

OLGU SUNUMU

AKCİĐERİN KONJENİTAL KİSTİK ADENOMATOİD MALFORMASYONU: Prenatal ve Postnatal Deđerlendirme

CONGENITAL CYSTIC ADENOMATOID MALFORMATION OF THE LUNG:
Prenatal and Postnatal Evaluation

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ÖZET

Akciđerin konjenital kistik adenomatoid malformasyonu (KKAM) ender görülen konjenital kistik bir akciđer lezyonudur.

Bu yazıda bir KKAM (Stocker tip 1) olgusu literatür bilgileri eşliğinde sunulmaktadır. Olgu fetal MR ile antenatal tanı almıştır ve ek konjenital anomali veya hidrops saptanmadığından intrauterin girişim yapılmamıştır. 36 gebelik haftasında doğan olgunun Apgar skoru 8 ve O₂ satürasyonu %92 olarak izlenmiştir. Solunum sıkıntısı ve takipne bulguları olan olgunun tanısı BT ile doğrulanmıştır. Olgu solunum sıkıntısı nedeniyle opere edilmiş ve lezyon tümüyle rezeke edilmiştir.

Anahtar Sözcükler: Kistik adenomatoid malformasyon, Konjenital kistik akciđer lezyonu, Postnatal tanı, Prenatal tanı

SUMMARY

Congenital cystic adenomatoid malformation (CCAM) of lung is a rare congenital cystic lung lesion..

We report a case of CCAM (Stocker type 1) with a brief review of literature. The patient was diagnosed antenatally with fetal MR. As no other associated anomalies or fetal hydrops were observed, no intrauterine intervention was planned. The baby was born at 36 weeks of gestation with an Apgar score of 8 and O₂ saturation was 92%. He had respiratory distress and tachypnea. The diagnosis was confirmed with CT examination. The patient was operated due to respiratory distress and the lesion was totally resected.

Key Words: Congenital cystic lung lesion, Cystic adenomatoid malformation, Postnatal diagnosis, Prenatal diagnosis

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INTRODUCTION

Congenital cystic adenomatoid malformation (CCAM) of lung is a rare congenital cystic lung lesion that results from excessive proliferation of tubular bronchial structures (1-3). The reported incidence ranges from 1 in 11,000 to 1 in 35,000 pregnancies and it represents 25% of congenital lung malformations and 95% of congenital lung lesions (1-3). Almost eighty percent of the lesions are recognized in neonatal period; however, there are reports even in adult population (1-3).

Five types of CCAMs are described based on cyst size, number and pathology which differ based on the embryologic level of origin and the histologic features (4).

We report a case of CCAM (Stocker type 1) which was diagnosed antenatally with fetal MR. The diagnosis was confirmed with CT examination after the birth and the patient was operated consequently.

CASE REPORT

A 33-year-old patient with 32 weeks of gestation (G3 P2) was referred to our hospital for fetal MR examination due to suspicion of a lung mass in fetal ultrasound performed in another clinic. The patient was not on routine follow up and this was the first ultrasound examination after the first trimester.

She had two healthy children of nonconsanguineous marriage with a history of two uneventful pregnancy. Her past medical history and family history were unremarkable.

The fetal MR was performed with a 1.5 Tesla scanner (Avanto, Siemens AG Medical Solutions, Erlangen, Germany) with high-performance gradients (maximum amplitude, 40 mT/m, slew rate, 200 mT/m/sec) with a multichannel phased-array surface radiofrequency coil.

The MR imaging protocol consisted of single shot HASTE GRE-T2W sequence on coronal, axial and sagittal planes. The MR images revealed multiple cystic lesions in the right lung with a maximum diameter of 30mm in the apico-posterior segment (Figure 1). The mediastinal structures were shifted to the left side and the left lung was seemed to be hypoplastic. We diagnosed the case as CCAM. No other associated anomalies or fetal hydrops was observed and the fetal development was correlated with the gestational age. Therefore, no intrauterine intervention was planned. The patient delivered a male baby with cesarian section at 36 weeks of gestation.

