

# A Case Report of Coexistence of Congenital Cystic Adenomatoid Malformation, Situs Inversus and Pectus Excavatum

## Konjenital Kistik Adenomatöz Malformasyon, Situs İnvversus ve Pektus Ekskavatum Birlikteliği Olgusu

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**ABSTRACT** As a rare variant of cystic congenital pulmonary disorders, congenital cystic adenomatoid malformation is characterized by marked proliferation of terminal respiratory structures and associated with various congenital anomalies such as pectus excavatum, cardiac and pulmonary lesions. We present a case that was treated surgically for congenital cystic adenomatoid malformation involving in the left middle lobe and pectus excavatum and situs inversus in a 3-year-old girl suffering recurrent infections and failure to thrive.

**Key Words:** Situs inversus; cystic adenomatoid malformation of lung, congenital; congenital abnormalities

**ÖZET** Konjenital kistik akciğer hastalığının bir türü olan konjenital kistik adenomatoid malformasyon (KKAM) dilate olmuş bronşiyol benzeri yapıların değişen boyuttaki proliferasyonu ile karakterize bir hastalıktır. Beraberinde pektus ekskavatum, kardiyak ve pulmoner lezyonlar bulunabilir. Bu yazıda konjenital kistik adenomatoid malformasyonun olduğu sol orta lobu cerrahi olarak tedavi ettiğimiz dekstrocardi, ve pektus ekskavatuumlu ve sık enfeksiyonlar ile büyüme zorluğu olan 3 yaşındaki kızda KKAM un klinik görünümünü sunuyoruz.

**Anahtar Kelimeler:** Situs inversus; akciğerin kistik adenomatoid malformasyonu, doğumsal; doğumsal anomaliler

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**A**s a rare pulmonary malformation, congenital cystic adenomatoid malformation (CCAM) is a dysplasia of overgrowing bronchioles and results from abnormal pulmonary mesenchymal development. CCAM is also considered a hamartomatous lesion of the lung with presence of skeletal muscular elements. Congenital anomalies such as intestinal atresias, diaphragmatic hernia, renal agenesis, pectus deformities and cardiac anomalies are associated with CCAM and cause the outcome poorer.<sup>1-3</sup> Affected children may be completely asymptomatic or present with severe respiratory distress in the newborn period. Most CCAMs present with respiratory distress and recurrent pneumonias in the infants.

### CASE REPORT

A three year old girl presented cough, sputum, frequent bronchopulmonary infections, tachypnoea and sometimes cyanosis. She was found to have dex-

trocardia and pectus excavatum on examination. The auscultation did not reveal any pathological sounds or murmurs. Her imaging tests revealed situs inversus in the thorax and multicystic lesion in the left lung (Figure 1, 2 and 3). The exact diagnosis of the reversal of lungs was not made until surgical exploration was performed. She underwent left middle lobectomy via left muscle-sparing thoracotomy (Figure 4). The cystic appearing lesion was type II regarding to Stocker's histologic classification of CCAM histopathologic classification. Some of the microscopic findings included enlarged alveolar and bronchial structures, absence of



FIGURE 1: Preoperative chest x-ray showing dextrocardia

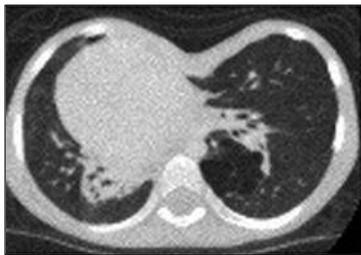


FIGURE 2: Pectus excavatum, CCAM and mediastinal shift on CT scan.



FIGURE 3: Postoperative chest x-ray.



FIGURE 4: Specimen of the left middle lobe.

septae cartilage in the masses and presence of inflammatory cellular infiltration. The patient had an uneventful postoperative course and she was discharged on the fifth day. For a better result, minimal invasive repair for her chest deformity was postponed until the age of 5 years.

## DISCUSSION

Congenital lung malformations occur rarely but could cause an important respiratory distress in the infancy. Congenital cystic adenomatoid malformation CCAM is characterized by dysplastic proliferation of bronchiolar structures and absence bronchial glands of matured alveoli and results from abnormal pulmonary mesenchymal development. CCAM is caused by a failure in the transition from the canalicular phase to the terminal saccular phase during the lung development. The estimated incidence is between 1/25,000 and 1/35,000.<sup>4,5</sup> CCAMs are 25-30% of all congenital lung malformations and approximately 30% of all patients with CCAM are at risk of respiratory distress at birth.<sup>6</sup>

Its pathologic variety correlate with symptoms. It can be presented as recurrent pulmonary infections. CCAM can lead to compression or lung hypoplasia as with other chest occupying lesions, the most common symptom is respiratory distress. The severity of symptoms depends on the degree of hypoplasia. It is not associated with chromosomal abnormalities. Other congenital defects such as diaphragmatic hernia, renal agenesis, pectus deformities and cardiac anomalies may be associated

with CCAM The most of cases involve one lobe, preferentially the lower one. The large mass inside the lung can restrict lung growth, can produce mediastinal shift, cardiovascular compromise and cava obstruction leading to non-immune hydrops fetalis. Most patients become symptomatic within the neonatal period and the less patient have symptoms in the later life. It causes respiratory distress at the times of stress, such as with infections and increased activity. The radiological features depend on the content, size, and number of cysts. The differential diagnosis of CCAM includes other cystic diseases including pulmonary sequestration, bronchogenic cyst and congenital lobar emphysema

According to the cystic characteristics, CCAM are divided into three types: Type I is macrocystic disease more than 2 cm in size and represents the majority of the cases. Type II is mix disease with small and medium-sized cysts of about 1 cm. Type III is referred to as microcystic disease.<sup>7</sup>

Failure to thrive due to recurrent pulmonary infections reminds a search for CCAM. It may be diagnosed by chance as patients are examined for

other reasons.<sup>[3]</sup> The treatment for CCAM is surgical resection even in asymptomatic patients because with longstanding disease, bronchioalveolar cancer and rhabdomyosarcoma have been reported.<sup>8</sup> Separation of cystic formation from normal tissues during surgery could be difficult. Therefore, incomplect resection is potential in procedures out of lobectomy.<sup>9</sup> This condition could also result in recurrent pulmonary infection or residual lesion. Surgical resection of the lesion as the appropriate management is required when respiratory distress is present. As complications, air leaks, pneumothorax, bronchopleural fistula, wound infections and pneumonia can occur.

CCAM is a rare but challenging pulmonary anomaly, which can be found at different ages and presents with various clinical settings. It should be considered in children with recurrent pulmonary infections or accompanying congenital malformations. Resection of all cystic adenomatoid malformations at the time of diagnosis is recommended because ongoing infections and malignant shift have been identified in children with this longstanding congenital abnormality.

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