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INTRAMEDULLARY SPINAL DERMOID TUMOR PRESENTING WITH EQUINOVARUS DEFORMITY: A RARE PEDIATRIC CASE

Neurosurgery

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ABSTRACT

Intramedullary spinal dermoid cysts are exceptionally rare, particularly in children and in the absence of spinal dysraphism. We report the case of a 12-year-old girl who presented with progressive right foot equinovarus deformity and lower limb weakness. MRI revealed an intramedullary lesion extending from D10 to L1, and surgical exploration confirmed a dermoid cyst containing keratinous debris and hair. Subtotal excision was performed to avoid neurological injury. The case highlights the potential for atypical orthopedic presentations in spinal dermoids and emphasizes the importance of considering intramedullary lesions in children with long-standing foot deformities.

KEYWORDS

Intramedullary dermoid cyst; spinal cord tumor; pediatric neurosurgery; equinovarus deformity; thoracolumbar spine; subtotal excision; MRI spine; keratinous debris.

INTRODUCTION

Dermoid cysts are congenital, benign lesions that result from ectodermal inclusion during neural tube closure and are classified as hamartomas because of their mixture of dermal and epidermal derivatives [1]. They typically contain keratinous debris, hair follicles, sebaceous glands, and occasionally teeth or calcified material, reflecting their ectodermal origin [1].

Spinal dermoid cysts are rare, representing less than 1% of all spinal tumors, with intramedullary variants comprising only about 0.3% [1]. They may occur congenitally, due to defective neural tube closure, or acquired, following trauma, lumbar puncture, or previous spinal surgery [2]. Most lesions occur in the lumbosacral region,and are often associated with spinal dysraphism, dermal sinus tracts,or tethered cord.Although these cysts usually present in the second or third decade of life, pediatric and atypical presentations have been reported [3]. Such atypical cases are diagnostically challenging because they may present with non-specific neurological or orthopedic manifestations.

Reports of intramedullary dermoid cysts without dysraphism in children are exceedingly rare. Moreover, the association with a chronic orthopedic deformity such as equinovarus foot has not been widely described in literature.Early identification is crucial to prevent irreversible neurological damage.We present a rare dorsal intramedullary dermoid cyst in a 12-year-old girl without dysraphism,manifesting primarily as a long-standing equinovarus deformity,with clinicoradiological and histopathological correlation.

Case Presentation

A 12-year-old female, resident of Jhinjhak, Kanpur Dehat, presented with intermittent low backache for 3 years and gradually progressive right foot deformity over 2½ years.

On general examination, the patient was vitally stable. Neurological evaluation revealed bilateral lower limb weakness (power 4/5 left, 3/5 right), equinovarus deformity of the right foot, diminished reflexes (1/5), and saddle hypoesthesia with sphincter involvement. Laboratory tests were normal.MRI of the thoracolumbar spine revealed a well-defined intramedullary lesion extending from D10 to L1, causing fusiform expansion of the cord. The lesion appeared isointense to mildly hyperintense on T1-weighted sequences and predominantly hypointense with heterogeneous signal on T2-weighted sequences (Figures *1* and 2). Post-gadolinium sequences demonstrated poor enhancement. No associated spinal dysraphism, syrinx, or dermal sinus tract was seen.The patient underwent D10-L1 laminectomy with subtotal excision. Intraoperatively, greasy keratinous debris with hair was encountered, consistent with dermoid cyst (Figures *3* and 4).Gross specimen showed keratinous material and hair (Figure *5*).The postoperative course was uneventful, with neurological status unchanged. A preoperative clinical photograph demonstrated equinovarus deformity of the right foot (Figure 7).Histopathology showing cyst wall lined by stratified squamous epithelium with adnexal structures.

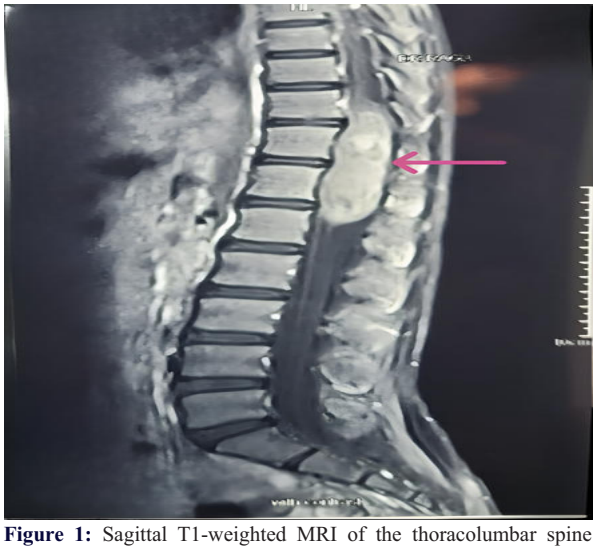

Figure 1: Sagittal T1-weighted MRI of the thoracolumbar spine showing a well-defined intradural extramedullary lesion extending from D10 to L1.