The baby was 2500 gr at birth. The Apgar score was 8. He had respiratory distress and tachypnea. Pectus excavatus deformity was present. Laboratory findings were Hb: 17.8 gr/dl, Htc: 54%, trombocyte: $311.000/mm^3$, leucocyte: $15.900/mm^3$, CRP ve blood biochemistry were normal. O₂ saturation was 92%. The chest X-ray obtained after birth showed cystic lesions in the right lung with a mediastinal shift (Figure 2). Physical examination findings were otherwise normal.



Figure 1. The fetal magnetic resonance (MR) image (coronal plane) revealed a multicystic lesion in the right lung with a maximum diameter of 3cm in the apex..

To confirm the antenatal diagnosis and to accurately localize the site of lesion within the lung segments for surgical planning, a thorax CT examination was performed the next day with a 64-slice scanner (Sensation 64, Siemens. AG Medical Solutions, Erlangen, Germany) with low dose protocol. In CT examination, almost all of the lower lobe of right lung was filled with cystic lesions with no visible normal lung parenchyma (Figure 3). The biggest cyst was 3cm in diameter and there was air-fluid levels in some of the cysts located in the basal segments. The aeration of the medial segment of middle lobe and apical segment of the lower lobe was partly preserved. The left lung was hypoplastic but bronchial and segmental distribution was

normal. The heart and mediastinum was shifted to the left. With these findings CCAM (Stocker type 1) was reported.

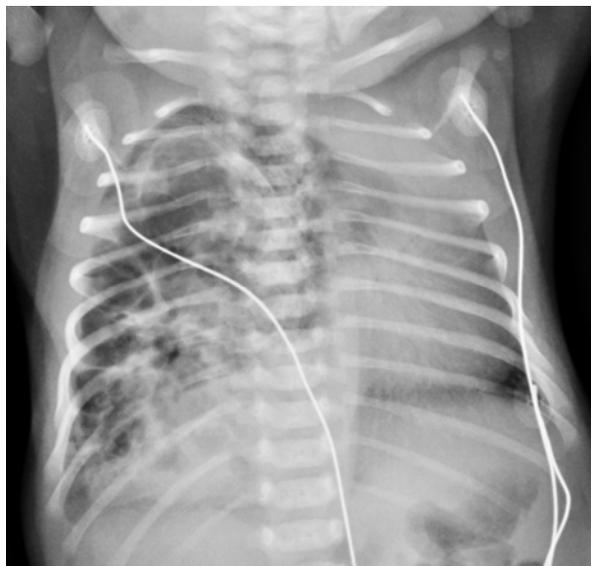


Figure 2. The chest X-ray obtained after birth showed multiple cystic lesions in the right lung and mediastinal shift .



Figure 3. The axial image of thoracic computed tomography (CT) examination demonstrated multiple cystic lesions in the right lung. The heart and mediastinum was shifted to the left.

As the respiratory distress continued on follow up, an urgent surgery was planned. The baby was operated on the fifth day. In the surgery, the middle and lower lobes of the right lung were totally resected. After the operation, no major complication was observed related to the operation. Our diagnosis was also confirmed with pathological correlation.

DISCUSSION

CCAM is a rare developmental, non-hereditary, hamartomatous abnormality of lung with unknown etiology presenting as cystic or solid lung masses (1,2). Most CCAMs present with respiratory distress or compromise during infancy or recurrent pneumonias in later years, including adulthood (2). The pathological feature of CCAM is adenomatoid proliferation of the bronchioles forming cysts (1-4).

It was classified into 3 subtypes in 1977 and expanded into five types and renamed as congenital pulmonary airway malformation by Stocker in 2002 (4).

Type 0 CCAM is the rarest form and arises from the trachea or bronchus. The presentation is severe and usually lethal. Cysts are small (<0.5 cm).

Type 1 CCAM is the most common form, representing 50% to 70% of cases, and it arises from the distal bronchus or proximal bronchiole. There are usually a small number of large echolucent cysts, measuring 3 to 10 cm. A single dominant cyst may also be seen. Cyst walls are thin and are lined by ciliated pseudostratified epithelium. Because these CCAMs may be large, they may have significant mass effect, which can lead to hydrops.

Type 2 CCAMs account for 15% to 30% of cases and arise from terminal bronchioles. They are composed of smaller cysts, measuring 0.5 to 2 cm, as well as solid areas that may be difficult to distinguish from surrounding tissue. These are lined by ciliated cuboidal or columnar epithelium, and elements of bronchioles or alveoli may be seen. Type 2 CCAMs have the highest incidence of associated anomalies, up to 60%, and prognosis depends on these findings. Associated anomalies are most commonly cardiac and renal and they include, cardiac anomalies (ventriculoseptal defects, tetralogy of Fallot, and truncus arteriosus), renal agenesis/dysgenesis, gastrointestinal defects and skeletal anomalies.

