

Congenital Cystic Pulmonary Malformations in Children: Our Experience with 19 Patients

Çocuklarda Konjenital Kistik Pulmoner Malformasyonlar: 19 Hastalık Tecrübemiz

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Abstract

Purpose. Congenital cystic pulmonary malformations (CPM) are rare anomalies. The purpose of this study was to present our experience with CPM patients who were surgically treated in our clinic and to discuss our findings along with those from the literature.

Materials and Methods: Surgical treatment was performed on 19 patients under the age of 16 who were diagnosed with CPM in our clinic between January 1995 and December 2008. The diagnoses, ages, gender, symptoms, locations of the lesions, surgical method used, hospitalization times, complications, and the results of all patients were retrospectively evaluated.

Results. The distribution of diagnoses was as follows: bronchogenic cyst(s) (BC) - 5 patients; pulmonary sequestration (PS) - 6 patients; congenital lobar emphysema (CLE) - 4 patients; congenital cystic adenomatoid malformation (CCAM) Type I - 3 patients, and PS and CCAM coexistence - 1 patient. All patients underwent resection. No operative mortality occurred. The mean postoperative hospitalization time of the patients was 6.9 days (range 4–17 days).

Conclusion. CLE, CCAM, and PS may lead to life-threatening respiratory distress in infants. BC, CCAM, and PS, on the other hand, often progress with recurrent pneumonia in children and adults. Surgery is needed to improve severe symptoms, prevent fatal complications, and establish a histopathological diagnosis.

Keywords: Congenital, Cystic pulmonary malformations, Diagnosis, Surgery

Özet

Amaç. Konjenital kistik pulmoner malformasyonlar (CPM) nadir rastlanan anomalilerdendir. Bu çalışmanın amacı cerrahi olarak tedavi ettiğimiz CPM'li olgularımızı sunmak ve literatür eşliğinde tartışmaktır.

Gereç ve Yöntem. Ocak 1995 ve Aralık 2008 arasında kliniğimizde CPM tanısı konan 16 yaş altı, 19 olguya cerrahi tedavi uygulandı. Tanı, yaş, cinsiyet, semptom, lezyonların lokalizasyonu, kullanılan cerrahi metod, hastanede yatış süresi ve komplikasyonlar geriye dönük olarak değerlendirildi.

Bulgular. Olguların 5'i bronkojenik kist (BC), 6'sı pulmoner sekestrasyon (PS), 4'ü konjenital lobar amfizem (CLE), 3'ü konjenital kistik adenomatoid malformasyon (CCAM) Tip I ve 1'i PS+CCAM idi. Tüm olgulara rezeksiyon uygulandı. Operatif mortalite gözlenmeyen olguların hastanede kalış süresi 6,9 gün (4-17) idi.

Sonuç. CLE, CCAM ve PS infantlarda hayatı tehdit eden solunum zorluğu ile çocuklar ve erişkinlerde tekrarlayan enfeksiyonlara neden olabilir. Semptomları gidermek, fatal komplikasyonları önlemek ve histopatolojik tanıyı koymak için cerrahi tedavi uygulanacak en iyi yöntem gibi gözükmektedir.

Anahtar Kelimeler: Konjenital, Kistik pulmoner malformasyonlar, Tanı, Cerrahi

Introduction

Congenital cystic pulmonary malformations (CPMs) develop due to disruptions of the mesoderm and ectoderm in the 20th and 24th weeks of intrauterine life [1]. They may be detected at any point of life. With advances in ultrasound technology, the diagnosis rate of these anomalies in prenatal life has increased [2]. CPMs in newborns and infants are usually recognized after respiratory deficiencies arise; however, they may be completely asymptomatic in adults. They are often detected during an investigation into recurrent pulmonary infections or on evaluation of an incidentally obtained pulmonary graph [3]. In some instances, the anomaly may itself present with life-threatening symptoms and thus require urgent intervention [4].

This study retrospectively evaluated the distribution of the pathologies, symptoms, radiological findings, surgical procedures, and results of the patients with CPM who were surgically treated in our clinic.

Materials and Methods

Nineteen patients were diagnosed with congenital CPMs in our clinic between January 1995 and December 2008. The diagnostic methods included pulmonary graph (PG), computer tomography (CT), and, in some patients, magnetic resonance imaging (MRI). All of the patients underwent surgical treatment depending on the type of lesion. The records of the patients were evaluated in terms of age, gender, clinical findings, type of malformation, location of cyst, surgical procedure, postoperative hospitalization time, and surgical outcome.

Results

The study included 11 male and 8 female patients (N=19), with a mean age of 7.7 years (range: 40 days to 16 years). Of the 19 patients with congenital CPM, 5 (26.3%) had bronchogenic cysts (BC). Of these 5 patients, 4 were male and 1 was female. The mean age of these patients was 9.6 years (5 months to 16 years). The symptoms of patients with BC included: coughing in 3 patients (60%); chest pain in 2 patients (40%); dyspnea in 2 patients (40%); discharge of phlegm in 1 patient (20%) and fever in 1 patient (20%). The lesions were intraparenchymal in 3 patients (2 right and 1 left) and mediastinal in 2 patients (1 right paratracheal area and 1 posterior mediastinum). Three of the intraparenchymal lesions were treated with cystectomy and capitonage. Two mediastinal lesions were resected through right thoracotomy.



