Arch Bronconeumol. 2017;53(9):517

Sociedad Española de Neumología y Cirugía Torácica SEPAR

ARCHIVOS DE Bronconeumología

www.archbronconeumol.org

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

Zuhal Bayramoglu[a,*], Emine Caliskan[a], Emine Dilek Yilmazbayhan[b]

[a] Department of Radiology, Istanbul Faculty of Medicine, Istanbul University, Fatih 34093, Istanbul, Turkey
[b] Department of Pathology, Istanbul Faculty of Medicine, Istanbul University, Fatih 34093, Istanbul, Turkey

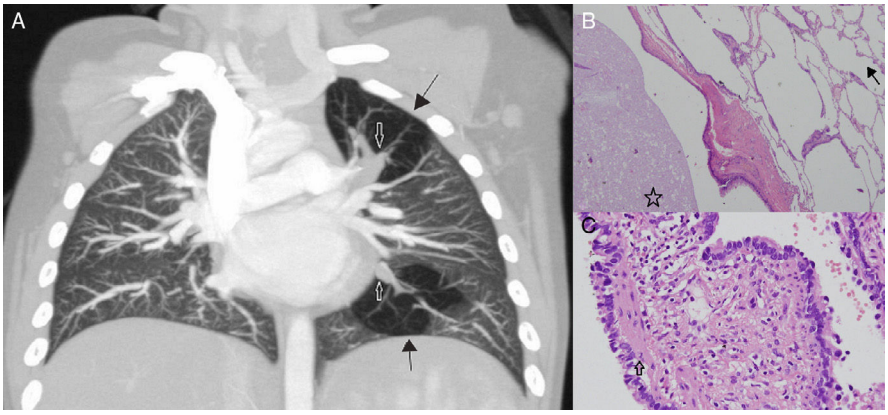

Fig. 1. (A) Contrast-enhanced, coronal plane, chest computed tomography. Closed arrows show mucoceles and black arrows show air-trapped segments. (B) Haematoxylin–eosin, ×100 magnification. The air trapping areas (open arrow) and mucocele (asterix), confirming bronchial atresia. (C) Haematoxylin–eosin, ×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 mucocele formations were observed in the left perihilar region, together with bi-segmental air trapping and oligaemia, suggestive of bi-segmental bronchial atresia (Fig. 1A). The patient underwent bi-segmentectomy. The histopathology report included mucocele 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

1. Garcia-Pena P, Guillerman RP, editors. Pediatric chest imaging. 3rd edition Berlin: Springer; 2014. p. 204–5.
2. David M, Lamas-Pinheiro R, Henriques-Coelho T. Prenatal and postnatal management of congenital pulmonary airway malformation. Neonatology. 2016;110:101–15.

* Corresponding author.
E-mail address: incezuhal@yahoo.com (Z. Bayramoglu).

http://dx.doi.org/10.1016/j.arbres.2017.02.010