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**Case Report**

# Epidermoid Causing Split Cord Malformation in a Case of Jarcho Levin Syndrome (JLS)

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## Abstract

Jarcho-Levin syndrome (JLS) (costovertebral syndrome) is a rare congenital anomaly with multilevel vertebrocostal deformities. Intraspinal epidermoid cyst is a rare tumor. The incidence in adults is lesser than 1% and in children lesser than 3%. Epidermoid is predominantly seen at the dorsal spinal level. These may be congenital or acquired with known association with spinal dysraphism. We hereby report a rare case of split cord malformation with epidermoid in a 7-year female with JLS.

The patient presented with complaints of progressive scoliosis, since3 years of age. On Neurological examination revealed no motor deficit. Patient had a dorsolumbar scoliosis with a hairy patch. X-ray revealed irregular fusion of left sided ribs and scoliosis of dorsolumbar spine with convexity towards left. Magnetic resonance imaging (MRI) and computerized tomography (CT) of the spine were done which revealed bifid spine from D11-L1, diplomyelia with two hemicords in a single dural sac with no membranous or bony septum.

Intra operatively incompletely fused spinous process were seen from D11-L1. The patient underwent D10 to L2 laminectomy; there was a fibrous band which was leading into the bifid spine which was followed till the dura after removal of the laminae. After opening the dura this fibrous band was seen extending into the split cord. To our surprise split cord was continuing above the fibrous band following it revealed an epidermoid at the junction of the split cord, which was excised completely and fibrous band also removed. Histopathology of lesion confirmed as epidermoid.

**Keywords:** Epidermoid; Jarcho levin syndrome (JLS); Congenital scoliosis; Diplomyelia; Spondylo costal dysostosis (SCD); Spondylothoracic dysostosis (STD).

## Introduction

Jarcho-Levin syndrome (JLS) is a rare congenital anomaly of the thoracic cage and vertebrae with an estimated incidence of 1 per 40,000 births. JLS is a clinico-radiological entity characterized by vertebral and rib malformations. Only few cases of JLS with diplomyelia have been reported in the literature [1,2]. Spinal epidermoids are uncommon [3].

They comprise between 0.5% and 1% of all spinal tumors [4]. Although present since birth, congenital epidermoid tumors are often not discovered until the second to fourth decade of life since they are mostly asymptomatic. Few cases of dermoids and epidermoids with diplomyelia have been reported [5,6]. Diplomyelias are most frequently found in the lumbar spine, followed by the thoracic region [7].

The authors describe a rare case of the JLS along with congenital intradural extramedullary epidermoid with diplomyelia which has not been reported yet in the available literature.

## Case report

A 7year-old Female child born of full-term normal delivery after a non-consanguineous marriage was referred to us with complaints of progressive scoliosis, since3 years of age. On examination the child had short neck, grove in the left intercostal region, a hairy patch near to the dorso lumbar region, with no motor deficit and Gait was normal. X-ray revealed irregular fusion of left sided ribs and scoliosis of dosrolumbar spine with convexity towards left [Figure 1 (A-B)].

The magnetic resonance imaging (MRI) and CT imaging showed diplomyelia extending from D11- L1 with split cord at D11 with single dural sac with no bony or membranous septum. No CVJ anaomolies were seen [Figure 2A-B].

The patient underwent D10 to L2 laminectomy. Intra operatively bifid spine was seen from D11-L1. There was a fibrous band which was extending from dermal lesion into the bony elements. It was dissected and traced into the dura.

On opening the dura, the two hemicords were enveloped in a single dural sac (Pang Type 2) and the fibrous band was ending between the split cord. To our surprise the band was not causing the splitting of the cord. Split cord was extending proximal to the entry of the band. The split cord was followed up proximally to an epidermoid at the level of the split.

The epidermoid (15mmx15mm) was on the surface at the split. The epidermoid and fibrous band was excised completely [Figure 3 A-E] and sent for histopathology analysis which confirmed it to be an epidermoid [Figure 4]. The patient had no neurological deficit postoperatively, on day 14 the sutures were removed. Follow-up review was done after 6 months.


