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A rare case of intradural extramedullary epidermoid with focal diastematomyelia

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ABSTRACT

Diastematomyelia is a rare congenital deformity, more common in females usually associated with cutaneous manifestation of spinal dysraphism with congenital scoliosis. We report a female patient 26 years old with scoliosis in the dorsolumbar spine with dorsolumbar focal diastematomyelia type-2 with intradural extramedullary epidermoid which is a rare entity.

Key words: Diastematomyelia; Spinal dysraphism; Scoliosis; Intradural extramedullary epidermoid

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INTRODUCTION

Diastematomyelia is a variety of spinal dysraphism in which there is a congenital splitting of some part of the spinal cord or rarely of more than one part. The name is derived from a Greek word meaning "separation" and "marrow" (i.e., spinal cord).[1] Diastematomyelia with septum is more common in females than in males about 3.5–13. Cutaneous manifestation of spinal dysraphism comprising in descending order of frequency, localized hypertrichosis (40%), subcutaneous lipoma, nevoid skin, atretic meningocele, and dermal sinus.[2] Diastematomyelia with septum with or without neurological deficit may be associated with congenital scoliosis. Vertebral anomalies associated with diastematomyelia with septum are central bony spur, spina bifida, vertical laminar fusion, block vertebrae, split vertebral body, hemi vertebral hypoplasia, fused or deformed spinous process, scoliosis, kyphosis, or lordosis.[3]

Intradural extramedullary tumors are uncommon with an incidence of 3–10/100,000 population.[4] These tumors occur predominantly in the middle decades and the sex distribution is about equal except high incidence of meningioma in females. The most common intradural extramedullary tumors are the nerve sheath tumor, which constitutes 30% of spinal tumors, and the meningioma which constitutes approximately 25% of the tumors, the most common intramedullary tumors having about equal incidence, are the astrocytoma and ependymoma. A variety of other intramedullary tumors, including hemangioblastoma, dermoid, epidermoid, and mixed tumor, are common. Ninety percent of all intradural spinal cord tumors are benign and potentially resectable.[5] Therefore outlook after surgical

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therapy is excellent. Similarly, severe defects due to spinal cord compression in the restively young individual can be reversed by removing these tumors, with the expectation of improvement of neurological function.[6]

CASE PRESENTATION

We report a female patient 26 years old presented to the neurosurgery outpatient department with a complaint of low back aches for 1 month and weakness in both lower limbs for 20 days with bowel and bladder incontinence for 15 days and decreased sensation in both lower limbs below groin region for 15 days. She does not have any known comorbidity and was on painkillers for low backache. On presentation, her pulse was 78/min, blood pressure 100/70 mmHg, respiratory rate of 16/min, and oxygen saturation of 99% on room air. On systemic examination, bilateral lung field air entry is equal, per abdomen soft on palpation. On neurological examination, the patient's Glasgow Coma Scale was conscious, confused, and obeying commands. The pupil was bilateral normal size, and normal reactive. On motor examination power in the lower limb is 2/5 at all joints in the right lower limb and 3/5 at all joints in the left lower limb, deep tendon reflex (DTR) +3 in both lower limbs with bilateral plantar extensor with bowel and bladder involvement. The sensory level of involvement is below the L2 level. Magnetic resonance imaging of Dorso's lumbar spine with contrast report suggests – a small well defined intradural extramedullary non-enhancing cystic lesion T2 hyperintense T1 hypointense with a T2 isointense area in the posterior dependent part of the lesion causing compression of the spinal cord and anterior epidural space anteriorly not extending to neural foramina seen at D11-12 level possibility of epidermoid (Figure 1). Partial block vertebrae at D3-4 and D12-L1 level with partially fused and bifid vertebrae. Low-lying cord with conus medullaris terminating at level of L4-5 level (Figure 2).

There is a longitudinal focal split in the lower dorsal cord from the D12-L1 intervertebral disk to the L3 vertebral level with cerebrospinal fluid cleft between the hemi cords. No obvious fibrous/osseous septa or syrinx was noted in this region suggestive of focal diastematomyelia type 2.

