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CASE REPORT

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Contiguous diastematomyelia with tethered cord, intradural extramedullary dermoid tumor, and lipomyelomeningocele: A unique case of spinal dysraphism

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Key Clinical Message

Diastematomyelia, tethered cord, intradural extramedullary dermoid tumor and lipomyelomeningocele such disease entities themselves are rare in their own form and concurrent presentation of all those pathological states in a single individual can be considered one of the rarest forms of spinal dysraphism globally. Moreover for prompt management with optimal prognosis needs refined neurosurgical intervention guided by intraoperative neuromonitoring so as to bring about the best quality of life in the patient.

KEYWORDS

diastematomyelia, lipomyelomeningocele, spinal dysraphism, tethered cord

1 | INTRODUCTION

Spinal dysraphism encompasses a diverse group of congenital abnormalities of the spine. Dermoid cysts are benign congenital anomalies arising from the entrapment of ectodermal elements along the lines of embryonic closure and are considered hamartomas. These tumors develop early in life in ectodermal and mesodermal tissues. Histologically, dermoid cysts consist of keratinous debris and hair follicles and are coated with a dermis, which contains skin appendages such as hair and sebaceous glands.[1] While commonly affecting the head and scalp in the pediatric population, intraspinal dermoid is extremely rare, accounting for around 1.1% of intraspinal tumors.[2] Diastematomyelia is a congenital anomaly of the spine due to defective formation of the primitive notochord, giving rise to two notochordal processes.[3] Here, we report a case of an intradural extramedullary dermoid cyst in a child with lipomyelomeningocele and the presence of a diastematomyelia with tethering of the cord, emphasizing the pathologic investigations and surgical treatment.

2 | CASE HISTORY/EXAMINATION

A five-year-old male presented with swelling over the lower back region with medial rotation of the right foot for the last 3 years. The patient also complained of difficulty walking. There was no history of trauma, tingling or burning sensation of limbs, weakness, or bowel and bladder involvement. There were no other medical, surgical, or localizing features. His developmental milestones were unremarkable. On physical examination, the patient is alert and oriented; the power of the upper limbs was 5/5, the left lower limb was 5/5, and the right lower

limb was 4/5. Sensations were diminished over the right lower limb compared to the left lower limb, reflexes were normal, and the planter reflex was bilaterally intact. The rectal tone was normal, and no saddle anesthesia was noted. Examination of the back revealed a large soft, non-tender, non-pulsatile swelling in the lumbosacral region measuring approximately 4×4 cm without a tuft of hair (Figure 1).

3 | METHODS (DIFFERENTIAL DIAGNOSIS, INVESTIGATIONS AND TREATMENT)

MRI of the spine showed an intradural nodular lesion (Figure 2) in the posterior aspect of the spinal canal at l2 level with cord tethering, features of diastematomyelia (Figure 3) below d12 level, and dorsal dermal sinus at l4-5 level with spinal cord tethering. The CT scan (Figure 4) of the patient showed defect in posterior arch of l2, l3, l4, l5, and all sacral sol in spinal canal, vertebrae, partial fusion noted in the right l4/5 facet joint, and a thin linear fibrotic like tract noted between spinal canal and skin at l4/5 level likely dermal sinus. Screening of the craniospinal axis was done to rule out hydrocephalus and Arnold-Chiari malformation. Management Based on the clinical and radiologic findings, the patient was scheduled for surgery. Surgery was carried out through a midline skin incision in the dorsolumbar region. On exposure, a type I split cord malformation (SCM) was confirmed, and an extradural bony spur was located at the D12-L2 vertebral level. Each hemicord had its own dural sac and associated independent lipomyelomeningocele. Excision of the bony spur was done microscopically using a high-speed drill extradurally. The dura mater was opened in an inverted, Y-shaped fashion. Debulking of the lipomatous tissue and untethering of the cord was done to the white line (interface between the lipomatous and the cord tissue) by using meticulous micro neurosurgical dissection. The patient underwent untethering of the cord with excision of the intradural extramedullary dermoid mass with excision of lipoma (Figures 5 and 6). The intraoperative simulation produced no significant responses to the perianal sphincter and the lower limb muscles. The patient did undergo intraoperative somatosensory and motor evoked potential neuro monitoring, which did not reveal any reduction in the evoked potentials intermittently. The pathology of the mass showed a dermoid cystic type of lesion.

