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Retroperitoneal Extralobar Sequestration Of Lung With Congenital Cystic Pulmonary Airway Malformation And A Concurrent Duplication Cyst Stomach – A Rare Combination

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Abstract

An extralobar pulmonary sequestration occurring with congenital cystic pulmonary airway malformation and gastric duplication cyst is an unusual combination. We report a case of a 4 year old child wherein USG and CT findings was that of a retroperitoneal lymph cyst. Laprotomy excision was done and histopathology demonstrated three distinct lesions; extralobar pulmonary sequestration with areas of congenital cystic pulmonary airway malformation occurring together with a gastric duplication cyst.

Keywords: Extralobar, sequestration, retroperitoneal, Gastric duplication, congenital cystic adenomatoid malformation

Introduction

Bronchopulmonary sequestration is a rare malformation, and is defined as a mass of pulmonary parenchyma that does not communicate with the tracheobronchial tree and has an anomalous arterial supply, usually from a systemic vessel. [1, 2] Sequestrations may be intralobar and extralobar types. By definition, extralobar sequestration is a discrete mass of pulmonary parenchyma that lies outside the pleural investment of the lung. They can be located at any level from thoracic inlet to diaphragm and even the abdominal cavity. The anomalous tissue closely resembles immature lung and is pink-tan and spongy covered by thin translucent wrinkled pleura. Microscopically lesion consists of immature lung parenchyma with dilated bronchioles and small cystic spaces formed by abortive bronchi.[1] Area resembling congenital adenomatoid malformation are not uncommon.[1, 2, 3]

Gastric duplication (GD) is a rare anomaly and presents as a unilocular or multilocular cyst lined by gastric mucosa. [2, 4] To qualify as a duplication cyst, the following criteria need to be satisfied: lining with the gastrointestinal mucosa, attachment to the gastrointestinal tract, and the presence of a smooth muscle coat.[4] Extralobar pulmonary sequestration associated with gastric duplication cyst are very rare and there are only a few case reports in English literature.

Case Report

A 4 year old female child presented with sudden onset of multiple episodes of vomiting since one day. There was no history of fever, cough, loose stools or burning micturition. There was no relevant past history or family history. Her antenatal period was uneventful. On examination, abdomen was soft, non tender and bowel sounds heard.

USG abdomen showed a well encapsulated multiloculated cystic lesion in anterior pararenal space and the possibility of retroperitoneal lesion was considered.

CECT abdomen (Figure 1) was done and plain and contrast enhanced serial helical sections of the abdomen were stuided which revealed a large retroperitoneal multiloculated cystic lesion on left side with thin/thick septations and solid components in close relation to the tail of the pancreas and posterior wall of stomach displacing the pancreas anteriorly and inferiorly. The lesion was extending upto the left diaphragmatic crus posteriorly. The spleen and bowel loops are displaced laterally and ipsilateral kidney is displaced inferolaterally by the lesion. All other organs were normal in size with no focal lesions. Based on the radiological findings the features were likely to represent a retroperitoneal lymphangioma with differential diagnosis of dermoid cyst / enteric duplication cyst.

Following which an open laprotomy was done, lesser sac opened and the whole cyst was excised and sent to histopathology. We received a pale brown encapsulated nodular spongy tissue measuring 4.5x3.5x2.5 cm with an attached cyst measuring 4x3x1 cm. Cut section of the nodular tissue was brownish and spongy showing pale brown septae. The attached cyst, on cut section was pale brown with mucosa showing rugal folds (Figure 2).

On microscopy, the encapsulated nodular lesion was composed of immature lung tissue with variably dilated bronchiloes and alveoli (Figure 3). Sections from the adjacent cyst wall show normal gastric mucosa and underlying musculature (Figure 4). The final histopathological diagnosis was given as a retroperitoneal extralobar sequestration of lung tissue with areas showing congenital cystic adenomatoid malformation and the attached cyst was consistent with duplication cyst stomach. Further, the child was investigated for any other congenital anomalies and is on routine followup.

Discussion

Lung sequestration is a nonfunctioning lung tissue which is not in continuity with the tracheobronchial tree and deriving its blood supply from systemic circulation. It constitutes 0.15–6.4% of all congenital pulmonary malformations.[3, 5] Extralobar pulmonary sequestration occurs more commonly in males than females with a male-to-female ratio of 4:1.[3] Clinical manifestations usually occur in the newborn period or early infancy with signs and symptoms of respiratory distress, repeated infections, and spontaneous pulmonary or pleural hemorrhage. Complete surgical excision of the extralobar sequestration is the main treatment.[3, 5] Congenital anomalies are more frequently seen in patients with extralobar lung sequestration than intralobar sequestration. These anomalies include congenital cystic adenomatoid malformation, congenital diaphragmatic hernia, vertebral anomalies, congenital heart disease, pulmonary hypoplasia, colonic duplication, tracheoesophageal fistula, bronchogenic cyst, pericardial defects, and congenital megacolon. The most common congenital anomaly being congenital diaphragmatic hernia with the esophagus or stomach. In approximately 15 – 25% of patients with extralobar sequestration, Type II congenital cystic adenomatoid malformation of the lung has been reported. [1, 3]