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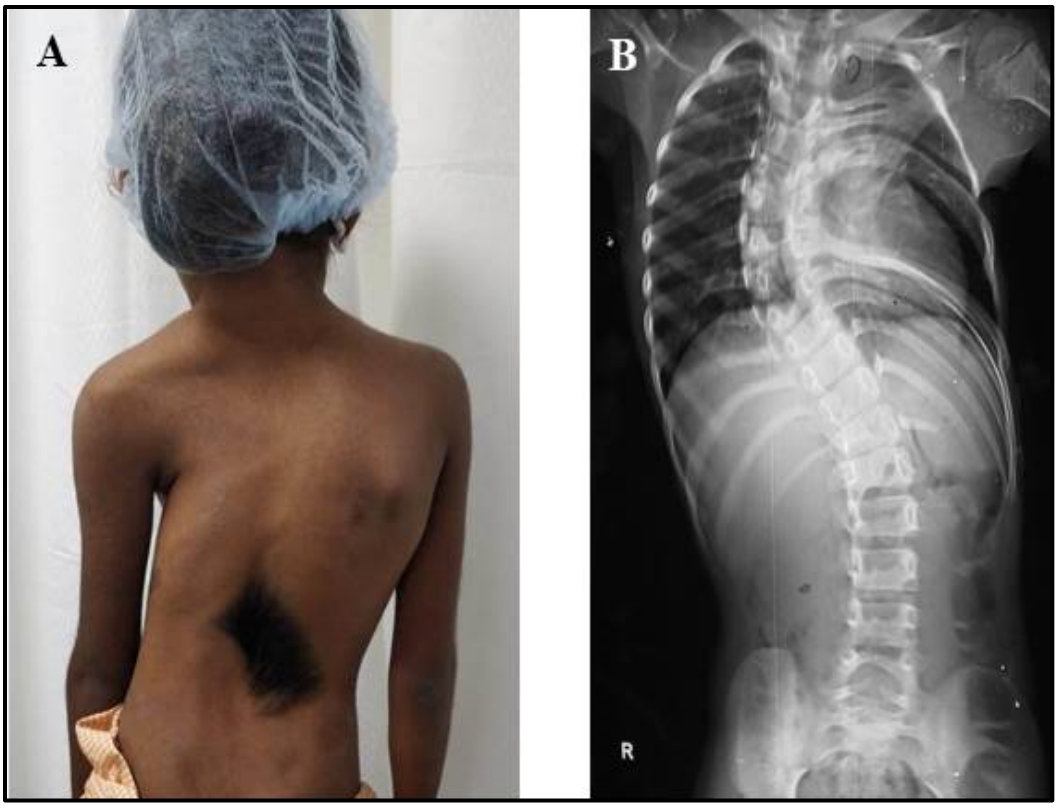