4 | CONCLUSION AND RESULTS (OUTCOME AND FOLLOW-UP)

Post-operatively, the patient's sensation over the right lower limb has improved, and there is also mild improvement in mobilization. The patient did not develop any new neurologic defects or cerebrospinal fluid leak after surgery. She was discharged on postoperative day 10. At the end of follow-up at 18 months, the patient showed mild improvement in bladder and bowel sensations; however, there was no improvement in her limb weakness. Spinal dysraphism's are themselves unique and rare forms of neurosurgical groups of disease conditions. The concurrent association of several types of spinal dysraphism in a single individual has been reported very scarcely in medical literature. Moreover, the management of such conditions is considered very arduous as well as they require prompt neuro monitoring guided surgical management, which is not that readily available with ease in developing lower-middle-income countries. But with meticulous surgery, the desired results and prognosis can be achieved in most of the cases.

5 | DISCUSSION

Spinal dysraphism's are congenital abnormalities of the spine and spinal cord arising from aberrations in the processes of gastrulation, primary neurulation, and secondary neurulation. A study conducted by Chellathurai et. al identified posterior myelomeningocele as the most common type of spinal dysraphism encompassing

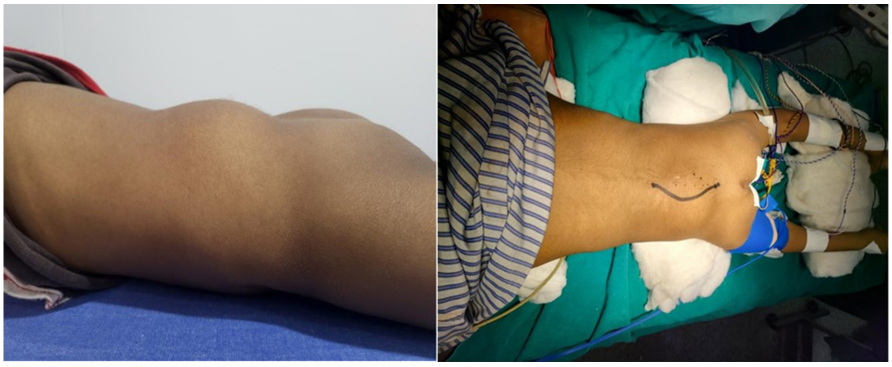

FIGURE 1 Swelling in the lumbosacral region suggestive of lipomyelomeningocele.

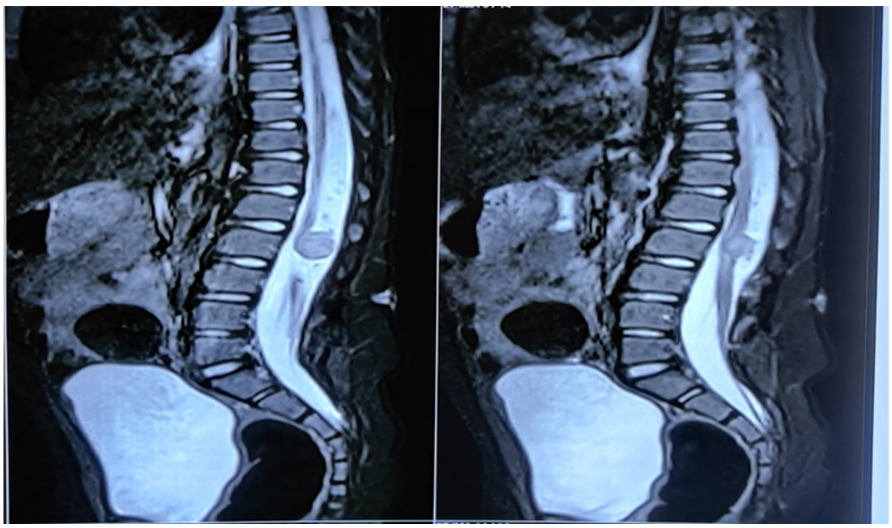

FIGURE 2 T2 MRI of the spine showing approximately 1.6 × 1.4 × 1.1 cm sized well-circumscribed nodular lesion is seen in the intradural location at L2 vertebral level posterior to the spinal cord suggestive of intradural extramedullary dermoid tumor with the spinal cord tethered in the anterior aspect of the lesion.