Gastrointestinal duplication is a rare congenital malformation, which is more commonly seen in ileum (35%), stomach (9%) and colon (7%). Gastric duplication (GD) commonly occurs in female, and with a male-to-female ratio of about 1:2-8.[4, 6] GD can be divided into two types - cyst type and tubular type, with the former being more common.[1, 4] There are no specific clinical manifestations for GD, and the common clinical manifestations include nausea, vomiting, difficulty in feeding, vague stomachache, abdominal mass, and weight loss. Hence, clinical diagnosis depends greatly on imaging. [4, 6] At present, there are many opinions on which kind of imaging examination leads to a diagnosis. Some studies consider that CT is the optimal imaging method [7], wherein it is seen as a mass with smooth border, fluid attenuation in the cavity, thick cyst wall with one side adhering closely to the gastric wall, and an intensified border; while in other studies it is believed that CT can hardly diagnose the nature of mass in an accurate way. [8]

Conclusion

A gastric duplication cyst occurring together with extralobar pulmonary sequestration with congenital cystic adenomatoid malformation is an unusual

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combination. A multidisciplinary approach aids in proper management, and surgical resection is the definitive management for all three anomalies.

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Figure 1: CECT Abdomen showing large retroperitoneal multiloculated cystic lesion on left side in close relation to the tail of the pancreas and stomach

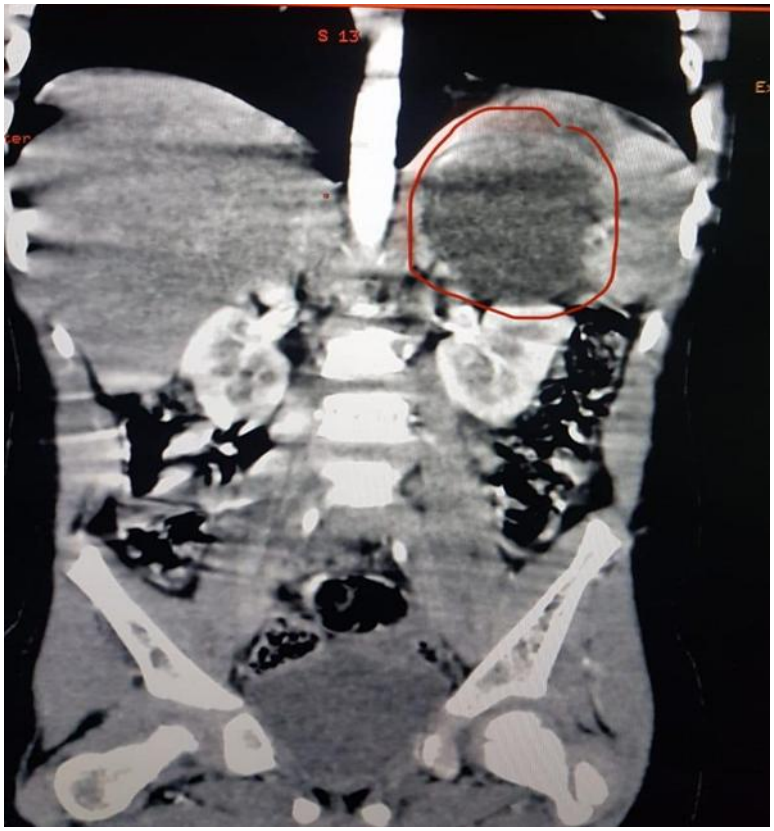

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Figure 2: Gross picture showing an encapsulated nodular spongy tissue with an attached cyst with mucosa showing rugal folds

