

# Neurenteric Cyst of Posterior Mediastinum in an Infant: A Case Report

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## Abstract

Neurenteric cysts are one of the rare congenital disorders that present within first 5 years. Neurenteric cysts are the enterogenous type of duplication cyst when associated with vertebral anomalies. These can be located anywhere in the body from intracranial to abdomen, but the posterior mediastinal neurenteric cysts are very rare, which presumably arise from misplaced epithelium of nasopharynx and intestinal tract. It is rare in incidence, with male predominance. Neurenteric cyst represents a failure of complete separation of the notochord from the foregut during embryogenesis. There are many types of the bronchopulmonary foregut malformations among which neurenteric cysts are the least common. The most common form consists of a simple epithelium resting on a delicate fibrovascular capsule. It is septate and has different varieties of epithelium (ciliated or non-ciliated, columnar, intestinal glands, pancreatic and salivary gland tissue) and a muscular wall with smooth mucosal lining. The treatment is complete resection of the cyst.

**Key words:** Infant, Neurenteric cyst, Posterior mediastinum

## INTRODUCTION

Neurenteric cysts are rare congenital anomalies, with only about 35 cases reported in the literature.<sup>1</sup> Neurenteric cysts develop during the 3<sup>rd</sup> week of development due to the abnormal connection between the primitive ectoderm and endoderm. These cysts are the result of the failure of complete separation of the notochord from the foregut. It is assumed that a cyst lined with enteric and neural tissue is formed when the foregut becomes incorporated into the notochord tissue.<sup>2,3</sup> Foregut duplication cysts are presently classified into three subtypes: (1) Enterogenous cysts (lined by intestinal epithelium), (2) Bronchogenic cysts (lined by respiratory epithelium), and (3) Neurenteric cysts (associated with apparent vertebral anomalies).

Neurenteric cyst can manifest in any age group (usually discovered during first 5 years of life) and can be found anywhere from intra-cranium to the abdomen, but many of the times they are located in the posterior mediastinum.<sup>2,3</sup> Neurenteric cysts are usually associated with vertebral anomalies and hemivertebrae may also be included.<sup>4</sup> Cervical and upper thoracic vertebrae are usually affected.

Neurenteric cyst can be diagnosed antenatally using ultrasound as early as 18 weeks gestation.<sup>2,3</sup>

## CASE REPORT

A 7-month-old female child with the history of a cough, fever, and left upper lobe pneumonic patch was referred to Krishna hospital for further management. The history revealed that a cough and fever were since last 2 months, for which she took treatment, symptoms subsided for few days and then recurred again. She also had dull pain in the front and back of the left chest. A clinical examination showed normal respiratory findings. Abdominal examination and spine were normal.

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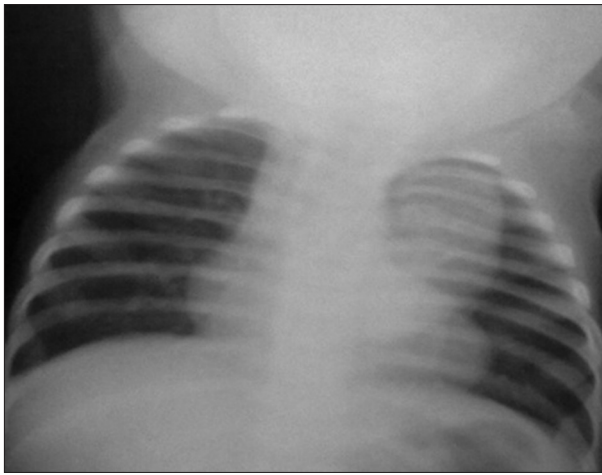
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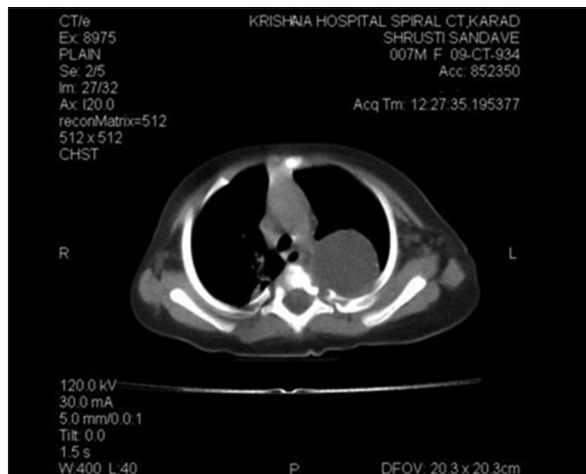
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**Figure 1: Posteroanterior roentgenogram of chest reveals a well-defined radio-opaque density with smooth sharp margins in the left hemithorax, paraspinal region with density less than that of heart as well as vertebral anomalies (hemivertebrae) with the adjacent vertebrae**



