Infradiaphragmatic Extralobar Pulmonary Sequestration: Masquerading as Suprarenal Mass

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ABSTRACT
Pulmonary sequestration is a rare malformation, wherein a portion of lung is non-functional and is not in normal continuity with the tracheo-bronchial tree, and may derive its blood supply from systemic vessels. Two types are described: Intralobar and extralobar types. Intralobar sequestration is more common type, which shares visceral pleura of the involved lobe and is localized within the normal pulmonary parenchyma. Whereas extralobar forms are uncommon and are totally separate from the lung and usually have own covering. Infra-diaphragmatic pulmonary sequestration is of extralobar type and is extremely rare, and usually is associated with other congenital malformations. We present an extremely rare case of isolated infra-diaphragmatic pulmonary sequestration which was antenatally detected and followed up with postnatal CT scan, where it masqueraded as suprarenal mass, and was surgically treated. This case emphasises to add a differential diagnosis of malformation in congenital supra-renal masses, which remain stable in size and appearance, and hence avoid immediate surgery.

Key words: Extralobar, intralobar, sequestration, sub-diaphragmatic, suprarenal mass

INTRODUCTION
Pulmonary sequestrations are rare malformations encountered in approximately 1-1.8% of all lung resections. Infra-diaphragmatic pulmonary sequestration belongs to the extralobar type, and most cases have been reported in association with other congenital malformations. We report a rare case of sub-diaphragmatic retroperitoneal extralobar sequestration without other associated malformations, which was detected antenatally and masqueraded as a suprarenal tumor.

CASE REPORT
Antenatal ultrasound scan of single live intrauterine fetus at 20 weeks in a primigravida was found to have a well-defined echogenic mass in left suprarenal area consistent with radiological diagnosis of neuroblastoma. The mothers’ medical history was unremarkable. This lesion was stable in size and morphology on two subsequent follow-up ultrasounds. A 2.96 kg full term live male baby was delivered by caesarian section. His physical examination was unremarkable with no other malformations.

Contrast-enhanced chest and abdominal computed tomography (CT) scan on postnatal day 3 showed a well-defined lobulated, mildly enhancing, heterogeneous mass in left suprarenal area, which was comparable with prenatal size (3.5 × 2.3 × 3.1 cm). The lesion was predominantly solid with a small cystic component and with no calcification [Figure 1]. Medially the mass was abutting the aorta with a small feeding vessel from the aorta [Figure 2]. The left hemi-diaphragm was indented and with compression on gastric fundus. Left adrenal gland was not visualized, and there was mild inferior displacement of the left kidney. Radiological differential diagnosis of retroperitoneal teratoma (due to cystic component) was added to the initial diagnosis of neuroblastoma. No other malformations were noted in the chest and rest of the abdomen and pelvis on CT scan.

Laprotomy and complete excision of the mass and left adrenalectomy was done on 5th postnatal day. The mass was adherent to left diaphragm and stomach fundus. Diaphragm was dissected uneventfully, however, stomach wall was perforated and had to be sutured. Baby withstood surgery well and recovery was uneventful.

Histopathological examination (HPE) of removed mass...
Figure 3] revealed cystic spaces of varying diameter lined by columnar epithelium with cartilaginous tissue around it resembling bronchi and bronchioles. Surrounded tissue had dilated spaces lined by low cuboidal epithelium resembling alveoli. Hemorrhage and macrophages were seen in lumen of alveoli. There was thin fibrous tissue all around the lesion. Features were consistent with pulmonary sequestration. Section from adrenal tissue was unremarkable.

**DISCUSSION**

Pulmonary sequestrations are nonfunctional lung tissue, which lacks normal communication to the trachea-bronchial tree and usually has its blood supply from systemic vessels.[1,2] On the basis of the morphologic patterns, it is further classified as intralobar or extralobar types. Intralobar sequestration is more common type (75-85% of cases) and is localized within the normal lung parenchyma and shares visceral pleura of the involved lobe. Extrapulmonary forms are uncommon (15-25% of cases) and are separate from the normal lung and will have their own pleural covering.[1-5] Sequestrations may have communication with the esophagus or stomach and hence have been also included in the spectrum of broncho-pulmonary-foregut malformations. The original communication with foregut usually regresses, but occasionally may persist as patent tract or as obliterated fibrous cord.[6]

Extrapulmonary pulmonary sequestrations are most commonly found intrathoracically within the pleural space and usually on the left side and typically in the posterior costophrenic sulcus. They may be rarely found in the mediastinum. Only 10-15% of extrapulmonary sequestrations are found below the diaphragm.[5] In extrapulmonary sequestrations reported incidence of various other congenital anomalies is 50-65%.[5] Most common associated anomalies are those of the diaphragm like congenital diaphragmatic hernia, diaphragmatic eventration, etc.[1,2,7] Other frequently associated anomalies are bronchogenic cyst, congenital cystic adenomatoid malformation, foregut duplication, or diverticulum. Cardiovascular, genitourinary, and gastrointestinal anomalies have been rarely reported.[6,8] Extrapulmonary sequestrations typically (80% of cases) draw blood supply directly from thoracic or abdominal aorta (systemic arteries). In 15% of cases it is by smaller systemic arteries like splenic, gastric, subclavian, and intercostal branches. Rarely (<5% of cases) it may receive supply from branches of the pulmonary artery or by both the pulmonary and systemic circulations.[4,6,7] Venous drainage is commonly via systemic veins into the right atrium (80% of cases) via azygos-hemiazygos veins or vena cava. Uncommonly, it may drain into the portal, intercostal, suprarenal, or other abdominal veins. Very rarely, drainage may happen into left atrium through the pulmonary veins.[4]
Extralobar sequestration in intraabdominal location is extremely uncommon. In most cases it has been reported close to the diaphragm, usually in the suprarenal area. Other congenital malformations has been reported in 50% of cases. [3,9] Diagnosis of intraabdominal pulmonary sequestration is difficult and is mostly being diagnosed during the histopathologic study of a surgically resected lesion. Radiological differential diagnosis includes suprarenal neuroblastoma, teratoma, foregut duplication, adrenal hemorrhage, and mesoblastic nephroma. [6] However, in congenital suprarenal masses with atypical features like cystic changes, possibility of sequestration also may be considered, more so when they remain stable in size and morphology for many weeks. Very rarely in those having air or communication to stomach, the diagnosis of sequestration is straightforward.

Intralobar sequestrations are seen mostly within the lower lobes, and more on the left side. At radiological examinations, they typically appear as consolidation or mass and sometimes with cavitations. [4] A systemic arterial supply helps in the diagnosis. Venous drainage is mostly into pulmonary veins and to the left atrium. [4,6]

It is remarkable that our case of intraabdominal sequestration was an isolated malformation without other associated anomalies. It had few atypical features like cystic change, absence of calcification, and had remained stable throughout the course (20 weeks). Arterial supply from abdominal aorta was noted, but was interpreted as tumor feeder. Firm gastric adhesion at surgery was probably an obliterated fibrous communication, seen in these broncho-pulmonary-foregut malformations. In our case, a differential diagnosis of congenital malformation, and probably Fine Needle Aspiration Cytology (FNAC) or biopsy could have postponed or avoided an immediate surgery.

CONCLUSION

Extralobar intraabdominal pulmonary sequestration as an isolated anomaly is extremely rare. Usually it is associated with various other congenital anomalies. Radiological diagnosis is extremely difficult, most cases being diagnosed during the histopathologic study of a resected mass. However, in stable congenital suprarenal mass with atypical features, a diagnostic consideration of congenital malformation may postpone or avoid a needless urgent surgery.

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