(Arrow point to the lesion appearing hyperintense on T1-WI, consistent with fat-containing material, and demonstrates minimal post-contrast enhancement.)

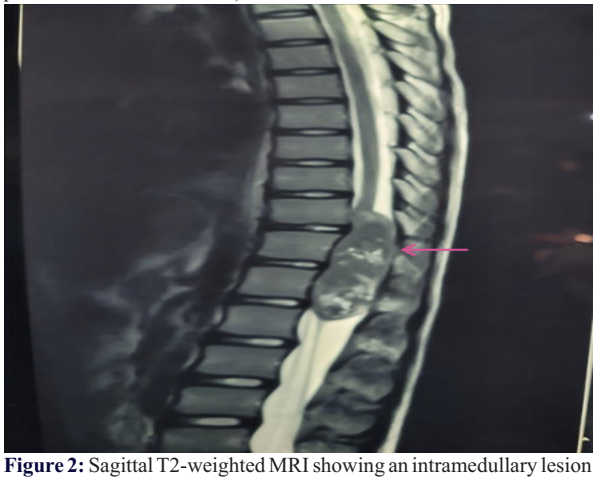

Figure 2: Sagittal T2-weighted MRI showing an intramedullary lesion

extending from D10–L1, with heterogeneous signal and cord expansion.

(Arrow indicates the lesion, consistent with a dermoid cyst.)

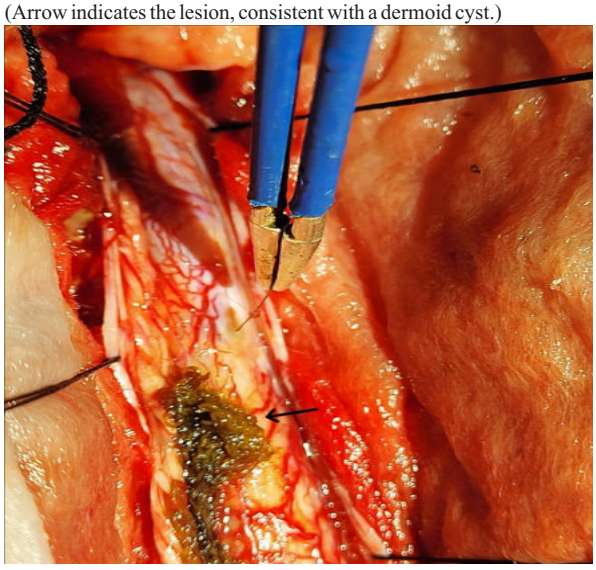

Figure 3: Intraoperative Image Showing Extrusion Of Keratinous Debris And Hair After Midline Myelotomy.

(Arrow points to the yellowish keratinous material and hair emerging from the cyst cavity, consistent with the contents of a spinal dermoid cyst.)

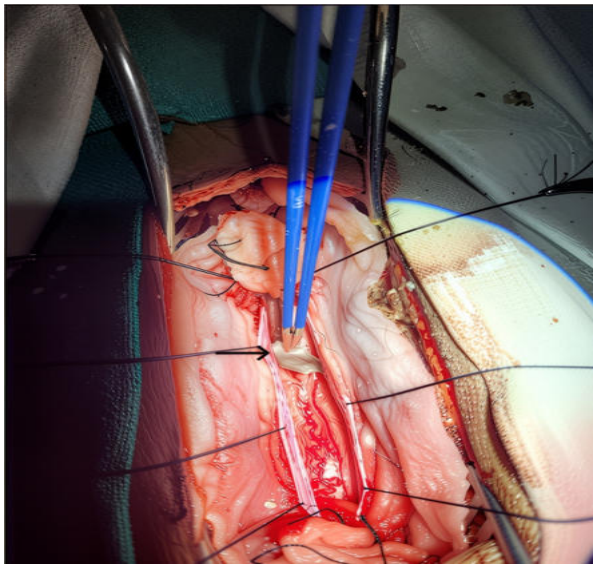

Figure 4: Operative Field After Subtotal Excision, Showing Cyst Cavity And Expanded Cord.

