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Thoracic Neurenteric Cyst Arising in Posterior Mediastinum in a Premature Male Infant- A Case Report

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Abstract | **Case Report**

Neurenteric cyst is a rare congenital malformation, which is enterogenous type of duplication cyst located anywhere in the body from intracranial to abdomen. Neurenteric cysts are formed due to failure of complete separation of notochord from the foregut during embryonal life. They are usually associated with vertebral anomalies. We present a case of neurenteric cyst arising in the posterior mediastinum in a 34 weeks male neonate with the clinical presentation of respiratory distress.

Keywords: Neurenteric cyst, Posterior mediastinum, Male infant.

INTRODUCTION

Neurenteric cysts are one of the rare congenital malformations which develop during the 3rd week of embryonal life resulting from abnormal connection between the primitive ectoderm and endoderm; and due to failure of separation of notochord from foregut[1].They manifest in any age group, but are usually detected during first 5 years after birth.We present a case of neurenteric cyst arising in the posterior mediastinum in a 34 weeks male neonate with the clinical presentation of respiratory distress.

CASE REPORT

A two day old, male premature baby was admitted in neonatal ICU for respiratory distress and abdominal distension. CECT Abdomen and Thorax revealed a fluid filled round to oval cystic thick wall structure noted in left thoracic cavity involving middle and posterior mediastinum with shift of trachea and mediastinum to contralateral side. Hemivertebrae and butterfly vertebrae were noted in upper dorsal vertebra (C7, T1 and T2).

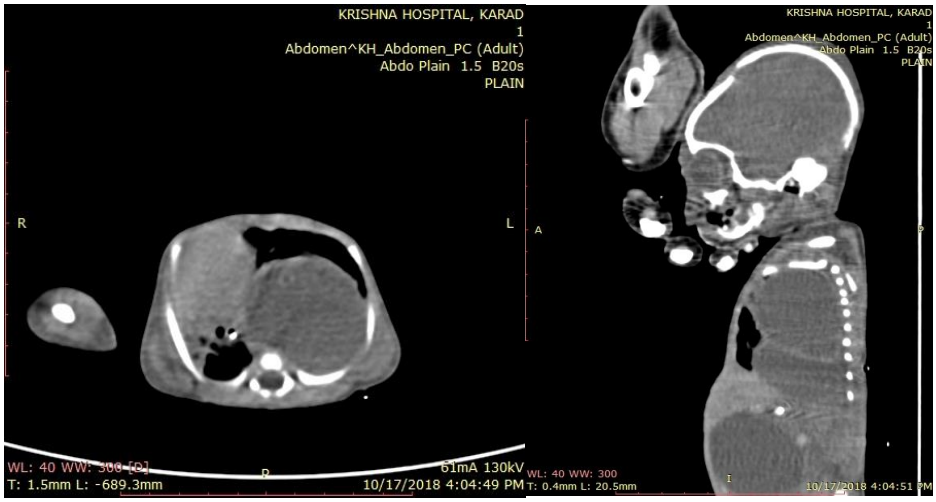

Fig-1: Axial and Sagittal view of CT scan showing cysts in left thoracic cavity involving middle and posterior mediastinum

In spite of critical care, the baby died because of respiratory distress on 2nd day of admission and was sent for autopsy.

AUTOPSY FINDINGS

External examination revealed a premature male baby with spina bifida.

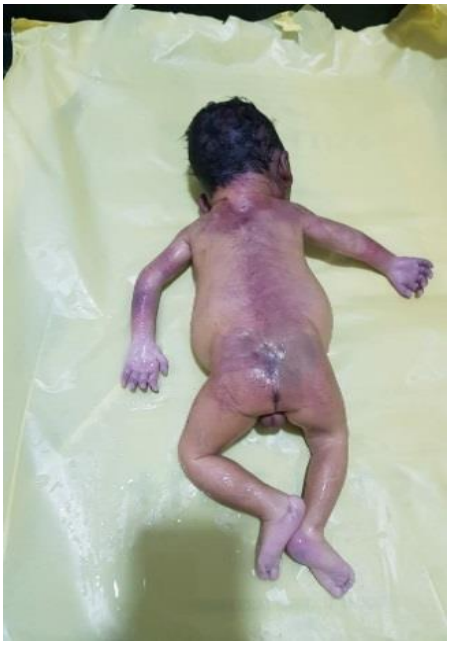

Fig-2: External examination showing spina bifida

In situ examination of thoracic cavity revealed two grey white cysts situated in posterior mediastinum, larger measuring 4.5 x 4 x 3 cm and smaller measuring 3.5 x 3 x 2 cm. cut section of cysts revealed unilocular cyst with serous fluid.

Fig-3: In situ examination of thoracic cavity showing two cysts

There was no communication of the cysts with any other organ. Hemivertebrae and butterfly vertebrae were noted in upper thoracic spine C7, T1 and T2.

Microscopy from cysts revealed a thick fibrocollagenous cyst wall lined by tall columnar epithelium resembling gastrointestinal mucosa. Considering all these features, the diagnosis was given as Neurenteric cyst with spina bifida and thoracic vertebral anomalies.

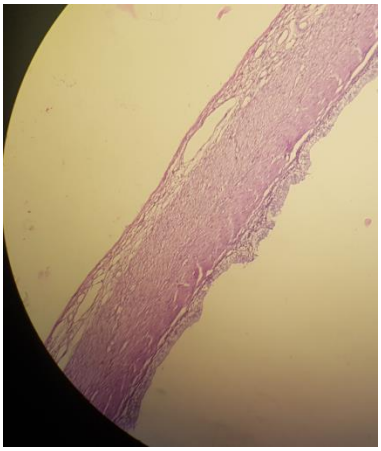

Fig-4: Microscopy of the cyst wall lined by tall columnar epithelium resembling gastrointestinal mucosa

DISCUSSION

Neurenteric cysts are developmental malformation which result from failure of complete separation of notochord from the foregut. They are associated with vertebral anomalies and hemivertebra [2]. Cervical and upper thoracic vertebrae are usually affected [3, 4]. Neurenteric cysts are commonly seen in the posterior mediastinum. Our case also revealed neurenteric cysts present in the posterior mediastinum with thoracic vertebral anomalies. The first account of neurenteric cyst was taken in 1937 in an autopsy case [5]. In 1976, neurenteric cysts were classified into 3 types by Wilkins and Odom based on microscopic features. Type A cysts are lined by pseudostratified columnar epithelium which resembles gastrointestinal or respiratory epithelium. Type B cysts show epithelial lining with complex glandular pattern producing mucin and also reveal smooth as well as striated muscle, cartilage, bone, fat, lymphoid tissue and nerve fibres. Type C cysts contain ependymal or glial tissue [5, 6]. In our case the cyst lining was mimicking gastrointestinal mucosa, hence classified as Type a neurenteric cyst.

One third of the neurenteric cystsremain asymptomatic and remaining present with respiratory distress due to their size compressing the lungs. Similar clinical presentation was seen in our case. The diagnostic features of neurenteric cyst are respiratory distress, with radiological features showing cervical or thoracic vertebral anomalies and posterior mediastinal cyst. Similar findings were noted in our case.

Primary management of neurenteric cyst is surgical resection; partial resection due to vertebral anomalies and adhesions may cause recurrence [7].

CONCLUSION

Neurenteric cyst is a rare clinical entity with a wide spectrum of congenital malformations. Correlation of clinical presentation with radiological features and histopathological examination help in making definitive diagnosis and giving better management.

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