Type 3 CCAMs account for 5% to 10% of cases and are thought to arise from acinar-like tissue. Type 3 CCAMs are composed of cysts that are so small the mass appears to be solid and highly echogenic on ultrasound. The tissue is acinar and shows adenomatoid elements consistent with distal airway. These masses may be large and may distort the thoracic contents; prognosis depends on the extent to which they do so.

Type 4 CCAMs account for 5% to 15% of cases. These CCAMs contain large cysts that may be as large

as 10 cm and have been associated with malignancy, specifically pleuropulmonary blastoma. They are alveolar in origin.

Prognosis also depends on Stocker type (1,5,6). Type I lesions have good prognosis. In type II lesions, the presence and extent of associated anomalies determine the prognosis. Type III lesions carry bad prognosis as they are usually large and presents with cardiovascular compromise (1-4).

Depending on the size of the lesion, other possible findings include polyhydramnios, mediastinal shift, pleural effusions, and hydrops. Large lesions may compress residual tissue, thus increasing the risk of pulmonary hypoplasia, which cannot be predicted by antenatal imaging (1,2,5). In general, associated congenital anomalies, bilateral involvement, presence of hydrops and other signs of cardiovascular compromise result in poor prognosis (2,5).

CCAMs are frequently detected on routine antenatal ultrasound (1,2) However, with similar appearances of different congenital lung and nonpulmonary lesions, a definitive diagnosis cannot usually be established antenatally with absolute certainty (1,2,5,6). Particularly in those cases of indistinguishable sonographic findings, fetal MRI is the modality of choice for proving the diagnosis and preliminary appraisal of intensive care therapy and extracorporeal membrane oxygenation postnatally. Furthermore, fetal MRI often facilitates assessment and planning of intrauterine surgical procedures (7).

Indications for prenatal intervention includes a large cyst more than 2 cm, the presence of mediastinal shifting and fetal hydrops. Intrauterine treatment options are cyst aspiration and thoracoamniotic shunting. Shunting is usually offered in more severe cases. If the cyst fluid reaccumulates rapidly after aspiration, fetal thoracoamniotic shunting should be considered. It may also be considered for non-hydropsic fetuses if the predicted risk of developing hydrops is high. Specifically, this includes fetuses with very large lung lesions that increase in size rapidly or are associated with polyhydramnios (5).

Serial ultrasound monitoring of congenital cystic lung lesions has demonstrated that some of these lesions decrease in size and may regress spontaneously; However, a significant proportion persist on postnatal imaging (5). Therefore, all prenatally detected patients

require thorough postnatal evaluation, including a chest computed tomographic scan (5,7).

The most common presentation of CCAM is respiratory distress immediately following birth (1,2,5,7). In the case of respiratory compromise, surgery is the accepted standard of care. Patients with large lesions may require emergency or urgent surgery in the neonatal period (5).

Some children are asymptomatic at birth, but present with recurrent respiratory infections later in childhood (1,2,5). Because of the risk of infection and of malignant transformation, most authors recommend resection of all antenatally diagnosed CCAMs, although often the surgery can be deferred until several months after birth (5). All removed tissue should be examined histologically. In stable patients, the timing of elective surgery is controversial(5). Some surgeons prefer a "wait and see" approach operating only on those patients who develop symptoms, but others operate on asymptomatic patients usually within the first year of life (5) Due to the potential of malignant transformation, children should have long term follow up (1,5).

Our patient was diagnosed as CCAM type 1. No hydrops was observed antenatally and no associated abnormality was detected. However, the patient presented with respiratory distress in the neonatal period and an urgent operation was performed.

CONCLUSION

CCAM is a rare developmental malformation of lung. With the increasing use of antenatal ultrasound, an increasing number cases are diagnosed antenatally. The prognosis of these lesions are variable and indications for prenatal and postnatal interventions must be determined according to the clinical presentations. Due to the potential of malignant transformation, even asymptomatic children should have long term follow up.

KAYNAKLAR

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