × 10 (HE × 10)

subdural juxtamedullary lumbosacral region and cauda equina, sometimes occurring in both locations [1].

Various hypotheses have been put forward to elucidate the development of dermoid cysts in the spine, which can be either congenital or acquired. Congenital dermoid is believed arise from embryological errors during neural tube closure, potentially explaining the associations with spinal developmental abnormalities discussed further. If dermal invagination into the neural canal occurs early in embryonic life, a dermoid tumor will result, if this occurs at a later stage, an epidermoid tumor will develop [6]. Conversely, some dermoid cysts are acquired and may develop following a lumbar puncture or injury [7].

The slow-growing characteristic of intramedullary dermoid cyst can make it asymptomatic with delayed diagnosis until the occurrence of nerve compression symptoms, or it can manifest within complications such as ruptured cyst, infection and abscess formation or, more rarely compressive hydrocephalus [8, 9].

MRI is the modality of choice for spinal cord exploration. Dermoid cysts are characterized by a heterogeneous appearance due to their mixed components, including tissue, cystic fluid and fat, often accompanied by calcifications. These features results in a varied signal on T1- and T2-weighted sequences with suppression of fatty portions on fat saturation sequences, and without restriction on diffusion-weighted sequences. Following Gadolinium injection, dermoid cysts typically exhibit either peripheral ring-like enhancement as seen in our case, or no enhancement, attributed to the presence of blood vessels in the connective tissue surrounding the tumor, which are absent within the epithelial wall of the dermoid [1, 3]. Enhanced mass appearance may suggest a complicated infected cyst, particularly if associated with a dermal sinus, which in rare cases can lead to abscess formation [9]. In some cases, the conus medullaris shows punctate images in T1 and T2 hypersignal arranged in a string of beads along the ependymal canal, this likely corresponds to lipid droplets found in cases of dermoid cyst rupture, as evidenced by enhancement of the ependymal wall highlighting meningeal inflammation [8].

MRI study can reveal associated subtle abnormalities. The association of dermoid cyst and split cord malformation is described in almost 13% patients with split cord malformations (SCM) according to a Turkish retrospective case review by E. Ozturk et al. [2]. Additional disorders can coexist such as low-lying cord, hydromyelia, meningoencephaloceles, thick filum, lipoma, epidermoid tumors or neuroenteric cyst [10]. When split cord is associated, it is capital to specify the type either I or II, for effective surgery management. In type I, the two hemicords are housed each in its own dural sac separated by a rigid osteocartilaginous septum which can be complete or incomplete. In contrast, in type II, both hemicords are housed within a common dural sac with no spur between the two hemicords [11].

CT can be more effective in evaluation of osseous spine structures, as observed in our case with an enlargement of the spinal canal, likely a response to slow tumor growth or associated imperfect midline fusion during development. However, MRI remains indispensable.

The main differential diagnoses of dermoid cysts in spinal cord on imaging include lesions with high lipid content such as lipomas and ependymomas. Ependymomas tend to be well-circumscribed on imaging and typically exhibit a low signal intensity on T1-weighted and high signal intensity on T2-weighted sequences. They often demonstrate homogeneous enhancement, in contrast with dermoid cyst. Additionally, they may contain hemosiderin deposits, related to hemorrhage, which can be mistaken for calcifications on MRI. Lipomas, on the other hand, are more homogenous fatty lesions, without cystic components or calcifications on imaging features [12]. Epidermoid tumors are usually reported to show low intensity on T1-weighted sequence and high intensity on T2-weighted sequence, similar to cerebrospinal fluid (CSF) unlike dermoids containing fat component [5]. The challenge of diagnosis may be encountered when facing an infected enhancing cyst. The presence of an abscess or a dermal sinus can redirect the correct diagnosis in association to clinical and biological findings.