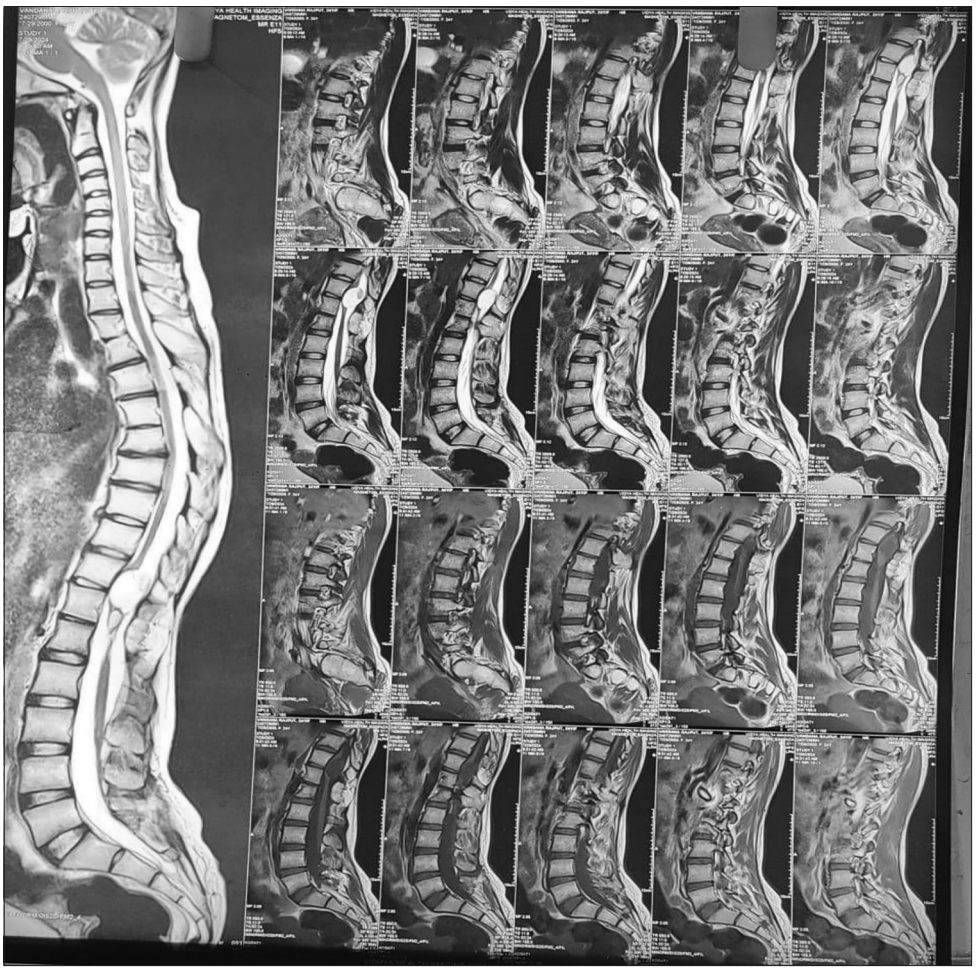

Figure 1: Magnetic resonance imaging of Dorso's lumbar spine with contrast sagittal view

