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Intradiaphragmatic hybrid lesion in an infant: case report

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Abstract Hybrid lesions have elements of both congenital cystic adenomatoid malformation and bronchopulmonary sequestration. We report an unusual case of an infant treated for an intradiaphragmatic hybrid lesion. Although computed tomography remains the criterion standard imaging examination for planning the operation, the exact localization of such lesions may be discovered only at surgical exploration.

Congenital cystic adenomatoid malformation (CCAM) and bronchopulmonary sequestration (BPS) belong to the spectrum of congenital pulmonary malformations. *Bronchopulmonary sequestration* is defined as a mass of nonfunctioning lung tissue that receives its vascular supply from a systemic artery and is separated from the tracheobronchial tree. Several cases of supradiaphragmatic or infradiaphragmatic extralobar sequestrations (ELSs) have been reported in literature, whereas intradiaphragmatic ELS is an extremely rare condition [1].

Congenital cystic adenomatoid malformation is considered a hamartomatous lesion of the bronchial tree; 3 different types had been identified based on histologic characteristics [2]. In contrast to ELS, CCAM is connected to the tracheobronchial tree, and its vascular supply comes from the pulmonary circulation.

Malformations with both of these components have been recognized and defined as hybrid lesions [3].

We report a case of an infant with an intradiaphragmatic hybrid lesion. To the best of our knowledge, such localization has not been previously reported for hybrid lesions.

1. Case report

A 25-year-old prima gravida was referred to our hospital after an ultrasound (US) examination at 24 weeks' gestation, which showed a cystic lesion of the left lower lung. A 3.660-g female infant was delivered vaginally at term, with normal physical findings. At birth, the patient was initially investigated by a plain chest radiograph, which showed neither cystic lesions nor displacement of the mediastinum. During the first months of life, the patient remained asymptomatic; follow-up chest radiographs at 3 and 7 months were considered normal. Chest computed tomography (CT) performed at 8 months of age showed a mass in the left lower lobe, measuring 28 × 11 × 32 mm, with an extension at the posterior costophrenic angle. An aberrant arterial supply from the celiac axis was identified. At age 11 months, the patient underwent surgical exploration. After a muscle-

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sparing left thoracotomy, the lower lobe appeared normal. The posterior diaphragmatic surface was slightly irregular owing to presence of a cystic lesion, measuring 30 × 28 × 11 mm. The superior aspect of the diaphragm was incised, and the lesion was identified. By means of sharp and blunt dissection, the lesion was dissected free from the diaphragmatic muscle fibers. There was no large feeding vessel supplying the caudal aspect of the lesion; instead, many small arterial branches entered the lesion and were controlled by bipolar coagulation. After removal of the small lesion, the dead space created in the diaphragm was minimal, drainage was not considered necessary, and the superior aspect of the diaphragm was repaired.

Histopathologic examination demonstrated multiple air-filled cysts lined by ciliated respiratory-type epithelium, consistent with the diagnosis of type II CCAM (Fig. 1).

The postoperative course was uneventful; the chest drain was removed on day 3, and the patient was discharged on postoperative day 4.

At a follow-up at 6 months, the patient was completely asymptomatic.

2. Discussion

With widespread use and the development of routine obstetric sonographic screenings, prenatal recognition of congenital lung abnormalities is more and more frequent.

Congenital lung lesions represent a wide array of developmental abnormalities including CCAM, BPS, bronchogenic cysts, and congenital lobar emphysema. Congenital cystic adenomatoid malformation is the most common lesion, with an incidence of 1:25,000 to 1:35,000 live births [4], whereas BPS has an estimated incidence of 0.15% to 1.7% in the general population [5].

Based on clinical, gross, and microscopic criteria, CCAM was initially classified by Stocker et al [2] in 3 types: type I (macrocystic, >2 cm), type II (microcystic, <2 cm), and type III (solid with microscopic cysts). In 2001, the classification was expanded, with the addition of type 0 (previously reported as acinar dysplasia of the lung) and type 4 (lung cyst manifesting with pneumothorax) [6]. Because some of these lesions are not always cystic or adenomatoid, they are now

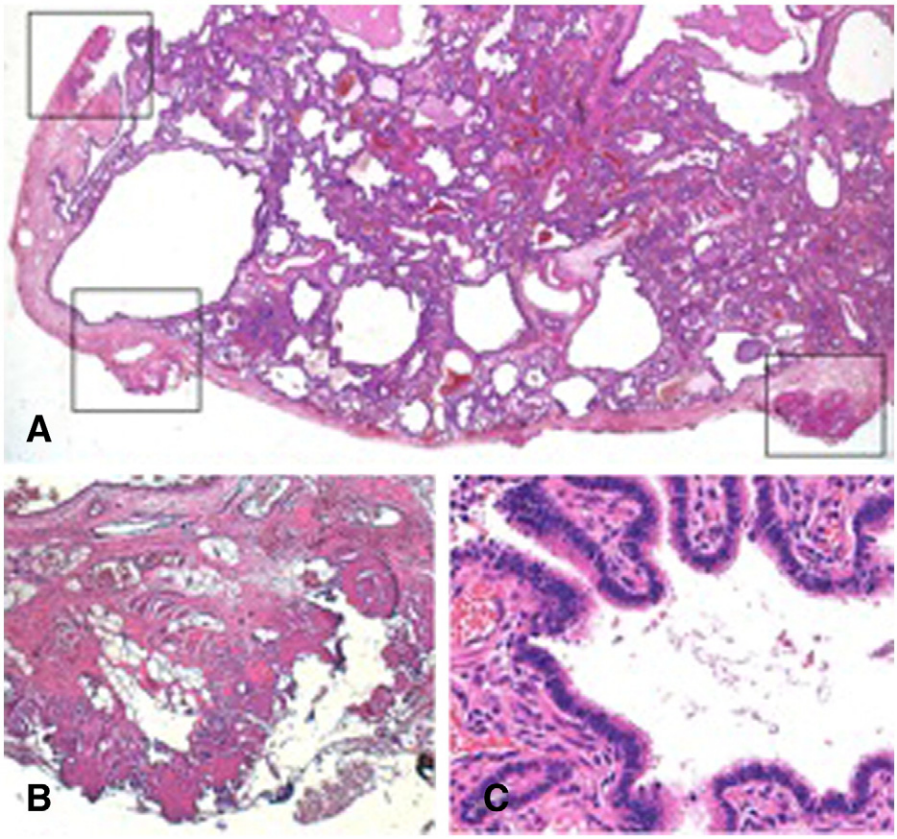

Fig. 1 Conventional histology of the specimen. A, Low-power micrograph of about half of the whole specimen, showing both cystic and relatively solid areas; the squares indicate the presence of diaphragm muscle tissue on both sides of the sample (H&E). B, High power of the lower left square of A, showing connective, fat, and striated muscle tissue of the diaphragm (H&E). C, High power of a cystic area detailing the normal bronchial type lining with ciliated cells (H&E).