The primary aim of surgery is to relieve pressure on the nerves in the cauda equina by removing the offending elements. Treatment typically involves a large laminectomy for decompression and surgical excision of the cyst. While the cystic component is generally straightforward

to remove, complete resection of the mass can potentially result in severe nerve damage due to the close connections that exist. Intraoperative considerations during cyst removal also include the potential contamination of the surgical field with spillage of cyst contents, which could lead to aseptic meningitis. Thus, achieving radical removal of the cyst is imperative to prevent postoperative chemical meningitis, which may arise from residual tumor material. Additionally, spinal deformity represents a common and significant complication associated with spinal surgery, warranting careful consideration [4]. Given the benign nature of these tumors and the inherent risks related to surgery, some authors advocate conservative management for asymptomatic lesions [13, 14].

When split cord and dermoid cyst are accompanied by other disorders, determining surgical priorities becomes crucial for simultaneous treatment of all the associated pathologies. In case of a tethered cord or a septum is present, these abnormalities should be excised first to reduce the traction on the conus. Otherwise, a sudden manipulation of the cord can result in spinal cord injury [15, 16]. Additionally, other associations, such as dermal sinus which is related to a high risk of infections, or spina bifida and meningomyelocele, should be addressed concurrently. Preoperative MRI assessment is essential for evaluating these conditions [4].

Given the high risk of intraoperative injuries related to surgery, intraoperative monitoring has been proposed as a valuable tool to protect neural structures during spinal surgery by recording a combination of electromyogram (EMG) and somatosensory evoked potential's activity (SSEP), which reflects the integrity of afferent and efferent pathways [4].

Dermoid cysts are renowned for their potential high risk of recurrence. The presence of residual cyst wall has been linked to postoperative recurrence, emphasizing the recommendation for total resection surgery [14, 15]. Additionally, although extremely rare, malignant transformation to squamous cell carcinoma has been described in spinal dermoids [17].

Assessing functional outcomes is crucial in managing spinal cord tumors due to their impact on neurological functions. Enhancing these outcomes improves patients' quality of life and a multidisciplinary approach helps optimize patient care. Postoperative improvements in pain symptoms and Frankel scores indicate a significant enhancement in functional outcomes after surgery [18].

In our case, complete surgery yielded gratifying results with a decrease in symptoms and no complications, given the absence of malformative associations heightening surgical-related risks. In other cases studies involving surgery for dermoid cysts, outcomes were similarly gratifying, with pain relief and neurological improvement observed. Notably, only one patient out of 26 experienced moderate deterioration in the postoperative period in a study led by Liu et al. Nevertheless, routine follow-up imaging is recommended, with a yearly MRI assessment, given the high risk of recurrence even before clinical finings appears [4].

Conclusion

Intradural dermoid cysts, which are uncommon sources of medullary masses causing cauda equina syndrome, are often linked to pre-existing malformations. MRI remains the preferred examination for diagnosis, playing an essential role in detecting associated lesions. Understanding these lesions is crucial for determining the surgical approach and strategy. Complete resection is always performed due to the high risk of recurrence associated with medullary dermoid cyst.

Abbreviations

MRI	Magnetic resonance imaging
CES	Cauda equina syndrome
SCM	Split cord malformations
CT	Computed tomography
CSF	Cerebrospinal fluid
EMG	Electromyogram
SSEP	Somatosensory evoked potential's activity

Author contributions

Hamza Retal helped in conception of the work, design of the work and acquisition of data. Soumya El Graini helped in acquisition of data. Hafsa El Ouazzani helped in acquisition of data. NADIA CHERRADI helped in acquisition of data. Meriem Fikri revising the work critically for important intellectual content. Najwa Echcherif El Kettani revising the work critically for important intellectual content. Mohamed Jiddane revising the work critically for important intellectual content. Firdaous Touarsa revising the work critically for important intellectual content and final approval of the version to be published.

Funding

The author(s) received no financial support for the research, authorship and/or publication of this article.

Declarations

Ethics approval and consent to participate

We confirm that our work has been conducted with the ethical approval of our institution.

Consent for publication

Written informed consent was obtained from the patient to be published in this article.

Availability of data and materials

All data generated or analyzed during this study are included in this published article.

Competing interests

The authors declared no potential conflicts of interest with respect to the research, authorship and/publication of this article.

