Reminder of important clinical lesson

Recurrent febrile respiratory distress in infant: which diagnosis?

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Congenital cystic adenomatoid malformation (CCAM) of the lung is a rare congenital anomaly. CCAM diagnosis can be made by prenatal ultrasound or in postnatal cases with regard to respiratory distress or recurrent respiratory infections. We present, for diagnostic purposes, a case of a patient, aged 4 months who had suffered since birth from recurrent bronchopulmonary infections. He was hospitalised several times for febrile respiratory distress. Chest x-rays previously performed were interpreted as normal. A final radiograph indicated the diagnosis and it was confirmed by CT. Surgical resection was performed with a positive result. CCAM responds to the prenatal diagnosis and requires frequent ultrasound checks during pregnancy and allowing for optimal management. The radiological assessment leads to the diagnosis and addresses the treatment which is based on a surgical cure.

BACKGROUND

A case report of unknown congenital cystic adenomatoid malformation (CCAM) until 4 months old, highlighting the importance of antenatal diagnosis and chest radiograph interpretation.

CASE PRESENTATION

A 4-month-old male infant presented to a tertiary children's hospital with acute onset respiratory distress. He had been born at term following an uncomplicated vaginal delivery. In the newborn period, he presented with moderate respiratory distress; he was discharged after 48 h of continuous positive airway pressure and coverage by two antibiotics. They were arrested after improvement of the newborn and the negative infectious samples. In the following 4 months he was hospitalised five times for respiratory illnesses. In all cases, a presumptive diagnosis of viral-induced lower respiratory tract infection was made. Chest radiographs on these admissions had been reported as normal.

Family history was unremarkable except for a history of childhood wheezing in the father.

Examination revealed a well-grown thriving child with normal growth parameters. Temperature was 39°C. Respiratory rate was 75 bpm, heart rate 142 bpm, saturations were 97% in room air. Cardiovascular examination was unremarkable and respiratory examination revealed moderated intercostal and sternal recessions with a few scattered rhonchi and decreased breath sounds at the right base.

INVESTIGATIONS

On the chest radiograph (figure 1), interpreted by an expert, there are two suspicious bulbous images of upper and lower lobes of the right lung, indicative of cystic formations. The heart is normal.

Exploration of these cystic formations requires the completion of a chest CT (figures 2 and 3).

Chest CT shows at the right upper lobe, a multicystic air formation, oval, thin-walled, measuring 40×38×28 mm (figure 2). At the right lower lobe, there is a unilocular cyst; thin-walled, measuring 31×28×25 mm. No aberrant systemic artery is found. The mediastinum is deviated to the left.

DIFFERENTIAL DIAGNOSIS

On the chest radiograph, the bulbous image suggests a cystic formation may be related to a bronchogenic cyst, CCAM of the lung, pulmonary sequestration or congenital emphysema.

However, the view on CT suggests CCAM.

Figure 1 Chest radiograph showing suspicious bulbous images at the right lung.

Figure 2 Chest CT showing at the right upper lobe of the lung, a multicystic air formation.

TREATMENT

The treatment of this pulmonary malformation requires a surgical cure. Before this, we treated the infection by amoxicillin-clavulanic acid and we completed the imaging tests searching for other malformations. The abdominal ultrasound and echocardiogram were normal. Surgery, by thoracotomy, was decided at 6 months. Following the exploration, it was found that the upper lobe was normal and there was a cystic formation of the lower lobe. The resection of this cyst was done without incident.

OUTCOME AND FOLLOW-UP

The postoperative course was uneventful. Histopathological examination confirmed the diagnosis of CCAM. After 2 months, the infant remains well. His respiratory function is good.

DISCUSSION

CCAM is a rare congenital lung disease with an incidence of 1/25 000–1/35 000 births.¹ It represents 25% of

Figure 3 Chest CT scan showing at the right lower lobe of the lung, a cyst filled with air.

bronchopulmonary malformations.^{1 2} It is often localised to one lobe^{1–4} especially the lower lobes.¹ Stocker's classification, the most widely used,² defines several types of CCAM based on pathological characteristics.^{4 5} It is characterised by an abnormal growth of the terminal bronchioles without bronchial gland and alveolar differentiation. This causes the formation of varying sized cysts lined with tall columnar mucous epithelium. It can be associated with other congenital abnormalities such as pulmonary sequestration⁴ or cardiac anomalies, digestive defect, kidney and brain malformation.^{2 4}

Diagnosis is currently highlighted by antenatal ultrasonography^{1 3 5} or by MRI in doubtful cases.^{5 6} The majority of these malformations diagnosed at this stage have no impact on lung development and does not require prenatal intervention.³ In 30% of cases they may regress or even disappear completely.⁵ Sometimes CCAM can grow, causing mediastinal compression, polyhydramnios, fetal hydrops, pulmonary hypoplasia² and heart failure.^{3 5}

CCAM which escapes prenatal diagnosis occurs after birth in the course of respiratory distress, recurrent infections or pneumothorax. Respiratory disorders increase while crying and breastfeeding.²

The clinical diagnosis, strongly indicated by imaging, is confirmed by histopathological examination.⁴ Radiologically, CCAM appears as a cystic formation or multicystic thin-walled anomalies with thickenings or levels of air-fluid.⁴ However, a normal chest radiograph at birth does not eliminate the diagnosis.⁷ CT that can be achieved at 30 days of life⁷ or 45 days,⁵ portrays the anomalies accurately, eliminates differential diagnosis and addresses treatment.² The scan confirms the presence of cysts, wall thickness, the presence of air-fluid levels and possibly infection or associated ventilatory disorders.

The treatment of these malformations is surgical. It can be achieved at the antenatal stage if symptoms of mediastinal compression are present.³ This treatment is difficult to perform and is only done in specialised centres⁵ with a significant perinatal morbidity and death rate.³ In postnatal cases, the surgery is advisable on account of the risk of development of secondary complications (recurrent infections, respiratory distress, pneumothorax) and malignant transformation.^{1 3 6} Surgical resection is the main treatment proposed. It is a safe method for a well-trained team⁶ and usually consists of a lobectomy or segmentectomy.^{1 6}

Learning points

- ▶ Recurrent respiratory infection and respiratory distress are frequent modes of discovery of congenital cystic adenomatoid malformation.
- ▶ Importance of valid interpretation of chest radiograph.
- ▶ Value of chest CT in exploration of recurrent respiratory infections in children.
- ▶ Importance of prenatal diagnosis.

Competing interests None.

Patient consent Obtained.

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