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Case Report / Olgu Sunumu

Video-assisted thoracoscopic lingulectomy for type 1 congenital pulmonary airway malformation in an adult patient: a case report

Yetişkin bir hastada tip 1 doğuştan pulmoner hava yolu malformasyonu için video yardımlı torakoskopik lingulektomi: Olgu sunumu

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ABSTRACT

Congenital pulmonary airway malformation is a rare embryogenic developmental anomaly of the lower respiratory tract in adulthood. In this article, we report a 38-year-old male patient with congenital pulmonary airway malformation, admitted with an 8 cm-diameter air cyst in the upper lobe of left lung. Air cyst was resected by video-assisted thoracoscopic lingulectomy. The histopathological examination of the lingular segment revealed type 1 congenital pulmonary airway malformation. Surgical resection is both diagnostic and curative for congenital pulmonary airway malformation. In cystic lesions, congenital pulmonary airway malformation should be kept in mind and appropriate surgical intervention should be performed.

Keywords: Adult; congenital pulmonary airway malformation; lingulectomy; video-assisted thoracoscopic surgery.

ÖZ

Doğuştan pulmoner hava yolu malformasyonu erişkinlikte alt hava yolunun nadir bir embriyonel gelişimsel anomalisidir. Bu yazıda sol akciğerin üst lobunda 8 cm çapında hava kisti ile başvuran, doğuştan pulmoner hava yolu malformasyonlu 38 yaşında bir erkek hasta sunuldu. Video yardımlı torakoskopik lingulektomi ile hava kisti rezeke edildi. Lingular segmentin histopatolojik incelemesinde tip 1 doğuştan pulmoner hava yolu malformasyonu olduğu görüldü. Cerrahi rezeksiyon doğuştan pulmoner hava yolu malformasyonu için hem tanısal hem de küratiftir. Kistik lezyonlarda doğuştan pulmoner hava yolu malformasyonu akılda tutulmalı ve uygun cerrahi girişim uygulanmalıdır.

Anahtar sözcükler: Yetişkin; doğuştan pulmoner hava yolu malformasyonu; lingulektomi; video yardımlı torakoskopik cerrahi.

Congenital pulmonary airway malformation (CPAM), previously known as congenital cystic adenomatoid malformation, is a rare but the most common embryogenic developmental anomaly of the lower respiratory tract.[1] While majority of the patients present with respiratory distress in the newbornhood, some cases may remain asymptomatic until later in life. The incidence of congenital lung cysts is in the range of 1/8,300 to 1/35,000 live births.[2] Congenital pulmonary airway malformation is still an uncommon disorder for adult chest physicians and thoracic surgeons. In this article, we report an adult patient with CPAM who underwent video-assisted thoracoscopic lingulectomy.

CASE REPORT

A 38-year-old male patient admitted with an 8 cm-diameter air cyst in the left upper lobe that was discovered on chest computed tomography while he was being evaluated for chest pain etiology. The wall of the air cyst was thin enough not to suggest a cavitary lesion in the etiology. Patient's medical history was not significant for any pulmonary diseases. According to clinical and radiological findings, our pre-diagnosis was an air cyst in the lingular segment (Figure 1). We previously performed thoracotomy and closure of bronchial openings with capitonnage in two patients who had air cysts like presented case, but the surgical results were not satisfactory. Therefore, we

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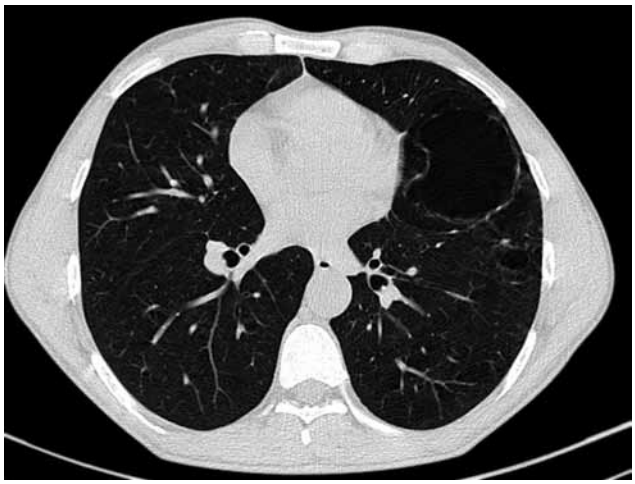

Figure 1. Chest computed tomography of patient revealed an air cyst located in lingular segment.

recommended video-assisted thoracoscopic resection of lingular segment including air cyst for this patient. We performed video-assisted thoracoscopic lingulectomy without any complication. The postoperative course was uneventful. Chest tube was removed on third postoperative day and he was discharged without any problem. The histopathologic examination of the lingular segment revealed a large cyst with thin wall. In most of the areas, cyst wall was lined with ciliated pseudostratified columnar epithelium including mucus secreting cells. In cyst wall beneath epithelium and in lung parenchyma very close to cystic structure, there were mucous gland proliferations (Figures 2 and 3). The patient was diagnosed with type 1 CPAM which has a good prognosis when compared with other types. Regarding that bronchoalveolar carcinoma has been reported in association with large CPAMs, we kept the patient under close follow-up.[2,3] We have not encountered any problems in the clinical follow-up of the patient yet. A written informed consent was obtained from the patient.