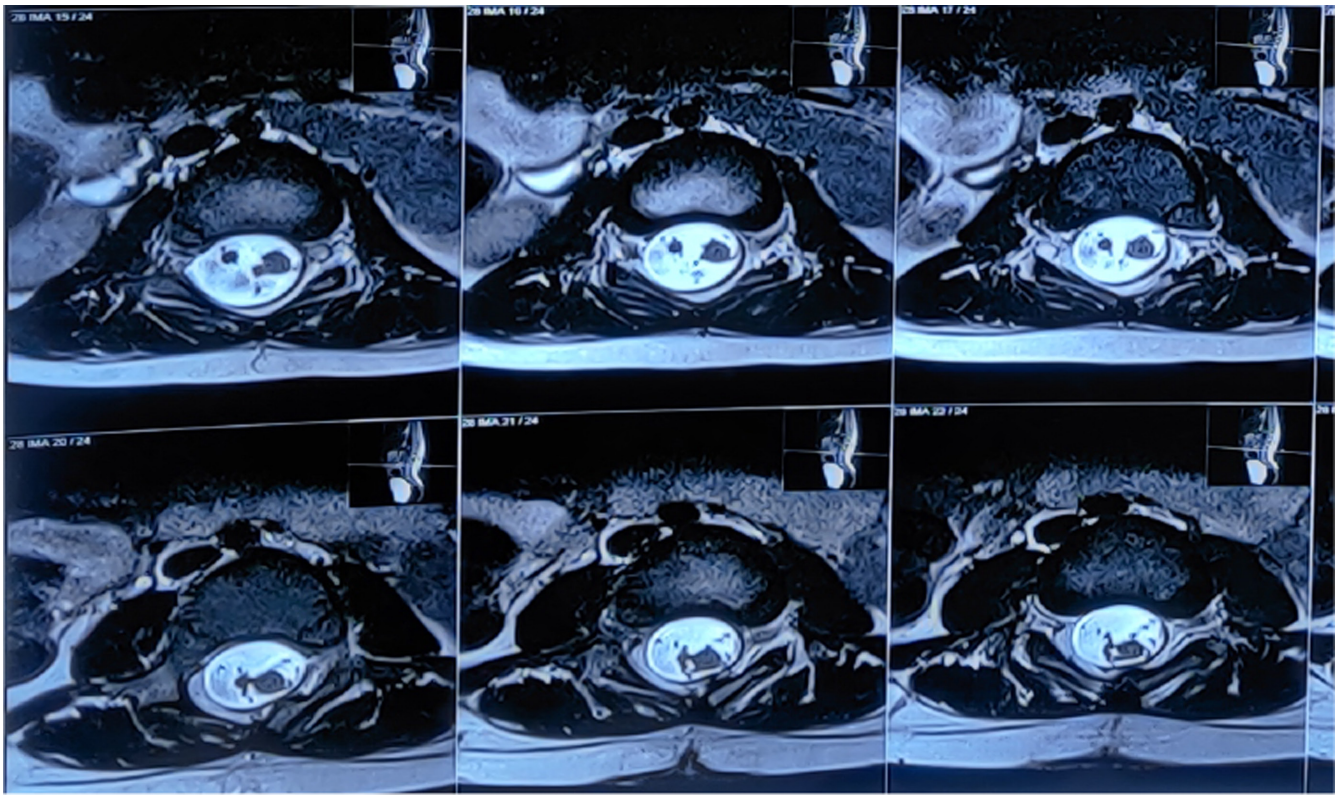

FIGURE 3 MRI spine axial view showing features of diastematomyelia characterized by a longitudinal splitting of the spinal cord into two hemicords below D12 level.

around 14.2% of the total cases, whereas posterior lipomyelomeningocele was found in 3.33% of cases, only 1.3% of spinal dysraphism was due to dermoid and Lumbar diastematomyelia was found in 6% of the cases.[3] To the best of our knowledge, there has not been a simultaneous concurrence of lumbar diastematomyelia, lipomyelomeningocele, and an intradural dermoid to date.

