Mediastinal Gastroenteric Duplication Cyst Presenting as Recurrent Retropharyngeal Abscess in a Child

Pushpa Gurudas Kini[1], Ganesh S Kamath[2], Sandeep Kumar[1], Deepak Nayak M[3], Karen Moras[1] and Priyanka Kini[4]

[1]Department of Paediatrics, Kasturba Medical College, Manipal Academy of Higher Education, Manipal, India

[2]Department of Cardiovascular and Thoracic Surgery, Kasturba Medical College, Manipal Academy of Higher Education, Manipal, India

[3]Department of Pathology, Kasturba Medical College, Manipal Academy of Higher Education, Manipal, India

[4]Department of Anaesthesiology, Kasturba Medical College, Manipal Academy of Higher Education, Manipal, India

Correspondence:
Sandeep Kumar
Department of Paediatrics,
Kasturba Medical College,
Manipal Academy of Higher Education,
Manipal,India.
Email: bksandydoc@gmail.com

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ABSTRACT

Mediastinal duplication cyst is a rare cause of mediastinal mass in early infancy. Recurrent retropharyngeal abscess is an uncommon presentation of mediastinal duplication cyst. We report a case of mediastinal duplication cyst in a nine year old girl who presented with recurrent retropharyngeal abscess. Surgical excision was performed successfully. The mode of clinical presentation and management has been discussed.

Key words: children; duplication cyst; mediastinal mass; neuroenteric cyst

INTRODUCTION

Gastro-enteric duplication cysts are often diagnosed in the newborn period or early infancy and are rarely seen in the older age group. The most common clinical presentation is mediastinal mass with or without respiratory distress.[1] We report a nine year old girl with mediastinal gastro-enteric duplication cyst who presented with recurrent retropharyngeal abscess.

CASE REPORT

A nine year old girl presented with prominence of the upper part of left chest which was noticed for two weeks. It was associated with fever, pain and redness over the area for three days. There was no cough, respiratory distress or stridor. The first presentation to our hospital was at five years of age with retropharyngeal abscess. There was past history of incision and drainage of the retropharyngeal abscess ten times through pharyngeal approach over a period of three years.

MR imaging of neck and chest done at the age of five years confirmed retropharyngeal abscess with prevertebral collection extending from C_4 to D_7 vertebra and revealed the presence of C_3 hemi vertebra with destruction of C_4, C_5, C_6 vertebrae. Surgical drainage was done through incision anteriorly over left side of neck. Biopsy showed evidence of granulation tissue with pigment laden macrophages. AFB (Acid fast bacilli) staining was negative. Due to the presence of destructive lesion of C_4, C_5, C_6 vertebra, the child was started on anti-tubercular drug therapy (Four drug regimen). The child was lost for follow up for next three years.

At presentation, for recurrence of retropharyngeal abscess at the age of nine years, the weight was 17 kg (below 3rd centile) and height was 120 cm (on 3rd centile). There was no neck swelling. There was prominence of the left infra-clavicular and mammary area with erythema in the left axillary region. Examination of other systems was normal. Investigations revealed normal blood counts with ESR 56 mm/hr and CRP 170 mg/L. Chest X-ray showed homogenous opacity in the left superior mediastinum (Figure 1). CT thorax revealed a well-defined, thick walled, septated cervico - thoracic cystic lesion measuring 4.6 x 4.9 x 7.0 cm extending inferiorly till upper border of D_6 level along prevertebral and left paravertebral space (Figure 2). Gastrografin study of the oesophagus ruled out aero - digestive tract fistula. Echocardiography and ultrasonography of the abdomen was normal.

Treatment was initiated with broad spectrum IV antibiotics. Left posterior thoracotomy and excision of the cyst was performed. Blackish mucoid secretion was seen in the cyst cavity. Post-operative period was uneventful. Histopathology of the cyst cavity specimen showed the presence of small bowel mucosa overlying a double layer of smooth muscle while the cyst wall revealed the presence of gastric foveolar epithelium with hypertrophied muscularis mucosa layer suggestive of gastro-