**Figure 1**: A) Hairy patch at dorsolumbar region, B) Fused left ribs with dorsolumbar scoliosis.

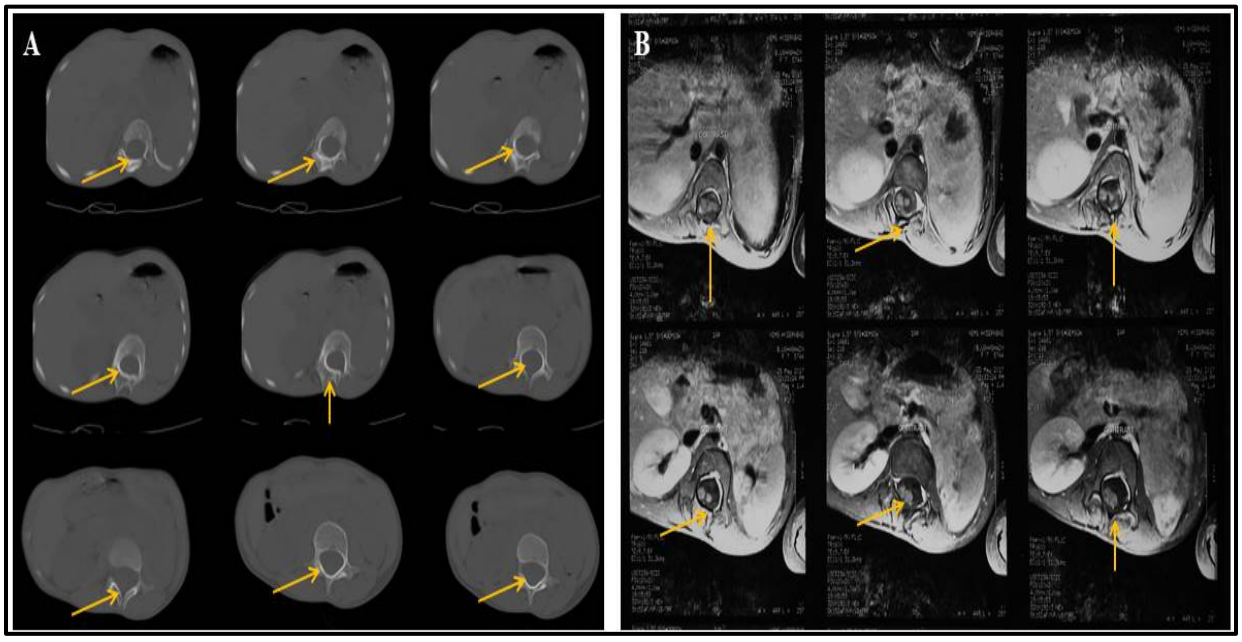


**Figure 2**: A) Axial CT showing bifid spine with no bony septum, B) Axial MRI showing diplomyelia with no epidermoid.


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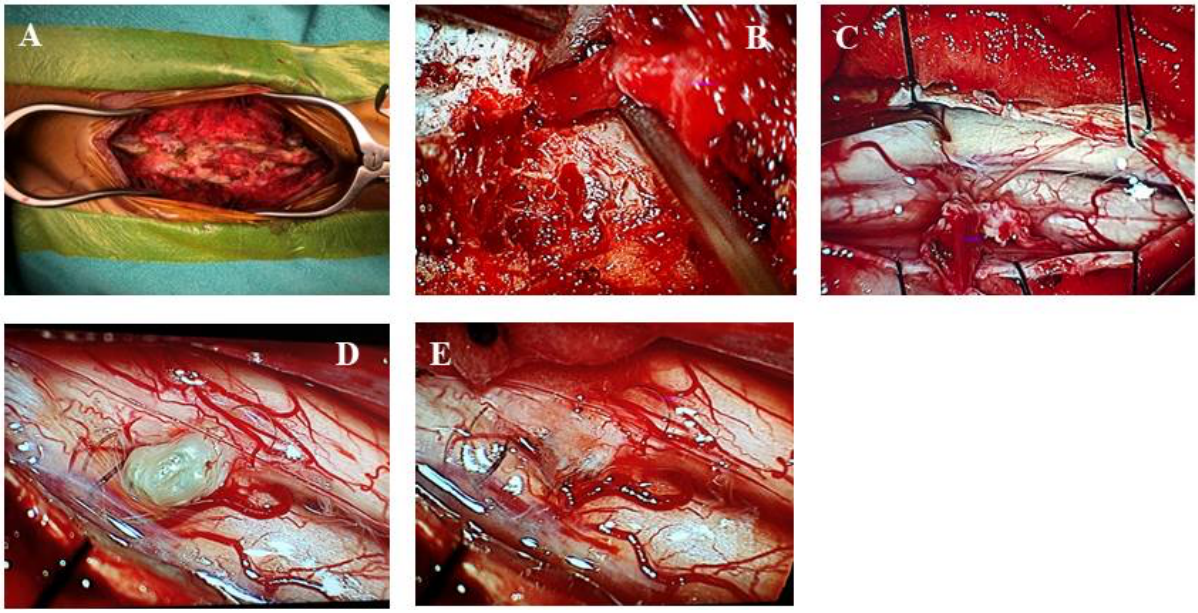


**Figure 3**: A) Bifid spine from D11-L1, B) Fibrous band entering the bifid spine, C) fibrous band extending into the split cord after opening the dura, D) epidermoid seen at the junction of diplomyelia, E) epidermoid completely removed.