DISCUSSION

Congenital pulmonary airway malformations, which are hamartomatous lesions, result from abnormalities of branching morphogenesis of the lung. The type variability of CPAM is thought to depend on various developmental anomalies at different embryological states of the tracheobronchial tree and lung.[1,2] While majority of the patients present with respiratory distress, some may remain asymptomatic until later in life.[2] Type 1 is the most common form of CPAM (60 to 70% of all cases). Considering that there is well-differentiated tissue within these cysts, this type is thought to originate relatively late during embryogenesis (7 to 10 weeks) and originate from the distal bronchi or proximal bronchioles. Type 1 CPAM has malignant potential especially for bronchoalveolar carcinoma, but the magnitude of the risk is not well delineated. Our patient also had type 1 CPAM with an air cyst 8 cm in diameter at the lingular segment and was consistent with the criteria described for type 1.[1,2]

McDonough et al.[3] identified 42 adult patients of CPAM in the literature up until February 2012. We detected 27 more adult patients with CPAM reported in the literature review from February 2012 to November 2015. However, the real prevalence of CPAM in general adult population has still not

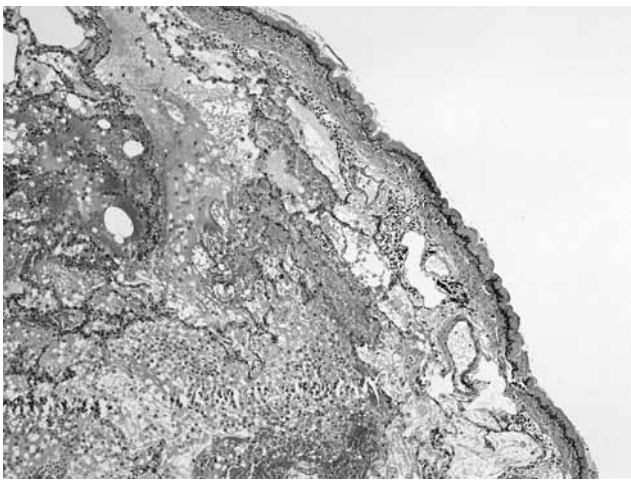

Figure 2. Cyst wall was lined with pseudostratified columnar epithelium including mucus secreting cells. Some epithelial cells were ciliated (H-E x 40).

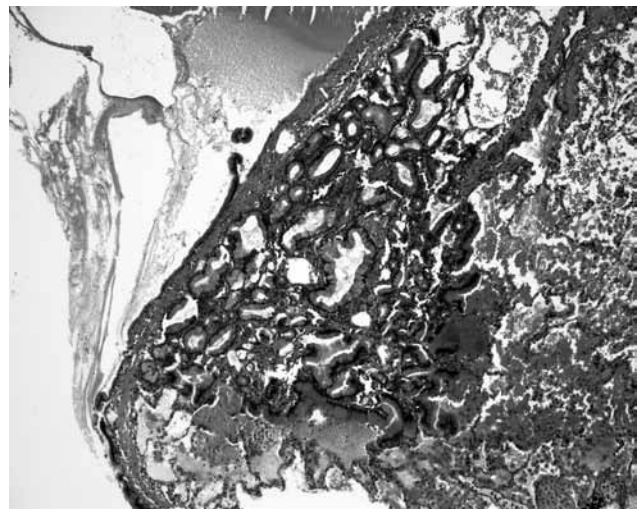

Figure 3. In some areas, beneath cyst epithelium in cyst wall, mucous gland proliferations were observed (H-E x 100).

been well-established. Due to its rarity, it is seldom suspected and adult physicians are not familiar with its clinical and radiological findings.

While CPAM may remain asymptomatic and sometimes be incidentally diagnosed, clinical presentation in adults is commonly characterized by recurrent pulmonary infections, pneumothorax, mycetoma, hemoptysis or bronchoalveolar carcinoma.[2,3] Among 42 adult patients identified by McDonough, 24% was asymptomatic with only radiologic abnormalities.[3] Our patient was also asymptomatic up to this age and admitted with chest pain.

The prognosis of CPAM presenting in adulthood depends on its type, pathological feature, and the potential for malignant transformation.[4] Although there is no precise data on treatment modalities and prognosis in adult CPAMs, most experts recommend surgical resection to confirm the diagnosis and reduce the risk of infection or malignant transformation.[3,4] Traditionally, lobectomy has been preferred because of the fear of incomplete removal of the pulmonary malformation and complications like air leak associated with lung sparing surgeries.[5,6] On the other hand, some authors suggest close observation of the lesions instead of prophylactic surgeries. They state that the risk of malignancy is overemphasized in these cases and prophylactic resection of CPAM lesions might not always be fully protective.[4,6] Considering the reports stating malignant transformation developing after prophylactic resection, close follow-up is absolutely necessary after resection of the lesion.[3,5,6] We performed diagnostic and prophylactic video-assisted thoracoscopic lingulectomy for our patient without any complication and kept him under close clinical follow-up. We performed surgical treatment previously for two patients who had air cyst and found many bronchial openings in the cystic lesions intraoperatively. Capitonnage used to be our surgical method, but unfortunately, the results were not satisfactory. Early surgical results were better and also to avoid lung cancer originating from this lesion, we recommend anatomic resection for patients with airway cystic lesions.

In conclusion, the current literature confirms that surgical resection is still the main treatment choice in congenital pulmonary airway malformation. Surgery is both diagnostic and curative in these patients. Resection of the lesion also reveals required specimen for histological examination and prevents infection and the potential neoplastic transformation. However, close clinical follow-up is necessary for these patients after resection of the lesion. In patients with cystic lesions, congenital pulmonary airway malformation should be kept in mind and appropriate surgical intervention should be performed.

Declaration of conflicting interests

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