

## Neurenteric cyst with herniation of spinal cord into the posterior mediastinum

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### ABSTRACT

Neurenteric cyst is a developmental abnormality. It accounts for 0.7-1.3% of spinal axis tumors. This rare lesion results from the inappropriate partitioning of the embryonic notochordal plate and presumptive endoderm during the third week of human development. It is usually associated with other abnormalities like vertebral defects and dorsal enteric cyst. It is one of the components of the split notochord syndrome.

**Key words:** neurenteric cyst, split notochord syndrome, spinal tumor

### INTRODUCTION

Neurenteric cyst is a rare benign endodermal lesion of the central nervous system, also referred to as enterogenous cysts.<sup>1</sup> It is one of the components of split notochord syndrome. This syndrome is a spectrum of congenital malformations that results from the "splitting" of notochord due to a persistent connection between endoderm and dorsal ectoderm. Other components of the split notochord syndrome are dorsal enteric fistula, dorsal sinus, and dorsal diverticula.<sup>2</sup> Here we report the CT and MRI features of a mediastinal neurenteric cyst.

### CASE REPORT

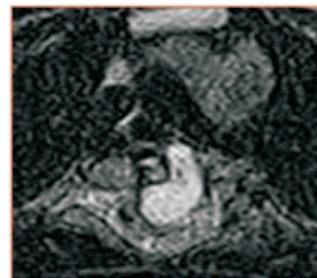
A four year old boy presented with backache. He was having deformities of spine and chest as well as developmental delay. There was history of a single episode of seizure at the age of one year. Neurological examination did not reveal any significant abnormality. He was subjected to CT and MRI investigations. CT scan of cervico-dorsal spine showed multiple segmentation anomalies of the upper thoracic vertebra with widening of interpedicular distance and posterior vertebral defects with dural sac extending up to the subcutaneous plane- suggestive of myelomeningocele. There was a well marginated intraspinal solid and cystic lesion with extension into the posterior mediastinum through a segmentation defect of dorsal vertebra – Neurenteric cyst (**Image 1**). There was diastematomyelia at D1 vertebral level with intra

medullary fat density suggestive of lipoma.



**Image 1:** Coronal CT reformed image shows: Splitting of the Spinal Cord

MRI of cervical spine showed multiple dysplastic upper dorsal vertebrae with associated ectasia and kyphotic deformity. A well marginated, extramedullary, intraspinal solid and cystic lesion with a small focal fatty component extending from C7 to D2 vertebral level was observed. The lesion was displacing the spinal cord posteriorly and extended ventrally into the posterior mediastinum through midline segmentation defect at D2 vertebra. The MRI features were consistent with mediastinal neurenteric cyst. (**Image 2, 3**)



**Image 2:** MRI T2 Axial image: anterolateral spina bifida



**Image 3:** MRI T1 Axial image ventral displacement of the dysplastic thickened cord into mediastinal cyst

## DISCUSSION

The embryologic mechanism of split notochord syndrome was first described by Saunders in 1943.<sup>3</sup> In the pathogenesis of split notochord syndrome, an adhesion occurs between endoderm and ectoderm along the route of notochord. The notochord splits around the adhesion creating a defect in the vertebral column. Adhesion between endoderm and ectoderm also results in potential connection between yolk sac and dorsal ectodermal surface, this connection persists as a tractus in the dorsal intestinal fistula which is the most severe form of split notochord syndrome. Sometimes this connection may obliterate at any point. This results in formation of dorsal enteric sinus, diverticula and neurenteric cyst.<sup>3,4,5,6</sup> Neurenteric cyst usually occurs at the cervico-thoracic junction or in the region of the conus medullaris. Its origin is similar to the posterior

mediastinal or abdominal cyst which often co-exists. The cyst may lie anterior or posterior to the spinal cord intradurally or it may lie in an intramedullary location. Commonest location of neurenteric cyst is posterior mediastinum. They are well delineated, thin walled fluid-filled masses containing elements derived from endodermal cells. The fluid within the cyst is nearly identical to CSF. They lie ventral to the spinal cord and associated with anomalies of vertebral segments. Neurenteric cysts are lined by gastrointestinal or respiratory epithelium. Anterior and posterior spina bifida, butterfly vertebrae and hemivertebrae frequently accompany the neurenteric cysts.<sup>1,3</sup> Prevertebral neurenteric cysts may be connected to the meninges or spinal cord by a tube or a fibrous neurenteric band via an anterior spina bifida and dura defect.<sup>7</sup> In conclusion we report MRI and CT features of a rare case of Neurenteric cyst with spinal cord herniation.

## AUTHOR NOTE

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