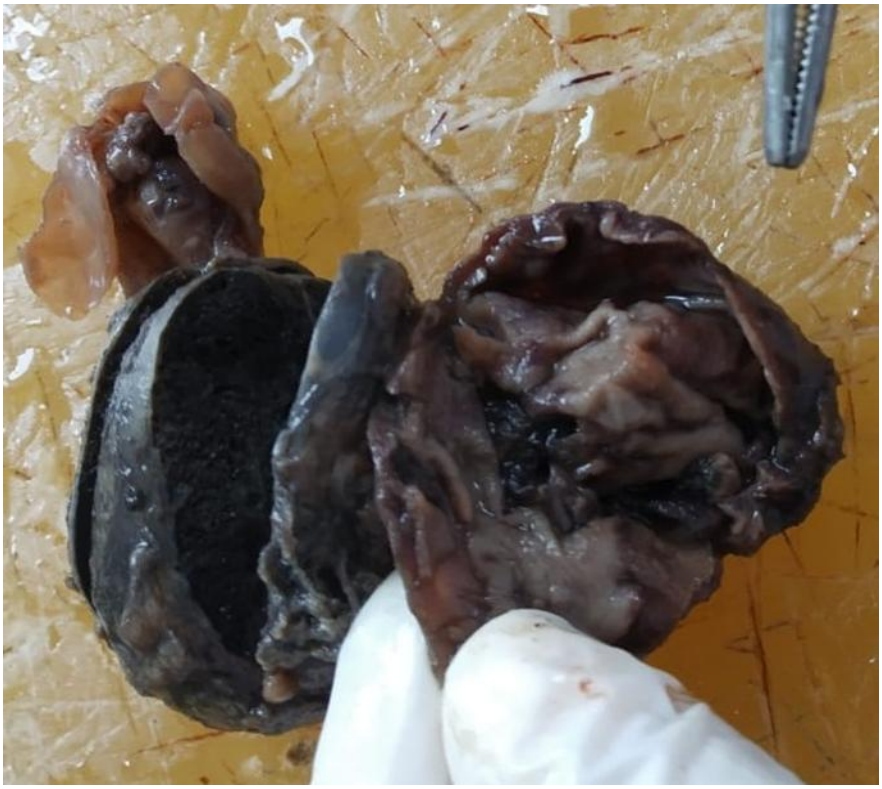

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Figure 3: Photomicrograph showing immature lung tissue with variably dilated bronchiloes and alveoli (H&E, 10x)

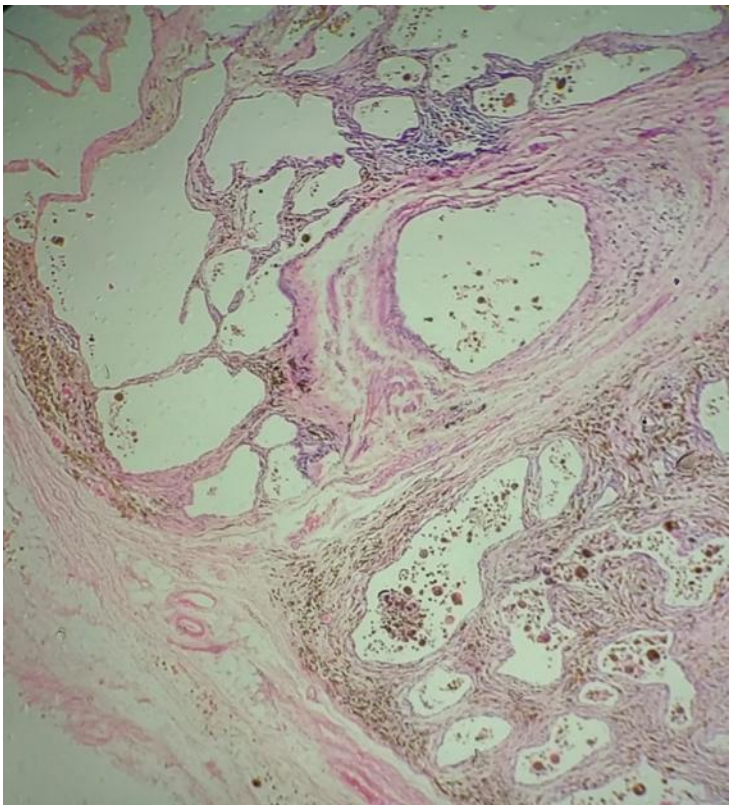
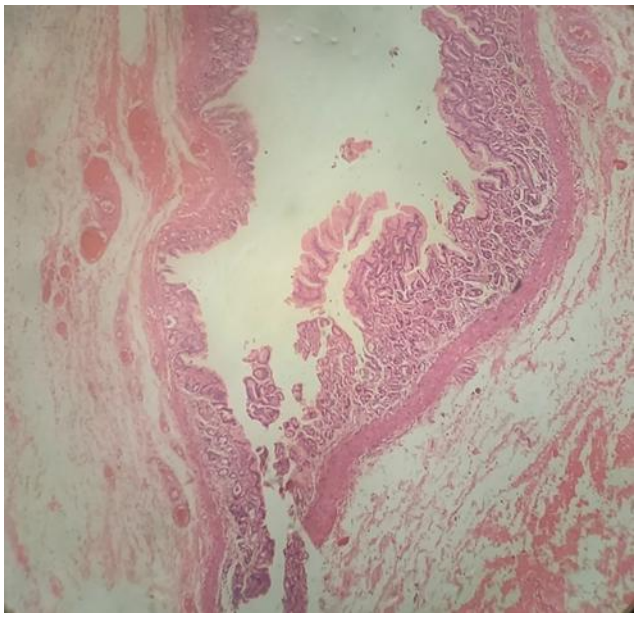


Figure 4: Photomicrograph from attached cyst wall showing normal gastric mucosa and underlying musculature (H&E, 10x)



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