**CASE FOR DIAGNOSIS**

**A boy with chest deformity**

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**Case report**

An 11 year old boy with a right hemithorax deformity and diminished breathing sounds in the right lung was referred to our department, following examination by a physician due to knee pain caused by flat-footedness. He had a history of recurrent viral infections of the upper airways in early childhood since the age of 3 yrs. He has been free of pulmonary complaints. Bronchitis was diagnosed only once. Pregnancy and delivery were normal. In the neonatal and infantile period the boy was in good health. The right hemithorax deformity was diagnosed at 4 yrs of age, but was interpreted as a complication of rachitis.

On physical examination, the patient appeared well. He was thin (weight 24 kg, height 134 cm; 25th and below 3rd percentiles, respectively), had a markedly enlarged right hemithorax with a slight reduction of wall movement, respiratory rate 16 breaths·min⁻¹, blood pressure 110/70 mmHg, and pulse rate 80 beats·min⁻¹. Heart sounds were normal. Lung auscultation revealed weakness of breathing sounds over the right lung field. Intensification to percussion was noted over the lower two thirds of the right hemithorax. Clubbing was not observed. In laboratory evaluation and blood gas analysis, no abnormality was found. An electrocardiogram revealed incomplete right bundle branch block. Pulmonary function tests showed forced expiratory volume in one second (FEV₁)=74% of predicted value and vital capacity (VC)=77% pred; all other parameters (including residual volume (RV) 132% pred, functional residual capacity (FRC) 106% pred, total lung capacity (TLC) 91% pred, and flow-volume curve) were normal. The patient’s exercise activity was normal.

A chest radiograph and computed tomography (CT) scan, are shown in figures 1 and 2.

Fibrebronchoscopy revealed a small diverticulum (depth 2 mm) at the site of the right ninth segmental bronchus. Bronchial culture was negative. The right bronchographs are shown in figure 3a and b.
BEFORE TURNING THE PAGE: INTERPRET THE CHEST RADIOGRAPH, CT SCAN AND BRONCHOGRAPHS. SUGGEST A DIAGNOSIS AND PROPOSE A TREATMENT.
The chest radiograph (fig. 1) shows: the overall multicystic lesion with three air-fluid levels in the expanded, hyperlucent right hemithorax; mediastinal shift and pulmonary herniation to the left; and inferior displacement of the hemidiaphragm.

**Interpretation of the computed tomography**

The CT section through the base of the lungs (fig. 2) shows multiple pulmonary cysts of various sizes in the right lower lobe, with air-fluid levels in some of them.

**Interpretation of the bronchographs**

Right bronchography (fig. 3) shows morphologically normal bronchi but compressed and displaced by the expanded part of the lower lobe. A small diverticulum was visible in the proper site of the right ninth segmental bronchus (fig. 3b).

**Diagnostic considerations**

In the differential diagnosis of the present case, bronchogenic cyst, pneumatocele, lobar emphysema and lobar pulmonary interstitial emphysema were considered. A diagnosis of congenital lung cystic adenomatoid malformation was initially made.

**DIAGNOSIS: "congenital lung cystic adenomatoid malformation"**

**Treatment**

The right lower lobe, with increased volume (9x8x6 cm) that occupied two thirds of the right hemithorax, was resected. It contained a small area of normal lung parenchyma and multiple cysts of various sizes. Some of them were partially filled with thick mucus.

Histological examination revealed multiple cysts lined by ciliated pseudostratified columnar epithelium. Their walls, with the absence of inflammation, contained smooth muscles. Normal alveoli were seen between the cysts. Histological evaluation was compatible with type I congenital cystic adenomatoid malformation (CCAM).

The postoperative course was complicated by a persistent air-leak, which was resolved with repeated drainage.

**Discussion**

CCAM is an uncommon congenital malformation. Approximately 200 cases of CCAM have been described so far [1, 2]. Most cases are found in infants younger than 1 year, though the lesions have been seen in children up to 14 yrs of age [3]. Cloutier et al. [1] concluded that only 17% of cases have been reported in children more than 1 year of age. Older children with CCAM most often present clinical symptoms of recurrent chest infection [3, 4]. Although CCAM are of three different pathological types [2], the only one usually compatible with survival over the neonatal period is type 1. A few asymptomatic children younger than 6 months of age were diagnosed incidentally due to abnormal chest radiographs performed for other reasons.

Chest radiography is always abnormal [1, 2, 4] and provides the main diagnostic clue. The type 1 variant of CCAM has a typical radiographic, multicystic pattern consisting of many cystic spaces containing air [2]. Fluid may be present within cysts, and on upright radiography an air-fluid level is visible.

Air diffusing from well-ventilated lung tissue through the alveolar Kohn’s pores is the cause of air-trapping in the cysts. The air pressure in the lesion is sufficient to cause distention of the occupied segment, which compresses other parts of the lung. The best treatment for CCAM is a surgical excision, offering good results by decompressing the thoracic viscera and allowing a proper expansion of the remaining pulmonary parenchyma.

Our patient was unusual because he presented deformity of the chest and no history of pulmonary disease. As in the case presented by Cloutier et al. [1], our child had low weight of unknown aetiology. The probable cause could be a chronic, subclinical impairment of ventilation due to compression and displacement of the normal pulmonary tissue. Radiographic analysis of the typical multicystic pattern suggested the diagnosis preoperatively. The CT scan of the chest clearly showed the morphology of the lesion, demonstrated the lack of any associated vascular abnormalities and confirmed unilateral disease. Bronchography localized the lesion more precisely and showed bronchial atresia. The gross appearance of the resected lobe corresponded closely to the radiographic image. Histological evaluation confirmed the initial diagnosis.

Congenital cystic adenomatoid malformation is a developmental abnormality resulting from an embryological insult up to 49 days [2]. In the case presented, co-existence of congenital cystic adenomatoid malformation and segmental bronchus atresia made it possible to define this more accurately and suggest that the insult took place between days 34 and 36. Bronchial atresia may be a primary defect leading to the development of congenital cystic adenomatoid malformation.

**Keywords:** Chest deformity, congenital cystic adenomatoid malformation.

**References**