## Discussion

JLS or Costovertebral dysplasia (CVD) is a rare congenital anomaly of the thoracic cage and vertebrae with incidence of 1 per 40 000 births [8]. Till date over 400 cases were reported in worldwide literature and over 18 cases were in reported in Indian literature [9]. The mode of inheritance is both autosomal dominant and recessive. Jarcho and Levin were the first to describe it in 1938 as a type of segmental costovertebral malformation [10]. Later classified cases of JLS into two phenotypes depending on the type of rib malformation as spondylo costal dysostosis (SCD) and spondylothoracic dysostosis (STD) [10,11]. Spondylo costal dysostosis (SCD) is associated with vertebral anomalies, intrinsic rib malformation (bifurcation, abnormal orientation, and fusion), flaring of iliac bones. Spondylothoracic dysostosis (STD) is associated with vertebral anomalies, crab or fan like rib configuration but no intrinsic rib malformations [12]. Mutations in any of the four genes that are important components of the notch signaling pathway (DLL3, MESP2, LFNG and HES7) are responsible for the development of spondylocostal dysostosis (costovertebral malformation) in JLS [13,1].

Diplomyelia is stated as a phenomenon where two hemicords share a single dura but separated by a bony or fibrous septum. During embryogenesis, the attachment between ectoderm and endoderm leads to formation of an endomesenchymal tract that splits the notochord, forming two hemineural plates and hemineural tubes.

Dyplomyelia can be associated with scoliosis, which might be vertebral anamolies or due to syrinx [14]. It is imperative that the patient should undergo all imaging investigations (i.e


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Computed tomography (CT), Magnetic resonance imaging (MRI)) for proper and precise diagnosis of vertebral anomalies, for avoiding neurological complications when opted for the correction of the spinal axial deviation formed due to the presence of diastematomyelic septum.

The congenital epidermoid cysts can be formed due to the aberrant ectoderm inclusion at the time of neural tube closure during embryonic life (i.e. 3rd to 5th week of gestation), which lead to mid line location of most cysts and are associated with spinal dysraphisms, such as diastematomyelia, hemivertebra, dermal sinus tract, meningomyelocele, etc [11,3].

Histologically, epidermoid consist of stratified squamous epithelium supported by an outer layer of collagenous tissue. Progressive desquamation and breakdown of keratin from the epithelial lining into the interior of the cyst produce the characteristic content.

The present case describes the rare case of a 7year-old Female presented with complaints of progressive scoliosis, since3 years of age with no neurological deficits. Intra operatively spina bifida with incompletely fused spinous process were seen from D11-L1.

Due to axial rotation laminectomy was done carefully to prevent injury to thecal sac. There was a fatty tissue with fibrous band which was leading into bifid spine which was followed till the dura. Hemicords though were equal they appeared to be unequal due to axial rotation. The fibrous band was followed till the split after opening the dura and was getting fused into the ventral dura. Band and blood vessels were excised. No bony septum was seen. To our surprise split cord continued above the fibrous band. An epidermoid of size 15X15 mm at the split cord was noticed and excised completely. Histopathology of lesion confirmed it as epidermoid.

JLS with diplomyelia associated with epidermoid has not been reported in the existing literature and epidermoid could be the inciting factor for the split cord rather than the fibrous band.

Children presenting with vertebral and rib anamolies-syndromic association like JLS should be kept in mind and also should evaluate for diplomyelia, fibrous band, bony septum, syrinx and lesions like dermoid, epidermoid etc [15].

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**Conflict of Interest**

None

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None

**Patients consent**

Consent was acquired from the patient's parents for publication.


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