Received: 10 July 2024 Accepted: 9 August 2024
Published online: 19 August 2024

References

1. Falavigna A, Righesso O, Teles AR (2009) Concomitant dermoid cysts of conus medullaris and cauda equina. Arq Neuropsiquiatr 67:293–296. https://doi.org/10.1590/S0004-282X2009000200023
2. Ozturk E, Sonmez G, Mutlu H et al (2008) Split-cord malformation and accompanying anomalies. J Neuroradiol 35:150–156. https://doi.org/10.1016/j.neurad.2007.11.006
3. Arseni C, Dănăilă L, Constantinescu A et al (1977) Spinal dermoid tumors. Neurochirurgia. https://doi.org/10.1055/s-0028-1090363
4. Liu H, Zhang JN, Zhu T (2012) Microsurgical treatment of spinal epidermoid and dermoid cysts in the lumbosacral region. J Clin Neurosci 19:712–717. https://doi.org/10.1016/j.jocn.2011.07.046
5. McNamee J, Flynn P, O'Leary S, Love M, Kelly B (2013) Imaging in cauda equina syndrome—a pictorial review. Ulster Med J 82(2):100
6. Manno NJ, Uihlein A, Kernohan JW et al (1962) Intraspinal epidermoids. J Neurosurg 19:754–765. https://doi.org/10.3171/jns.1962.19.9.0754
7. Herman JM, McLone DG, Storrs BB, el al. (1993) Analysis of 153 patients with myelomeningocele or spinal lipoma reoperated upon for a tethered cord: presentation, management and outcome. Pediatr Neurosurg 19:243–249. https://doi.org/10.1159/000120739
8. Jidal M, Elkharras A, Aziz N et al (2005) Intramedullary rupture of a dermoid cyst of the conus: a case report. J Radiol 86:498–501. https://doi.org/10.1016/s0221-0363(05)81397-6
9. Vadivelu S, Desai SK, Illner A et al (2014) Infected lumbar dermoid cyst mimicking intramedullary spinal cord tumor: observations and outcomes. J Pediatr Neurosci 9:21. https://doi.org/10.4103/1817-1745.131475
10. Ghimire S, Shrestha S, Shrestha D, el al. (2024) Contiguous diastematomyelia with tethered cord, intradural extramedullary dermoid tumor, and lipomyelomeningocele: a unique case of spinal dysraphism. Clin Case Rep 12(4):e8783. https://doi.org/10.1002/ccr3.8783
11. Pang D, Dias MS, Ahab-Barmada M (1992) Split cord malformation: part I: a unified theory of embryogenesis for double spinal cord malformations. Neurosurgery 31:451–480. https://doi.org/10.1227/00006123-199209000-00010
12. Koeller KK, Rosenblum RS, Morrison AL (2000) Neoplasms of the spinal cord and filum terminale: radiologic-pathologic correlation. Radiographics 20:1721–1749. https://doi.org/10.1148/radiographics.20.6.g00nv151721
13. Bailey IC (1970) Dermoid tumors of the spinal cord. J Neurosurg 33:676–681. https://doi.org/10.3171/jns.1970.33.6.0676
14. Guo S, Xing Y (2014) A review on five cases of intramedullary dermoid cyst. Childs Nerv Syst 30:659–664. https://doi.org/10.1007/s00381-013-2281-7
15. Mahapatra A (2011) Split cord malformation—a study of 300 cases at AIIMS 1990–2006. J Pediatr Neurosci 6:41. https://doi.org/10.4103/1817-1745.85708
16 Sinha S, Agarwal D, Mahapatra AK (2006) Split cord malformations: an experience of 203 cases. Child Nervous Syst 22:3–7. https://doi.org/10.1007/s00381-005-1145-1
17. Kudo N, Hasegawa K, Ogose A et al (2007) Malignant transformation of a lumbar intradural dermoid cyst. J Orthop Sci 12:300–302. https://doi.org/10.1007/s00776-007-1125-2
18. Randhawa AS, Srivastava A, Agrawal S, Verma JS, Sharma BS, & Sherpa TD (2024) Functional outcomes in intradural extramedullary spinal tumors. Surg Neurol Int 15. 10.25259%2FSNI_689_2023.

Publisher's Note