A 3-year-old Girl with Recurrent Respiratory Tract Infections

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Section 2 – Answer

Case

A 3-year-old girl was referred to radiology clinic with complaints of cough, fever (38,5°C), sputum production, and respiratory distress. It was learned that the patient was previously followed up in the hospital due to recurrent respiratory tract infections. Physical examination of the patient revealed a decrease in respiratory sounds in the lower parts of the left lung during auscultation. Significant laboratory tests result: white blood cell count 16,350/μL, C-reactive protein 9, 8 mg/L. Ultrasonography (US) and a computed tomography (CT) was performed. On US a hyperechogenic, well-circumscribed mass, with cystic areas was detected in the basal part of the left lung [Figure 1].On color Doppler US, a branch extending from the thoracic aorta to the lesion was seen [Figure 2]. On CT images, branches of the thoracic aorta and the pulmonary vein extending toward the lesion were detected [Figures 3 and 4].

Interpretation

US and CT images revealed heterogeneous mass located at the basal part of the left lung, feeding from the thoracic aorta and containing cystic areas smaller than 2 cm. It was defined as a hybrid lesion because it contains both congenital cystic adenomatoid malformation (CCAM) and pulmonary sequestration (PS) features. Lobectomy treatment was recommended to the patient because of the poor prognosis of hybrid lesions. After the lobectomy hybrid lesion, diagnosis was confirmed with pathology result.

Discussion

Congenital bronchopulmonary malformations are rare diseases. PS is the second-most common congenital lung lesion detected antenatally (0.15%–6.45%).It is characterized by a part of the lung that does not adhere to the tracheobronchial tree and is supplied systemically from the thoracic or abdominal aorta (rarely from celiac, spleen, intercostal, subclavian artery).[1]

Two types of PS have been described: intralobar (ILS) (%75) and extralobar (ELS) (%25). Both ILS and ELS are more common in the left lung. The intralobar type is surrounded by the visceral pleura leaf of the lung and usually drains into the pulmonary venous system. The extralobar type has its own pleura leaf and systemic venous drainage.[2] The localization of ELS can be intrathoracic or subdiaphragmatic.

It has been reported that the incidence of anomalies associated with extralobar sequestration (65%) is higher than intralobar sequestration (11%).[3] Extralobar sequestration may be associated with congenital systemic anomalies such as congenital diaphragmatic hernia, heart abnormalities, pulmonary hypoplasia, or foregut duplication cysts.[1]

An abnormal source of systemic arteries can be seen in CCAMs.[4] Such lesions are often referred to as hybrid lesions and show imaging features of both CCAM and PS. The incidence of hybrid lesions is very low and association with ELS is more common in cases reported in the literature.[5] Hybrid lesions are more common on the left side. Most of CCAM cases were found to have a type 2 pattern on histological examination (small [0.5–2 cm] cyst bronchiolar origin lesion in type 2).[6] While PS is 3 times more common in males, hybrid lesions are more common in females.

Patients with PS may be asymptomatic at birth, but later develop cough, hemoptysis, and recurrent pneumonia, or may remain asymptomatic and be diagnosed incidentally. In prenatal US, the hybrid lesion is seen as a cystic hyperechoic mass in a paraspinal localization, mostly in the left lower thorax. Feeding artery arising from the descending aorta can be seen on color Doppler US. In some patients, these vessels may not be identified in Doppler US, making sequestration indistinguishable from a microcystic CCAM.[7] Diagnosis is confirmed by prenatal

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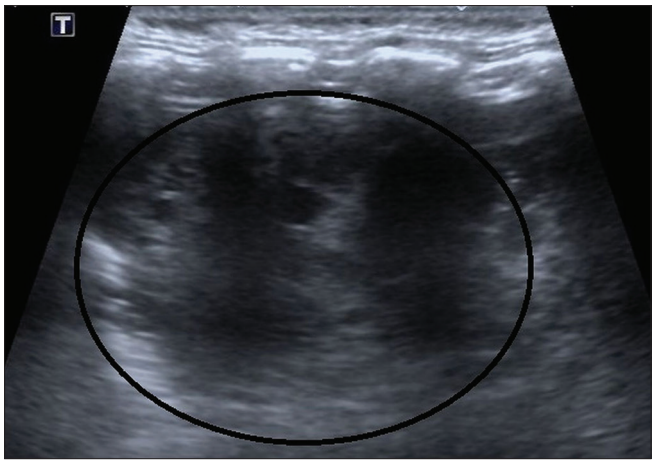