While Dermoid cysts are benign cystic lesions arising during the gastrulation stage of development with primary disruption of tissues derived from the surface ectoderm, lipomyelomeningocele forms because of premature dysfunction where the epithelial ectoderm detaches prematurely from the neural ectoderm, which allows the mesenchymal tissue to contact the inner surface of the developing neural tube.[4] As the lipoma tethers the cord to the adjacent soft tissue, the tethered cord can naturally coexist with lipomyelomeningocele in most cases. Lipomyelomeningocele can be associated with several abnormalities, with Hoffman et al.[5] reporting an

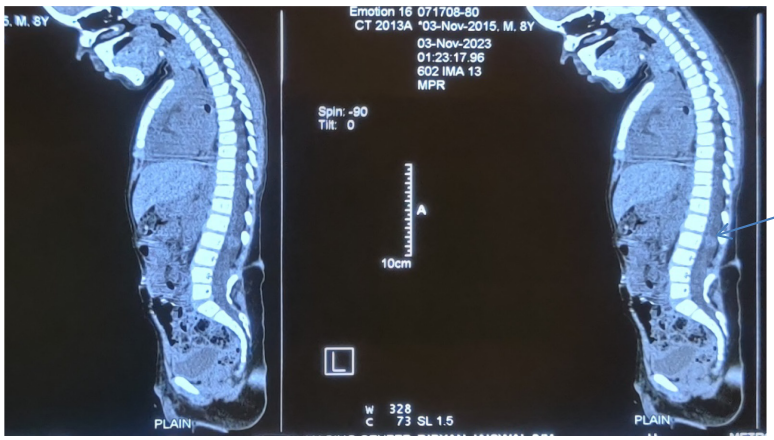

FIGURE 4 The CT scan of the patient showing a defect in the posterior arch of l2, l3, l4, l5, and sacral space occupying lesion in the spinal canal.

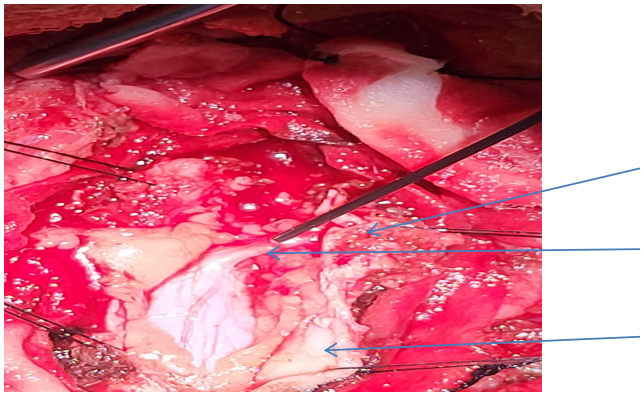

FIGURE 5 Intraoperative picture depicting a tethered cord attached to lipomatous lesion along with intraoperative neuromonitoring using neuromonitoring rod.

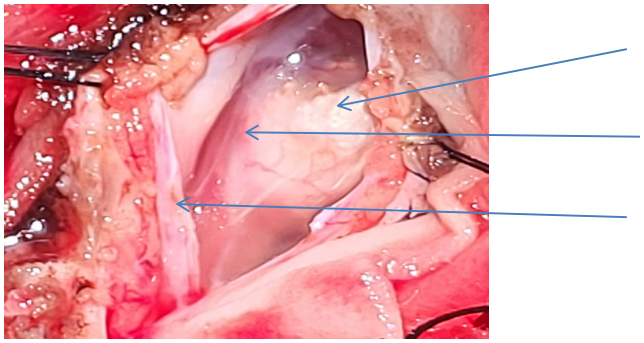

FIGURE 6 Intraoperative picture depicting the opening view of the duramater along with the arachnoid and underlying dermoid mass.

association with genitourinary tract abnormalities (4.1%) and dermoid cysts (3.1%), along with other associations. Diastematomyelia arises due to a defect in midline integration of the primitive notochord, such that there are two notochordal processes, each inducing the formation of separate neural plates.[3] Although diastematomyelia is commonly found to be associated with vertebral abnormalities, our patient had no such findings except for a swelling in the back along with a hairy tuft.

