Case Report

Extralobar sequestration of lung associated with congenital diaphragmatic hernia and malrotation of gut

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ABSTRACT

Extralobar sequestration of lung is a rare congenital malformation frequently diagnosed during repair of congenital diaphragmatic hernia. However, the combined association of congenital diaphragmatic hernia with both pulmonary sequestration and malrotation of gut is rare. We report a case of a 1-year-old girl with extralobar sequestration of lung and malrotation of gut detected during the repair of diaphragmatic hernia. The histopathological examination of the sequestered lobe revealed dilated bronchioles, alveolar ducts and alveoli along with dilated subpleural and peribronchiolar lymphatics and areas of type II congenital pulmonary airway malformation.

KEY WORDS: Congenital diaphragmatic hernia, congenital pulmonary airway malformation, extralobar sequestration of lung, malrotation of gut

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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.[1] It constitutes 1.1-1.8% of all pulmonary resections.[2] It is of two types, namely intralobar and extralobar. Intralobar sequestration is an acquired lesion that usually presents beyond 20 years of age and is rarely associated with other malformations. It is present within the visceral pleura of a lung. Extralobar sequestration is a congenital abnormality and is frequently associated with other congenital malformations. It lies outside the visceral pleura and is enclosed within its own pleural covering.[1,3]

We present the clinicopathological findings of a left sided intrathoracic extralobar pulmonary sequestration associated with congenital diaphragmatic hernia and malrotation of gut in a 1-year-old girl.

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CASE REPORT

A 1-year-old girl presented with breathlessness and failure to gain weight since birth. Breathlessness was relieved by upright position. The child was born by normal vaginal delivery at home. The milestones were delayed. On examination, the child was pale and afebrile with breathlessness and irritability. The pulse rate was 100 per minute and respiratory rate was 50 to 70 per minute. On systemic examination, there was decreased air entry and presence of crepitations on the left side of thorax. Chest X-ray was consistent with pulmonary hypertension and showed left-sided diaphragmatic hernia for which she underwent surgery.

Intraoperatively, there was a large diaphragmatic rent with migration of more than 50% of small intestine, spleen and a part of large intestine into the left thoracic cavity. There was also evidence of malrotation of gut with colon on the left side and small intestine on the right side. The left lung was hypoplastic. A separate lobe of lung with no bronchial communication was seen in left hemithorax which was attached to diaphragm [Figure 1a] and was supplied by descending thoracic aorta. The sequestered lobe was resected and was sent for histopathological examination. The contents of the diaphragmatic hernial sac were reduced and the diaphragmatic rent was repaired.

Unfortunately, condition of the patient deteriorated and she died 2 hours after the surgery due to terminal cardiorespiratory failure due to pulmonary hypoplasia with pulmonary hypertension.

Gross examination

We received an ovoid mass of lung tissue measuring 6 × 3.5 × 1.6 cm for histopathological examination [Figure 1b]. It was completely covered by pleura and showed a vascular pedicle without bronchus [Figure 1c]. Pleural surface showed congestion. Cut surface showed few tiny cysts.

Microscopic examination

Histopathological examination revealed lung tissue with dilated bronchioles, alveolar ducts and alveoli [Figure 2a]. Some of the bronchioles were tortuous with undulating cuboidal to columnar epithelium [Figure 2b] and abnormal cartilage plates [Figure 2c]. Some areas were characteristic of type II congenital pulmonary airway malformation [Figure 2d] and showed presence of closely situated irregular bronchiole like cystic spaces lined by cuboidal to columnar epithelium. The walls of the spaces contained smooth muscle fibers. The subpleural and peribronchiolar lymphatics appeared dilated mimicking congenital lymphangiectasia [Figure 2e and f].

DISCUSSION

Extralobar pulmonary sequestration (ELS) is a congenital abnormality that results from a supernumerary or separated lung bud from the developing tracheobronchial tree.[1] It is commonly accompanied by other congenital anomalies, and is frequently detected incidentally during the repair of diaphragmatic hernia. Other associated anomalies include pericardial defects, pectus excavatum, congenital heart disease and pulmonary abnormalities like pulmonary hypoplasia, congenital pulmonary airway malformation (CPAM) or congenital lobar emphysema.[3,4]

Congenital diaphragmatic hernia (CDH) is accompanied by other malformations in about 25% cases.[1] Our patient also had malrotation of gut with colon on left side and small intestine on the right side. The combined association of CDH with both pulmonary sequestration and malrotation of gut has been rarely reported.[5] The presence of intestine in the thorax during late fetal life causes malrotation.[6] There is also a report of two cases with pulmonary sequestration, bronchogastric fistula, malrotation of intestine and Meckel's diverticulum.[7]