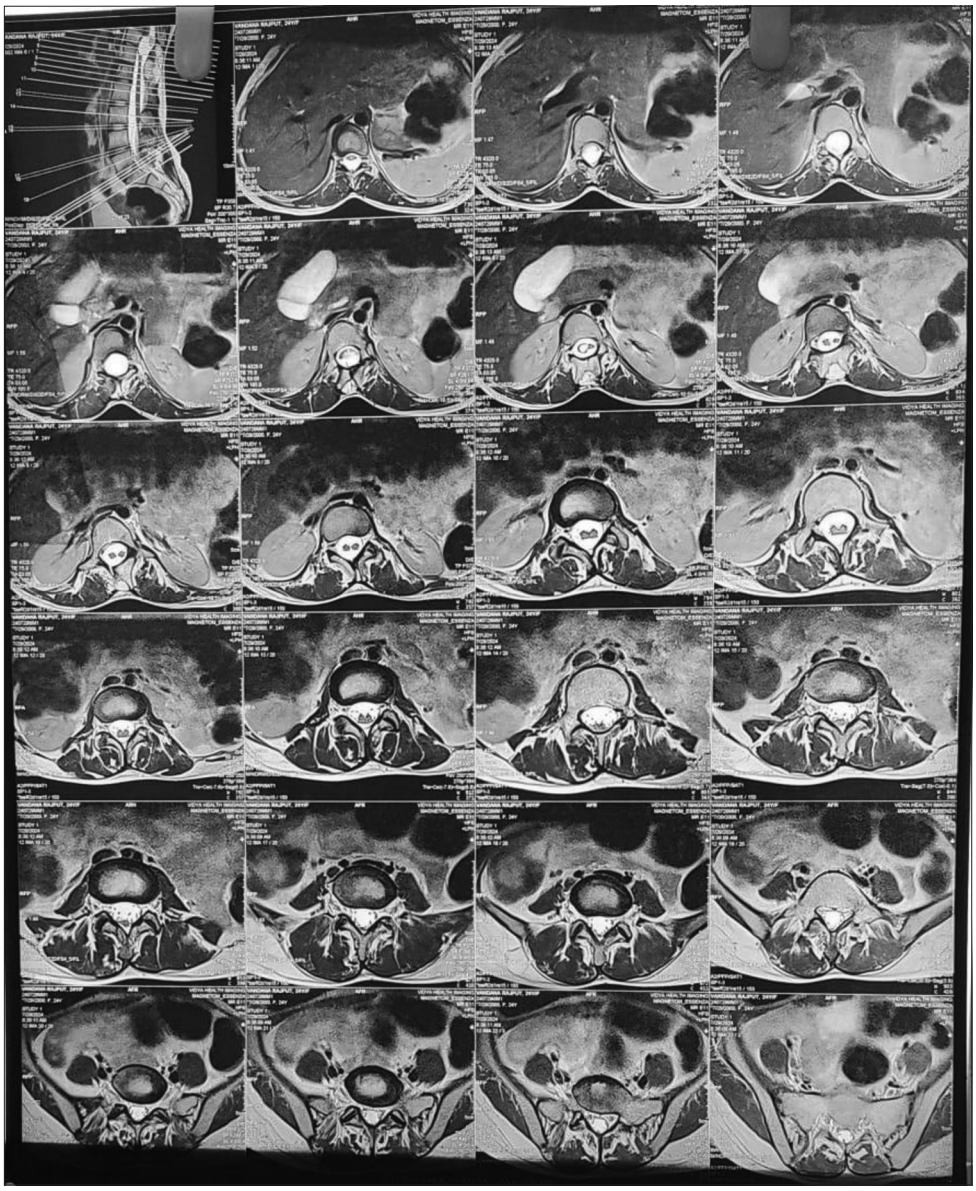

Figure 2: Magnetic resonance imaging of Dorso's lumbar spine with contrast axial view

The patient prepared for the surgery with proper consent and underwent D11-D12 laminectomy with excision of cystic structure with detethering of the cord by excising the filum terminale on October 08, 2024. Under general anesthesia in a prone position with intraoperative milky white fluid coming out of the cyst which was sent for histopathological analysis as shown in (Figure 3).

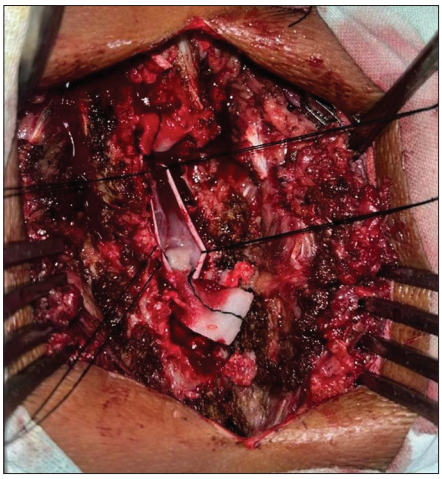

Figure 3: Intraoperative picture of cystic sol with milky white fluid coming through the middle of it

DISCUSSION

The immediate results and future prognosis in intradural extramedullary tumor, have been well established. These tumors are benign and if they are carefully and thoroughly removed, the patient should be cured, with an excellent prognosis[7]. Even in patients who

have been devastated neurologically by the growth of these tumors before surgical intervention, there is some hope, especially in young individuals that many of the neurological abnormalities may resolve slowly in the post-operative period. It may take 18 months to 2 years to maximize the resolution of these neurological deficits.[8]

Neurological improvement following an operation for diastematomyelia is usually modest and is restricted to symptoms and signs of recent onset. Children with dysraphism are never improved beyond their degree of congenital neurological deficit. However, pain in the back and legs and muscle spasms are always relieved and may be completely abolished. Urinary incontinence if of recent origin, also tends to improve. Foot deformity requires close, continued orthopedic supervision.[9]

CONCLUSION

Diastematomyelia usually presented with spinal dysraphism, this case report highlights the rarest presentation of diastematomyelia with tethered low-lying conus medullaris with the associated intradural extramedullary space-occupying lesion. Further research and studies are warranted to know the post-operative outcome of both diseases together.

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Authors Contribution:
HDP- Definition of intellectual content, literature survey, prepared the first draft of the manuscript, implementation of the study protocol, data collection, data analysis, and manuscript preparation and submission of the article; **AS-** Concept, design, clinical protocol, manuscript preparation, editing, manuscript revision, design of the study, statistical analysis and interpretation, and review manuscript; **VK-** Review manuscript; **SS-** Literature survey and preparation of figures; **AS-** Coordination and manuscript revision.

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