Because of multiple concurrences of spinal cord pathologies, it is usually expected to find several neurologic disturbances such as back pain, foot deformities, muscular atrophy, progressive weakness, extremity shortening, bowel bladder incontinence, paresthesia, paresis, reflex abnormalities, and gait disturbances.[6,7] Our patient complained of difficulty in walking along with a medial rotation of the right foot, and the right lower limb power was 4/5. However, there were no sensory or autonomic dysfunctions. The case series by Bradford revealed that most cases with spinal dermoid present with bladder symptoms, either retention or incontinence.[8] Our patient, despite having diastematomyelia and an extramedullary dermoid had a normal bowel and bladder functioning which was rather surprising.

Lipomyelomeningocele may be suspected clinically based on the presentation of a swelling in the back region and neurological symptoms, but spinal dermoid and diastematomyelia have non-specific clinical pictures. Plain X-rays may also show normal or nonspecific findings. MRI is a better imaging modality than CT scan because it helps detect the morphological patterns, signal intensities, and enhancement patterns of a tumor. Dermoid cysts on MRI present as an intraspinal tumor, and the differential diagnosis can be extensive (dermoid, epidermoid, meningioma, neurofibroma, glial neoplasm, hemangioblastoma, intramedullary metastasis, lipoma, infectious abscess, etc).[9] MRI of the spine of our patient showed an intradural nodular lesion in the posterior aspect of the spinal canal at l2 level with cord tethering, features of diastematomyelia below d12 level, and dorsal dermal sinus at l4-5 level. Because the differential diagnosis is too extensive, the only way to diagnose these lesions is via surgical excision and histopathological examination. STIR MRI can distinguish a dermoid from lipoma based on the non-fat component of the former pathology.[8] Histopathological examination is the most accurate way to diagnose spinal tumors. Dermoid cysts are coated with dermis and thus contain skin appendages such as hair and sebaceous glands.

Surgical excision is the treatment of choice in the case of spinal dermoids. Although extramedullary dermoid cyst can be totally resected, complications can arise in the case of intramedullary dermoids because of the attachment of the capsule to the spinal cord and proper judgment is imperative regarding its complete resection.[10] Our patient underwent untethering of the cord with excision of the intradural extramedullary dermoid mass with excision of lipoma. Vissarionov et al recommend that treatment of diastematomyelia should be based on the degree of deformity and the rate of progression. The presence of bony spur indicates a necessity for surgery because of the associated progressive neurological deterioration in these patients. The bony spur should be removed prior to surgical correction of the spine.[11] There is some doubt as to whether untethering should be undertaken in such a patient or not. The patient's neurologic deteriorations only included right lower limb weakness and slight diminution of sensory functions, and bowel bladder functions were normal. Accordingly, whether an intra-spinal procedure including untethering would turn out to have more benefit or more harm should be carefully weighed before surgery. Our patient did undergo surgical untethering and postoperative sensory and motor functions had remarkable improvements and there were no complications.

AUTHOR CONTRIBUTIONS

Sagun Ghimire: Conceptualization; data curation; resources. **Ananta Maharjan:** Conceptualization; supervision. **Shikher Shrestha:** Conceptualization; resources; supervision. **Dinuj Shrestha:** Supervision; visualization. **Kajan Ranabhat:** Conceptualization; supervision. **Suman Bhattarai:** Conceptualization; supervision. **Prabin Chaudhary:** Resources; software. **Kritick Bhandari:** Writing – original draft; writing – review and editing.

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CONFLICT OF INTEREST STATEMENT

The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

DATA AVAILABILITY STATEMENT

Any data used in the manuscript can be made available if asked upon by the chief editor.

CONSENT

Written informed consent was obtained from the patient to publish this report in accordance with the journal's patient consent policy.

CONSENT TO PUBLISH

The participant has consented to the submission of the case report to the journal.

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