**Figure 3: Axial 5mm CT scan showing well defined cystic lesion with smooth sharp margins in the retroperitoneum on the left side anterior to left kidney and getting connected to L1 vertebral body**



**Figure 2: Axial 5 mm computed tomography scan showing well defined well-circumscribed hypodense (6-12 HU) cystic lesion in left hemithorax in posterior aspect in close proximity to vertebral body and posterior margin of rib, causing erosion of the rib**



**Figure 4: T2 weighted MRI showing well defined rounded hyperintense lesion with smooth, sharp and regular margins in the left paraspinal region extending from D2 – D7 vertebral bodies and getting attach to D3 – D5 vertebral bodies (which are hemivertebrae). Single septa present in the inferior aspect of the lesion**

Chest radiograph showed a left posterior mediastinal mass, which was extrapulmonary (Figure 1). Contrast-enhanced computed tomography (CECT) (Figures 2 and 3) and magnetic resonance imaging (MRI) (Figures 4 and 5) confirmed the presence of a cystic mass (5 cm × 3 cm × 3 cm in diameter) extending from D2-D7 vertebral bodies and getting attached to D3-D5 vertebral bodies that are hemivertebrae.

The CT and MRI also revealed another cystic mass (4 cm × 2 cm × 2 cm in diameter) in front and above the left kidney, showing a connection to L1 vertebrae. Other investigations were within normal limits. Pre-operative diagnosis of the neurenteric cyst was being made and by left posterolateral thoracotomy, the cyst was excised. Histopathology confirmed the diagnosis of the neurenteric cyst.

## DISCUSSION

Split notochord theory is the explanation for the neurenteric cysts.<sup>5</sup> Neurenteric cysts can appear in any age group, but it is the first 5 years of life in which they are mostly seen. The failure of separation of the notochord from the foregut during the 3<sup>rd</sup> week of development gives rise to these posterior enteric remnants. 1842 was the year in which Roth first reported vertebral column attachment to an enteric cyst, but Mc Ritchie, Purves, and Saunders coined the term “neurenteric.”<sup>6</sup> The first case reported in 1934 by Pusser.

Neurenteric cysts can be either multiloculated or septate and may look similar to gastric, duodenal or intestinal mucosa. These cells are mostly Periodic acid–Schiff positive and can contain mucus and globules, with occasional squamous cell

metaplasia. Neurenteric cyst lined with gastric columnar epithelium can develop hemorrhage, ulceration or erosion. Neurenteric cyst is also susceptible to infection, perforation or rupture.

In the pediatric population, one-third of the patient with mediastinal cysts remains asymptomatic while two-third present with complains of the respiratory system. These cysts are usually benign, but due to their size they can cause compression of the structures within its vicinity.

Neurenteric cysts are associated with cervical and upper thoracic vertebral abnormalities, such as hemivertebrae, as noted in our case, anterior and posterior spina bifida, the absence of vertebrae, scoliosis, and diastematomyelia. The most common symptoms were difficulty in breathing, stridor or a persistent cough.<sup>4,7</sup> Ganglion cells, lymphatic tissue, pancreatic tissue, salivary glands, or muscular tissue may be present in the cyst wall without serosa. The cartilaginous tissue is never present.<sup>8</sup>

A clinical trial of respiratory symptoms or distress, a chest radiograph demonstrating cervical or thoracic vertebral anomalies, and a posterior mediastinal cyst suggest neurenteric cyst. Neurenteric cyst can present with a vast spectrum of symptoms and can be life-threatening. When the gastric epithelium lines the cyst, hemorrhage, anemia, and pain can be the chief complaints. The majority of the children with these cyst presents with central nervous system symptoms such as back pain, sensory, or motor deficit or gait disturbances. A persistent cough and fever were the chief complaints of our case.

