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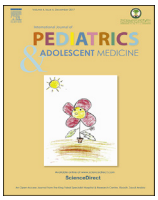

Images in pediatrics

Congenital cystic adenomatoid malformation

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ABSTRACT

Congenital pulmonary airway malformation of the lung is a rare lesion that commonly affects the lower respiratory tract, which manifests as progressive respiratory distress in the neonatal period. This case is highlighted to raise awareness of the variable evolving morphological and radiological appearance of this entity.

1. Case summary for images in pediatrics

A full term female patient was born in our hospital. The baby was suspected to have cystic parenchymal lung abnormality according to antenatal ultrasound. The baby was admitted to the NICU and she was clinically stable, maintaining saturation on room air. A chest radiograph was obtained on day 0 (Fig. 1) and it indicated no obvious cystic changes. Another chest radiograph was obtained on day 2, which also showed no definite lung cystic abnormality. The patient was discharged and given an appointment at a pediatric pulmonology clinic. Two weeks later, the patient presented to the ER with a history of coughing, vomiting, cyanosis, and fever for three days, so a chest radiograph was obtained. Fig. 2 shows the development of cystic changes in the background of air space opacity in the right middle lung zone. A CT scan (Fig. 3) of the chest was performed which detected multiple variable sized cysts in the right lower lobe, where the largest measured 1.16 cm, with no abnormal arterial supply or venous drainage. Overall, the findings were compatible with right lower lobe type 2 congenital cystic adenomatoid malformation (CCAM). The patient underwent right lower lobectomy and histopathological correlation confirmed the diagnosis of CCAM type 2 with surrounding acute and chronic inflammation.

2. Discussion

Five main types of CCAM, a congenital hamartomatous lesion of the lung, have been described based on the embryological level of origin and histological features.

2.1. Type 0

Type 0 CCAM is the rarest form, where it arises from the trachea or bronchus. The presentation is severe and usually lethal. Cysts are small [1].

2.2. Type 1

Type 1 CCAM is the most common form comprising 50%–70% of cases, where it arises from the distal bronchus or proximal bronchiole. Usually, a small number of large echolucent cysts are present, which measure 3–10 cm. A single dominant cyst may also be seen. The cyst walls are thin and lined with a ciliated pseudostratified epithelium, although other cell types such as cartilage may be found between the cysts. These CCAMs may be large, so they could have a significant mass effect, which can lead to hydrops [1].

2.3. Type 2

Type 2 CCAMs comprise 15%–30% of cases, where they arise from terminal bronchioles. They comprise small cysts measuring 0.5–2 cm as well as solid areas, which may be difficult to distinguish from the surrounding tissue. These CCAMs are lined with a ciliated cuboidal or columnar epithelium, and elements of bronchioles or alveoli may be seen. Frequently, the cysts are more evenly spaced than those in Type 1 CCAMs. Type 2 CCAMs have the highest

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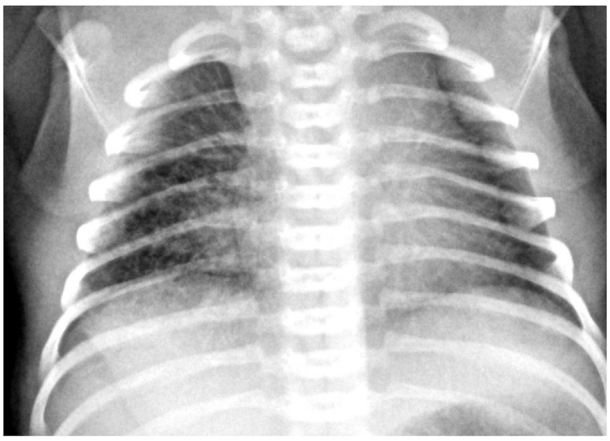

Fig. 1. Chest radiograph obtained on day 0 indicating no obvious cystic changes.

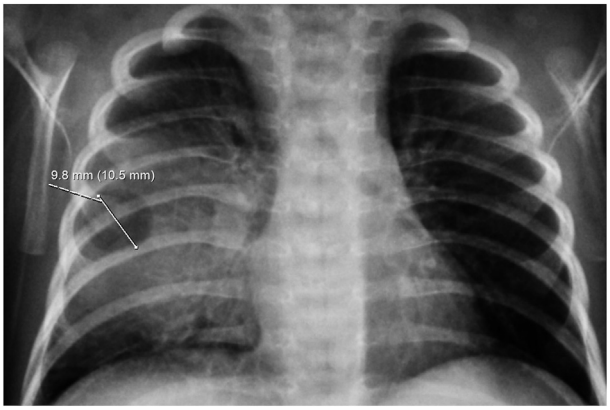

Fig. 2. Development of cystic changes in a background of air space opacity in the right middle lung zone.

incidence of associated anomalies at up to 60%, and the prognosis depends on these findings, which can include most organ systems [1].

2.4. Type 3

Type 3 CCAMs comprise 5%–10% of cases and they are thought to arise from acinar-like tissue. Type 3 CCAMs comprise cysts in macrocystic lesions and they are associated with a poorer prognosis. Types 1, 2, and 4 CCAMs are classified as macrocystic or both macrocystic and microcystic. Type 3 CCAMs are microcystic [1].

2.5. Type 4

Type 4 CCAMs comprise 5%–15% of cases. These CCAMs contain

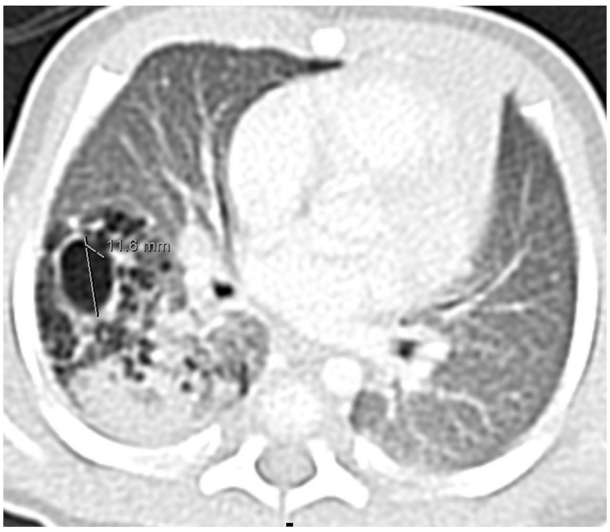

Fig. 3. CT scan of the chest showing multiple variable sized cysts in the right lower lobe, where the largest measured 1.16 cm.

cysts as large as 10 cm and they are associated with malignancy, specifically pleuropulmonary blastoma. They are alveolar in origin. Antenatally, CCAMs have been classified as microcystic (<5 mm) versus macrocystic (≥5 mm). Microcystic lesions are frequently significantly larger than teratomas and they tend to be more vascular, which may create more ultrasound shadowing [1].

3. Radiological findings

The imaging findings are variable and correlated with the type of the lesion imaged.

CT is a useful tool for differentiating cystic adenomatoid malformation from other entities that affect neonates in respiratory distress. CT is valuable for further characterizing the lesion in terms of its distribution and the internal structure of the affected lung [2].

4. Conclusion

Understanding the variable radiological appearance of this lesion may help to improve early detection to facilitate early diagnosis and optimal patient management.

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