(Arrow points to the opened cyst cavity within the cord parenchyma following removal of dermoid contents.)

Figure 5: Gross Specimen Of Pultaceous Keratinous Material And Hair.

(Arrow indicates a fragment of the cyst wall containing adherent keratinous material and hair.)

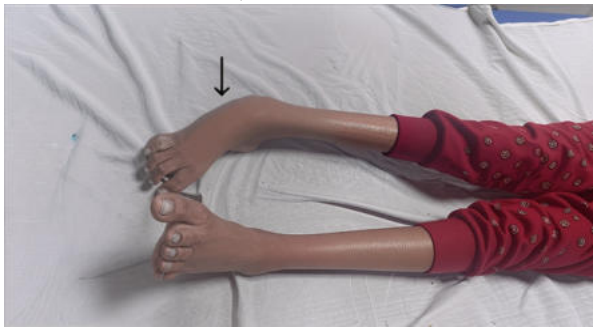

Figure 6: Preoperative photograph showing equinovarus deformity of the right foot.
(Arrow indicates the equinovarus deformity of the right foot.)

DISCUSSION

Intramedullary dermoid and epidermoid cysts are rare spinal lesions, accounting for less than 1% of tumors [1]. They arise either from ectodermal remnants trapped during neural tube closure (congenital) or secondary to trauma or surgical procedures (acquired) [2]. Cruveilhier first described spinal epidermoids in 1829 as “tumeurs perlées” [2]. While lumbosacral involvement is most common, intramedullary dermoids without dysraphism are exceptionally rare [3].

Several reports illustrate the spectrum of presentations.Patankar et al. described a cervical intramedullary dermoid in an 18-year-old female, managed with subtotal excision [3]. Miller and Chahlavi reported a 42 year old male with a lumbar dermoid at L1-L2, presenting with radicular symptoms and managed with gross total excision [4]. More recently, Anwer et al. highlighted a long-segment intramedullary dermoid extending from the cervical to dorsal spine, necessitating subtotal excision due to extensive involvement [5]. Lekgwara et al. reported a sub-axial cervical intramedullary dermoid in an adult, emphasizing the diagnostic and surgical challenges [6]. Gatam et al. described a rare adult intramedullary dermoid without trauma or dysraphism, reinforcing the sporadic nature of these lesions [7]. Similarly, Prachi et al. reported a dorsal intramedullary dermoid mimicking spinal tuberculosis [3]. In addition, cases of concomitant dermoid cysts at the conus and cauda equina have been described [8], and extramedullary dermoid cysts have also been reported in the literature [9]. Shah et al. further reported multiple intramedullary lipomas associated with a conal dermoid, highlighting radiological diversity [10] .Aggarwal et al. discussed two conus medullaris white epidermoids, stressing the risk of recurrence and the importance of surgical caution [2].

Our case stands out due to the pediatric age of presentation, absence of dysraphism, the presence of a long-standing equinovarus deformity, and atypical MRI findings (T2 hypointense, T1 isointense, poor enhancement). MRI characteristics of dermoids are variable depending on cyst contents, and thus histopathology remains the gold standard for diagnosis.Intraoperative rupture should be avoided to reduce the risk of chemical meningitis [2].

Taken together, our case reinforces that intramedullary dermoids can present atypically, MRI findings may be variable, histopathology is confirmatory, and safe subtotal excision provides decompression while minimizing morbidity.

CONCLUSIONS

Intramedullary spinal dermoid cysts are extremely rare lesions, particularly in the pediatric population and in the absence of associated spinal dysraphism. This case emphasizes the importance of maintaining a high index of suspicion when evaluating children with long-standing orthopedic deformities such as equinovarus foot, as these may be the initial manifestations of an underlying spinal pathology. MRI remains the imaging modality of choice, although findings can be variable depending on cyst composition. Intraoperative recognition of the cyst's characteristic keratinous contents confirms the diagnosis, and careful microsurgical excision provides effective decompression while minimizing neurological risk. Early diagnosis, individualized surgical planning, and long-term follow-up are key to

optimizing neurological and functional outcomes in such rare presentations.

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