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 çocukluçağı ve erişkin döneme gecikebilir. Burada tesadüfen akciğerin geç prezentasyonu tip 4 konjenital kistik adenomatoid malformasyonu tanısal alan 10 yaşında bir kız çocuğunu sunuyoruz.

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

SUMMARY
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Anteriorly gadolinium-enhanced T1-weighted images show a large lobulated mass arising from the left lower lobe with a heterogeneous background suggestive of a complex cystic lesion. The mass demonstrates intense enhancement along its margins, typical of hemorrhagic components. The surrounding lung parenchyma appears normal. The lesion is well-circumscribed and has a thin rim of enhancing tissue, consistent with a peripheral location. There is no evidence of invasion into the surrounding structures or distant metastases. The findings are most consistent with a benign cystic lesion, such as a simple cyst or a mucous cyst. Further imaging and clinical correlation are necessary to make a definitive diagnosis.
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, hemothysis 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.

**REFERENCES**