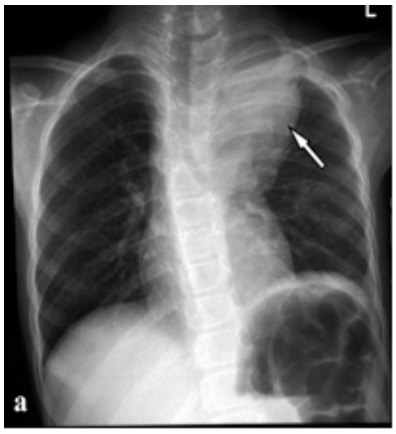

Figure 1. Chest X-ray showing mass in the left superior mediastinum

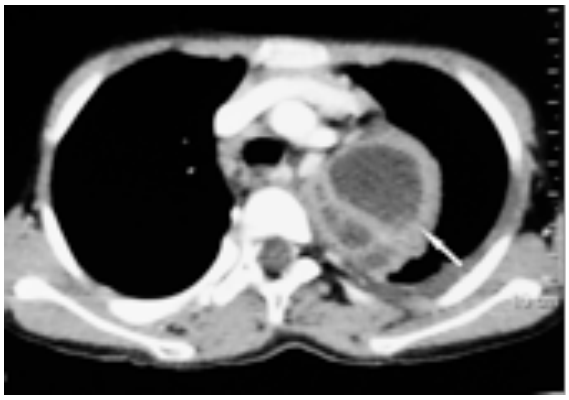

Figure 2. CT thorax showing thick walled, septate cystic mass

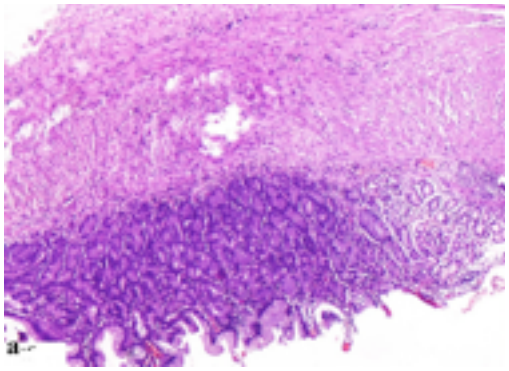

Figure 3. Cyst wall lined by gastric mucosa showing pit glands and muscularis layer (H&E; x40)

enteric duplication cyst (Figure 3). At six months follow up she is asymptomatic.

DISCUSSION

Duplication cysts can be found in any part of gastro-intestinal tract. Midgut duplication cysts are the most common (50%) followed by foregut (36%) and hindgut (12%). Other types of duplication cysts include retroperitoneal, spinal, oropharyngeal and biliary (2%). Cysts of foregut origin are further classified into oesophageal, gastric, enteric and bronchogenic cysts based on the epithelial lining of the cyst wall.[1] Gastro-enteric cyst refers to the combined presence of both gastric and enteric elements.[2] The term neuro-enteric cyst is used to describe foregut duplication cyst associated with vertebral anomalies.[3]

The clinical presentation depends on the age, location of the cyst and associated complications. Foregut duplication cysts account for 11 to 18% these mediastinal lesions.[4] The commonest clinical presentation in infancy is respiratory distress or stridor secondary to mass effect. Children present with recurrent respiratory infections, symptoms related to pressure effect such as stridor, dysphagia, chest pain, vomitings and poor weight gain. Acid secreted from the parietal cells of gastric mucosa results in peptic digestion of the cyst wall and haemorrhage.[4] Haematemesis occurs if the cyst communicates with oesophageal lumen. 15% of the cases may remain asymptomatic and diagnosed incidentally on chest imaging.[5]

The commonest site of foregut duplication cyst is right posterior mediastinum in the para-vertebral region.[6] In our case, cyst was located in the left posterior mediastinum. Vertebral anomalies including hemivertebrae, fusion defects, spina bifida and diastematomyelia are seen in up to 50% of the cases.[5,6] The case presented had C_3 hemi vertebra with destructive lesion of C_4, C_5, C_6 vertebrae. Other congenital anomalies associated include malrotation of gut, talipes equinovarus and congenital heart disease.[6]

Recurrence of retropharyngeal abscess is relatively rare in children. Recurrence should alert the presence of a congenital lesion such as aero - digestive fistulas, branchial cyst anomalies or lymphangiomas.[7] Immunodeficiency state and inadequately treated spinal tuberculosis are other causes of recurrent retropharyngeal abscess.

Chest X ray- postero-anterior view detects cystic lesion in 90% of the cases. Lateral view is helpful to diagnose vertebral anomalies.[6-8] CT chest is the modality of choice in older children. It helps in delineating the extent and borders of the cyst and its relation to the surrounding structures. MR imaging is preferred in infants as it avoids exposure to radiation. In addition, it helps to detect spinal extension precisely.[8] Definitive diagnosis depends on histopathological findings. The closest differential diagnosis includes benign mediastinal neoplasms especially cystic teratoma. The absence of sebaceous glands and hair follicles excludes its diagnosis.[8]

Surgical excision is the mainstay of treatment. The presence of pressure symptoms is an early indication for surgical removal.[9] Thoracotomy is preferred in case of large cysts or those attached to mediastinal structures or in the presence of neuro-enteric communication.[10]

CONCLUSIONS

Mediastinal foregut duplication cyst is usually diagnosed in infancy. Definitive diagnosis is by histopathology. The case discussed is distinct due to the late and unique mode of presentation. Following thoracotomy and complete excision of the cyst, there is no recurrence of the lesion.

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