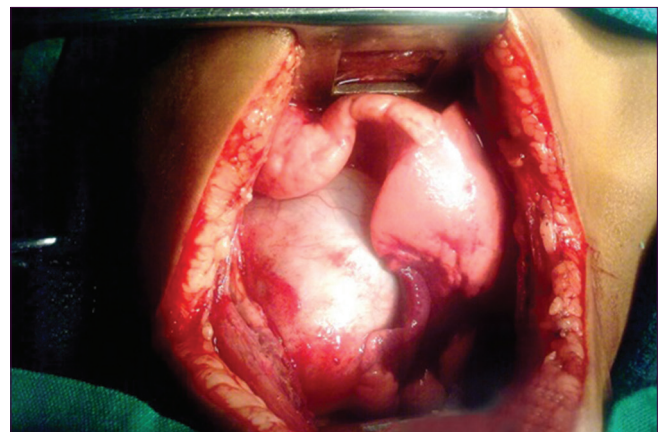
The radiological evaluation of the neurenteric cyst has evolved with advances in technology. Before MRI, CT metrizamide myelography has been the single best

diagnostic study in the diagnosis of the neurenteric cyst. CT and MRI both are very good in diagnosing the condition.

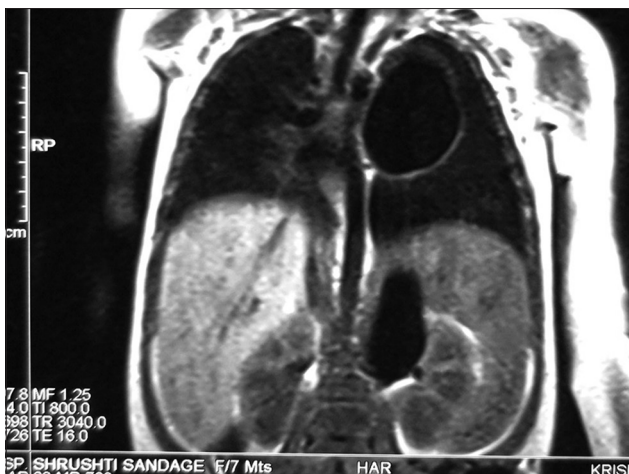
The surgery of these lesions is actually straight forward. If an asymptomatic or less symptomatic lesion is found out, elective excision is recommended unless operative risks are too high. The symptomatic lesion must undergo excision first as seen in our case. The treatment of choice remains complete excision of the cyst (Figures 6 and 7).<sup>8,9</sup> Neurenteric cyst is a type of foregut duplication cyst.<sup>10</sup>

## CONCLUSION

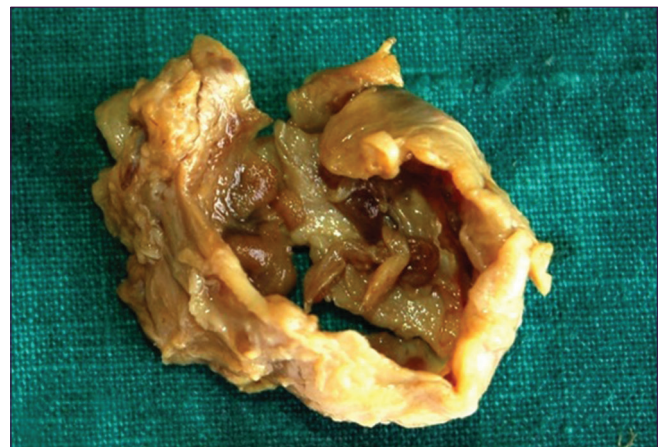
The aim of presenting this case is because of its rarity. Neurenteric cysts are one of the rare congenital disorders that present within first 5 years.<sup>11</sup> These are the enterogenous type of duplication cyst associated with vertebral anomalies. The intrathoracic cyst is within the



**Figure 6: Gross photograph showing posterior mediastinal cystic mass displacing left lung anteriorly.**



**Figure 5: Flair MRI showing the lesion is getting suppressed indicating cystic nature of the lesion.**



**Figure 7: Gross specimen of cut opened cyst showing smooth internal surface**

mediastinum, 90% posteriorly, as seen in our case and 60% of the cysts are seen superior to the carina. 66% of the cysts are seen on the right side.<sup>12</sup> Early and complete excision of the lesion is the treatment of choice.

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