Late presentation of cystic adenomatoid malformation type 4 of lung in an asymptomatic child

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ÖZET

Asemptomatik bir çocukta akciğerin geç prezentasyonlu tip 4 kistik adenomatoid malformasyonu

Akciğerin konjenital kistik adenomatoid malformasyonu genellikle perinatal dönemde tanı alan, nadir bir embriyonik gelişim anomalisidir. Nadiren prezentasyonu çocukluk çağı ve erişkin döneme gecikebilir. Burada tesadüfen akciğerin geç prezentasyonlu tip 4 konjenital kistik adenomatoid malformasyonu tanısı alan 10 yaşında bir kız çocuğu sunuyoruz.

Anahtar Kelimeler: *Konjenital kistik adenomatoid malformasyon tip 4, çocuk, akciğer.*

SUMMARY

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Congenital cystic adenomatoid malformation of lung is an uncommon embryonic developmental anomaly usually diagnosed perinatally. Rarely the presentation is delayed until childhood and adulthood. Herein we present a 10-years-old girl with a late presentation of congenital cystic adenomatoid malformation type 4 of lung who was diagnosed coincidentally.

Key Words: *Congenital cystic adenomatoid malformation type 4, child, lung.*

Congenital cystic adenomatoid malformation (CCAM) is a rare abnormality characterized by disorganized overgrowth of respiratory bronchioles and intercommunicating cysts (1). In the classification by Stocker of CCAM, five types were described, based on site of origin of the malformation and labeled 0 ± 4 to indicate their progression down the airway (2). At new classification the new name of CCAM is "congenital pulmonary airway malformation". Most of the cases are diagnosed in the neonatal period. A few cases have been diagnosed in childhood and adulthood (3,4). Herein, we report a late presentation of CCAM type 4 in an asymptomatic child.

CASE REPORT

A healthy ten years old girl was admitted to Pediatric Surgery Department of our hospital because of a giant cyst detected in the left lung. She neither got a history of lower respiratory tract problem nor she was taken a chest radiograph before. One day before the admittance, she fell down gently on his back and got a slight back pain. Because she got a slight fever and sore throat on the same day, she went to an outpatient clinic. The chest radiograph and the computerized tomography (CT) of thorax taken afterwards suggested congenital lobar emphysema. On admission she was well and in no respiratory distress. Pulmonary auscultation revealed decreased breath sounds over upper left lung. Laboratory studies included normal CBC and C-reactive protein levels. Chest radiograph showed a large hyperlucent area of the left lung. CT of thorax revealed a large thin walled cyst with dimensions of 10 x 8 x 6 cm covering upper lobe of the left lung (Figure 1). Thoracoscopic surgery was performed successfully by removing the cyst completely. The gross pathological examination revealed an unilocular cyst of a 9 cm maximum diameter torn in the surgery. Histologically the cystic lesion

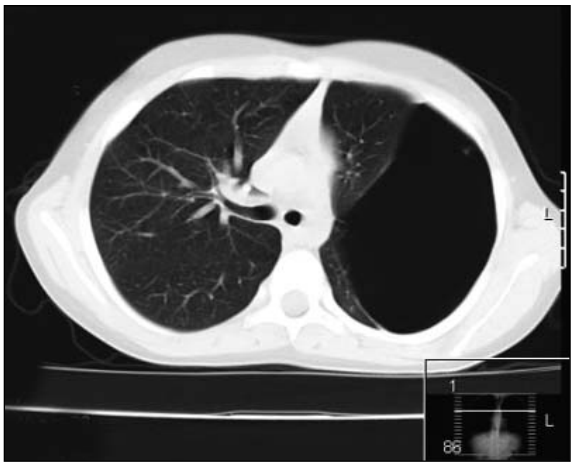

Figure 1. A large thin walled cyst in thorax CT of the patient.

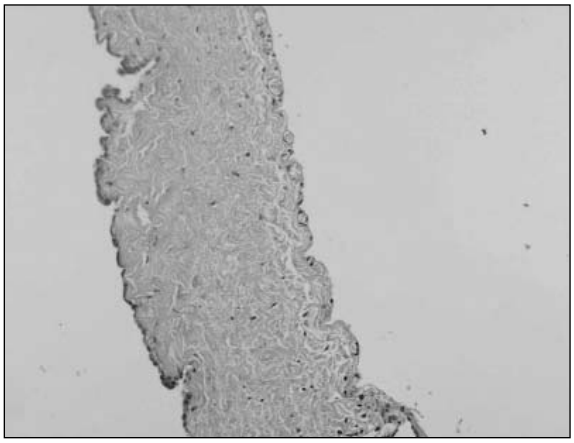

Figure 2. Histological findings of the patient.

looks like CCAM 4 with cysts that are composed of loose to dense collagen and contain somewhat unusual vessels. The lining of the cyst wall appears to be more like alveolar cells, which is a characteristic of the type 4 lesion (Figure 2).

DISCUSSION

A CCAM is characterized by a lack of normal alveoli and an excessive proliferation and cystic dilatation of terminal respiratory bronchioles (5). The reported incidence of CCAM is approximately 1 in 25.000 pregnancies (6). Type 1 accounts for 50% to 70% of cases. Type 4 is a rare form with a frequency of 2-4% in all CCAMs (5). They are characterized by large peripheral thin-walled cysts lined by type 1 pneumocytes and some more rounded cells representing type 2 pneumocytes. A low cuboidal-like epithelium may be present, but cilia are not present (2).

Cystic adenomatoid malformations are usually unilateral, affecting either lung equally (5). Eighty to 85% of all cases are diagnosed in the first 2 years of life (7). There are rare cases in which the lesion is first detected after infancy (4,8). The most common presentation is acute respiratory distress during the neonatal period (8). There are few described cases where CCAM have remained asymptomatic throughout life. The most common complication is pneumonia. Other complications include the development of malignancies, pneumothorax, hemoptysis or hemothorax (9). The importance of our case was being asymptomatic till the late presentation age.

Resection of the cystic lesion is advised in order to make the exact diagnosis of CCAM, to prevent recurrent infections, as well as the risk of developing malignancies in later life (10). In our case thoracoscopic surgery performed successfully by removing the cyst completely.

In summary, CCAM is a rare congenital malformation of the lower respiratory tract that may be diagnosed after infancy. It must be included in the differential diagnoses of any individual with lung cyst even if it is found as coincidentally on a chest radiograph

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