Patients with CDH also have pulmonary hypoplasia and

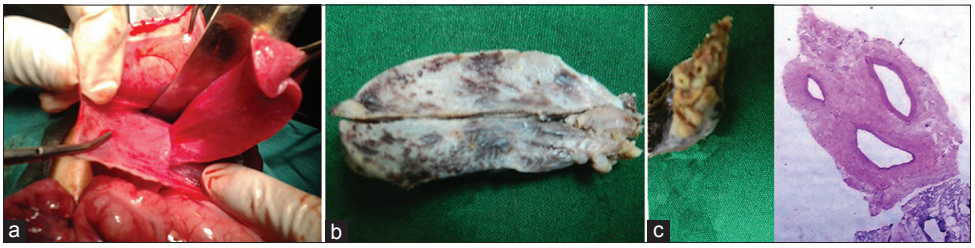

Figure 1: (a) Intraoperative photograph showing the sequestered lobe of lung attached to diaphragm. (b) Gross photograph of sequestered pulmonary lobe. (c) Gross and microscopic (H and E ×10) photograph of pedicle

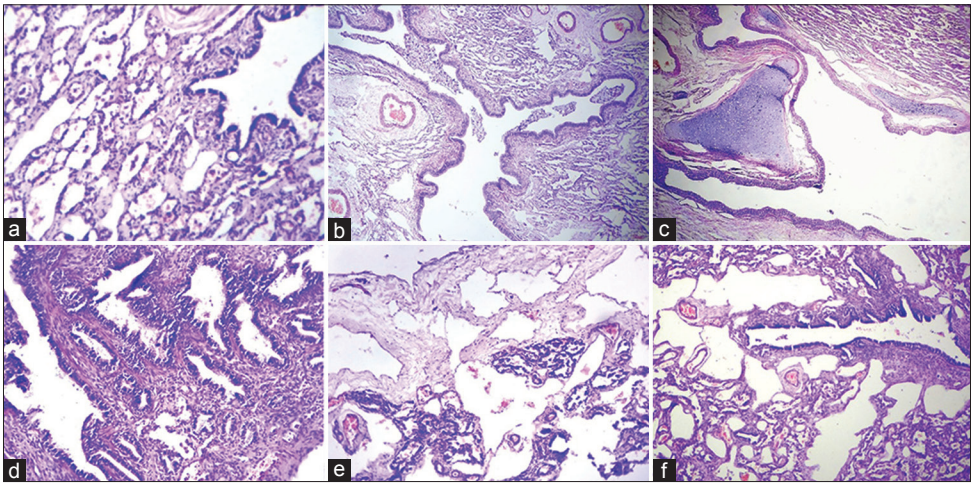

Figure 2: (a) Photomicrograph showing dilated bronchioles, alveolar ducts and alveoli (H and E ×100). (b) Photomicrograph showing tortuous bronchiole with undulating epithelium (H and E ×40). (c) Photomicrograph showing bronchiole with abnormal cartilage plates (H and E ×40). (d) Photomicrograph showing areas of congenital pulmonary airway malformation (H and E ×100). (e) Photomicrograph showing dilated subpleural lymphatics (H and E×100). (f) Photomicrograph showing dilated peribronchiolar lymphatics (H and E ×100)

pulmonary hypertension as in our case.[6] Pulmonary hypoplasia results from abnormal development followed by compression due to herniated viscera.[6,8,9] Pulmonary hypertension is due to reduced number and generations of airways, abnormal muscularization of the intra-acinar pulmonary arteries, reduced pulmonary vascular bed and abnormal pulmonary vasoconstriction.[8,10]

The association of ELS and CPAM is well documented in the literature. Forty percent of the extralobar sequestrations show histology of CPAM type 2.[1] Dilated subpleural lymphatics resembling congenital lymphangiectasia as in our case are also seen in over 33% of cases of ELS.[1]

CPAM may be present in the lung of the same or opposite side of the chest or within the sequestration.[11,12] The cases of ELS with associated CPAM (ELS/CPAM) differ in presentation from ELS cases that are not associated with CPAM. Conran and Stocker found that the ELS/CPAM cases were more frequently diagnosed within first 3 months of life and were more frequently seen on the left side.[13]

Antenatally, bronchopulmonary sequestration can be complicated by nonimmune fetal hydrops, or hydrothorax.[8,14] Postnatally, it may be complicated by infection and rarely torsion, infarction or hemothorax.[3] The prognosis of extralobar sequestration in the absence of severe anomalies is good. But the associated pulmonary hypoplasia can be fatal if severe.[11] When associated with CDH, the prognosis depends on the severity of pulmonary hypoplasia and therapy-resistant pulmonary hypertension.[9] Our patient had CDH with both pulmonary hypoplasia and pulmonary hypertension.

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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