

Clinical Image

Congenital Bi-segmental Bronchial Atresia with Areas of Congenital Pulmonary Airway Malformation



Atresia bronquial bisegmentaria congénita acompañada de áreas de malformación congénita de las vías respiratorias pulmonares

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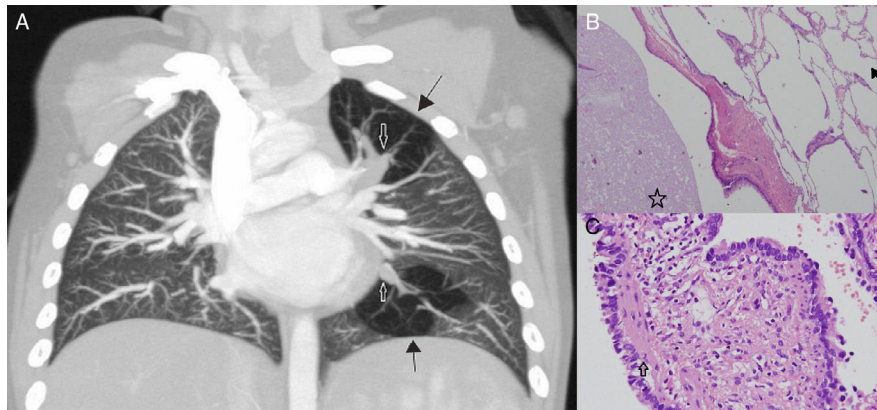


Fig. 1. (A) Contrast-enhanced, coronal plane, chest computed tomography. Closed arrows show mucocoeles and black arrows show air-trapped segments. (B) Haematoxylin-eosin, $\times 100$ magnification. The air trapping areas (open arrow) and mucocoele (asterisk), confirming bronchial atresia. (C) Haematoxylin-eosin, $\times 400$ magnification. CPAM areas (closed arrow) showed by respiratory epithelial lined cysts and a thin fibromuscular layer in the cyst wall.

A 2 year-old girl presented with a 6-month history of recurrent airway infections. On anteroposterior chest X-ray, focal radiolucent areas were seen in both the upper zone and paracardiac region of the left lung. On contrast-enhanced chest computed tomography, central mucocoele formations were observed in the left perihilar region, together with bi-segmental air trapping and oligoemia, suggestive of bi-segmental bronchial atresia (Fig. 1A). The patient underwent bi-segmentectomy. The histopathology report included mucocoele formations and emphysematous changes confirming bronchial atresia, together with respiratory epithelial lined cysts and a thin fibromuscular layer in the cyst wall, suggesting areas of focal congenital pulmonary airway malformation (CPAM) (Fig. 1B).

CPAM areas within bi-segmental bronchial atresia is an extremely interesting presentation.¹ To the best of our knowledge, this is the first such case reported in the literature. This case is an

example of type 4 CPAM that is generally part of the differential diagnosis of focal air-trapping. CPAM is associated with increased incidence of pulmonary malignancy in addition to pulmonary and extrapulmonary malformations. Pleuropulmonary blastoma, bronchioloalveolar carcinoma and rhabdomyosarcoma are the most common malignancies associated with CPAM.² The presence of CPAM areas within bi-segmental bronchial atresia requires long-term follow-up due to the increased risk of malignancy.

References

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