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Surgery

CASE REPORT - CONGENITAL DIAPHRAGMATIC HERNIA WITH PULMONARY SEQUESTRATION

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ABSTRACT A Congenital Diaphragmatic Hernia (CDH) is a congenital malformation of the diaphragm in which abdominal content herniate in thoracic cavity, resulting in respiratory distress and feeding difficulties in the newborn. Pulmonary sequestration is a rare association and found in only 15%-30% of patients with congenital diaphragmatic hernia. Extralobar sequestration are commonly associated with diaphragmatic hernia. We present diagnosis and management of one month old male child with left diaphragmatic hernia with intralobar pulmonary sequestration.

KEYWORDS : .

INTRODUCTION

Pulmonary sequestration is defined as the presence of a mass of abnormal lung tissue that does not communicate with the tracheobronchial tree through a normally located bronchus and is supplied by an anomalous systemic artery. An association of congenital diaphragmatic hernia with pulmonary sequestration is rare. Very few cases have been reported in literature. There are two types of pulmonary sequestration, intralobar and extralobar. Extralobar sequestration is more commonly associated with anomalies like CDH, congenital pulmonary airway malformations, pulmonary hypoplasia, congenital lobar emphysema, bronchogenic cyst, congenital heart disease, vertebral anomalies, and intestinal duplications.[6] We report a case of congenital diaphragmatic hernia which was associated with intralobar sequestration.

CASE REPORT

One month old male child presented to our institute with upper respiratory tract infection and respiratory distress of 15 days duration. There was history of refusal to feeding. Antenatal history was insignificant, ultrasound scans being normal. After optimum stabilization baby was evaluated. Chest radiograph revealed elevated left hemidiaphragm, suggestive of eventration or diaphragmatic hernia. Ultrasonography of chest and abdomen confirmed left sided congenital diaphragmatic hernia with suspected pulmonary sequestration with an anomalous transdiaphragmatic vessel supplying the sequestration.

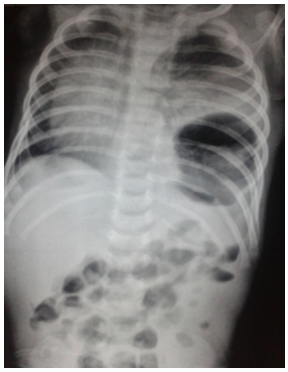

Figure 1 – Chest x-ray of the patient

Further evaluation with CT Angiogram confirmed presence of congenital diaphragmatic hernia with intralobar pulmonary sequestration which was supplied by anomalous vessel arising from descending aorta, and drained by pulmonary vein.

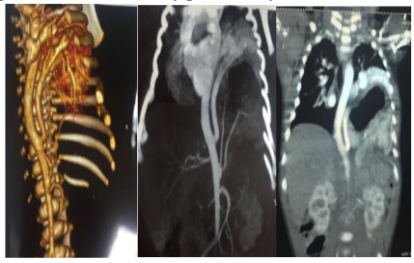

Figure 2 – CT angiogram showing the lung sequestration

Patient was taken up for surgery after evaluation. Intraoperatively hernial sac was found to be absent with small bowel and part of large bowel as content. Lower lobe sequestration was excised carefully and diaphragmatic defect was closed with nonabsorbable suture.

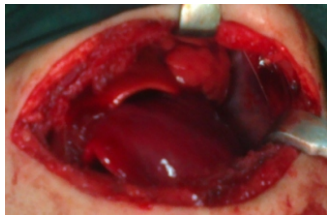

Figure 3 – Intraoperative photograph

Postoperative recovery was uneventful and patient discharged on postoperative day 10. Patient is on regular follow-up and doing well with adequate weight gain.

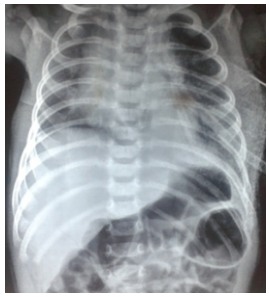

Figure 4 – Post operative radiograph of the patient

DISCUSSION

Pulmonary sequestration is a rare congenital lesion consisting of non-functioning lung tissue that is not connected to the normal tracheobronchial tree and is supplied directly from the systemic circulation [5]. Extralobar sequestration receive arterial supply from an aberrent vessel from aorta and venous drainage into systemic veins to the right atrium, vena cava, azygous vein; whereas intralobar gets arterial supply from lower thoracic, upper abdominal aorta and venous drainage into left atrium via Pulmonary veins. The relationship between pulmonary sequestration and congenital diaphragmatic hernia may be explained by the hypothesis that pulmonary sequestration, which develops at 4–5 weeks of gestation, can disturb the fusion of the diaphragm and closure of the pleuro-peritoneal canal occurring at 10 weeks of gestation [8]. Congenital diaphragmatic hernia (CDH) is accompanied by other malformations in about 25% cases.[9] Antenatally, bronchopulmonary sequestration can be complicated by nonimmune fetal hydrops, or hydrothorax.[11,12] Postnatally, it may be complicated by infection and rarely infarction or hemothorax.[10] The prognosis of intralobar sequestration in the absence of severe anomalies is good. But the associated pulmonary hypoplasia can be fatal if severe.[14] When associated with CDH, the prognosis depends on the severity of pulmonary hypoplasia.[13]

To conclude, congenital diaphragmatic hernia and the associated fetal lung lesions are interrelated. The possibility of lung lesions should be considered in the patients of congenital diaphragmatic hernia. Antenatal or early postnatal diagnosis and management are essential to reduce morbidity and mortality.

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