Figure 1: Axial sonography showing a cystic heterogen-hyperechogenic mass in the left lower lobe (inside the circle)

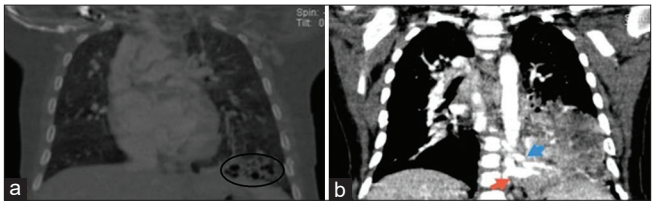

Figure 3: Coronal computed tomography image lung window (a) and soft-tissue window (b), There is a cystic mass in the left lower lobe (inside the circle), inside the lesion branches of thoracic aorta (red arrow) and pulmonary vein (blue arrow)

MR imaging.[8] In the neonatal period, diagnosis can be made by posteroanterior chest radiography (posteroanterior) and Doppler USG. Demonstrating systemic arterial nutrition and abnormal venous drainage with contrast-enhanced thoracic CT, magnetic resonance imaging, and angiography in older children is diagnostic.

The treatment of lung sequestration is surgical. Segmental resection of ELS is easier because it is completely surrounded by the pleura. ILS treatment is the segmental resection of sequestration, but lobectomy is generally compulsory because of chronic infection-related changes.[9] The treatment of hybrid lesions is segmental resection or lobectomy of the pulmonary lesion to prevent complications such as respiratory distress, infection, intrathoracic bleeding, hemoptysis, heart failure, and potential malignancy.[10]

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the legal guardian has given his consent for images and other clinical information to be reported in the journal. The guardian understands that names and initials will not be published and due efforts will be made to conceal patient identity, but anonymity cannot be guaranteed.

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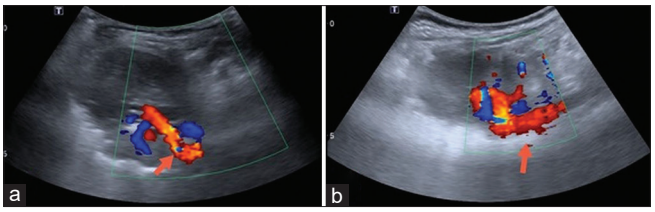

Figure 2: (a) axial and (b) longitudinal color Doppler ultrasonography images showing blood supply of the lesion originating from the thoracic aorta

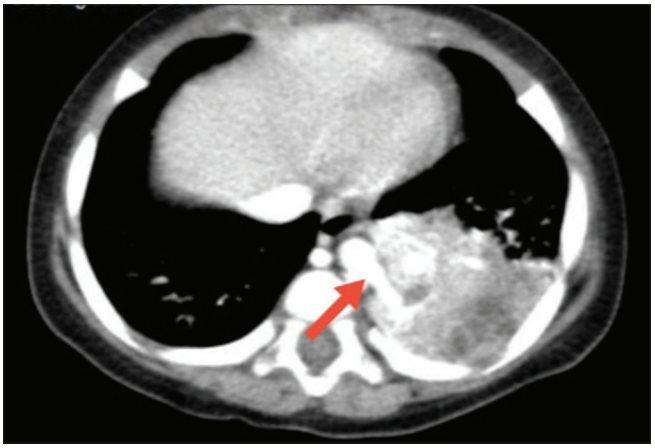

Figure 4: Axial computed tomography image, a cystic mass in the left lower lobe and a branch of the thoracic aorta (red arrow) extending into the lesion

Conflicts of interest

There are no conflicts of interest.

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