Fig. 1 — Radiograph of the chest of a 2-month-old infant with lobar emphysema involving the left upper lobe.

Pulmonary sequestration (PS) was detected in 6 of the patients (31.6%). The female/male ratio was 1, and the mean age of these patients was 9.5 years (range: 2 years-15 years). Of the 6 patients, 3 had recurrent pneumonia attacks. Other symptoms included coughing and dyspnea. The lesions were located in the left lower lobe in 3 patients and the right lower lobe in 3 patients. All patients underwent lower lobectomy. The mean age of the 2 male and 2 female patients (21.0%) with congenital lobar emphysema (CLE) was 11.0 months (40 days-3 years). Severe respiratory failure, developmental disorder, coughing, and fever were the major symptoms. In two patients whose CLE was located in the left upper lobe, upper lobectomy was performed (Figure 1). One patient with CLE in the right upper lobe underwent right upper lobectomy, and one patient with CLE in the right middle lobe underwent right middle lobectomy. In 3 patients (15.8%), congenital cystic adenomatoid malformation (CCAM) Type I was detected. The mean age of the patients (1 male and 2 female) was 11.0 years. Two of these patients complained of recurrent pneumonia attacks, and the other had abdominal pain and dyspnea. Cysts were located in the left lower lobe, right lower lobe, right upper and middle lobe in 1 patient each. Right lower lobectomy, left lower lobectomy and superior bilobectomy was each performed on one patient. A 5-year-old male patient presented with the complaint of recurrent pneumonia and was diagnosed with coexisting intralobar pulmonary sequestration (ILPS) and CCAM type I (Figure 2 a,b,c,d). The lesion located in the left lower lobe was removed through lobectomy.

The age, gender, diagnosis, cyst location(s), and treatment methods of all 19 patients, who were surgically treated

for congenital pulmonary malformation, are shown in Table 1. No complications occurred in the operative or postoperative periods of the patients. The mean postoperative hospitalization time of the patients was 6.9 days (range: 4–17 days).

Discussion

Congenital pulmonary malformations are rare lesions that form during the embryological development phase of the lungs. The rate of diagnosis for congenital pulmonary malformations in both the prenatal period and adulthood has increased as a result of the advances in imaging methods. In infants, it often presents with respiratory distress and requires urgent diagnosis and treatment. In older children and adults, it either remains asymptomatic or progresses with recurrent pulmonary infections. It will thus play an important role in future coronary artery stent imaging with its high spatial resolution and rapid acquisition.

Bronchogenic Cysts

BC is the most common form of congenital cystic lesions in the mediastinum. BCs account for 34% of the cysts, 6.3% of the mediastinal masses, and almost half of all congenital mediastinal cysts [5].

Bronchogenic cysts are benign congenital lesions that form as a result of abnormal budding from the primitive tracheobronchial tube during the developmental phase of the respiratory system. They may be detected in any age group. The cysts are usually solitary, thin-walled, and unilocular. Rarely, they may be multiple or multilocular. Three-fourths of the cysts are located in the mediastinum, while one-fourth are intrapulmonary. In the mediastinum, BCs are most frequently located in the pericarinal area followed by the right trachea and hilum. They are rarely found in the posterior and/or anterior mediastinum. Intrapulmonary BCs are usually located in the lower lobes. The cysts contain bronchial cartilage, smooth muscle, elastic tissue, and mucous glands. Internally, they contain a mucoid fluid. Histopathologically, the internal cystic wall is covered with a ciliary columnar or cuboidal epithelium.

Although BC is a congenital anomaly, it is often detected in the adults, particularly in adult males. It is most commonly detected between the ages of 30 and 40 years [6].

BCs may be symptomatic or asymptomatic. The symptoms vary depending on the location and adjacent structures. Of BC patients, two-thirds are asymptomatic, and the cysts are frequently detected in adolescents and adults [7]. The mediastinally located cysts generally have a more latent progression, whereas those located in the parenchyma become symptomatic because of their proximity to the bronchial tree. Of intraparenchymal cysts, 20% are accompanied by infection. The most common symptoms are chest pain, coughing, and dyspnea [8]. Our patients had similar symptoms. The

cysts are generally thin-walled, unless they are infected. When infection develops, the cystic wall becomes thicker and squamous metaplasia may develop on the epithelium that covers the internal cyst. Cysts have close relations with the tracheobronchial tree and esophagus. In case of compression of the tracheobronchial tree, infection, hemorrhage, or rupture, they may be life-threatening.

Mediastinal bronchogenic cysts are well demarcated and homogenous on pulmonary graphs. The cysts may contain thick fluid. During thoracic radiography of intrapulmonary cysts, air, fluid, or air-fluid levels can be observed [9]. BCs are rarely detected through prenatal sonography, and CT is highly useful in diagnosis and forming differential diagnosis. In suspected cases, MRI provides additional information in differentiating cystic from solid lesions. Barium studies can show the abnormal relation between the respiratory tract and esophagus or stomach.

On differential diagnosis, esophageal duplication cysts, neuroenteric cysts, CCAM and cystic teratoma, thymic cyst, or ectopic thyroid tissue that undergoes cystic transformation, particularly when BC is located in the anterior mediastinum, should be kept in mind [5].

BCs are histopathologically diagnosable and have a low malignancy potential. Thus, to avoid any complications, they require surgical treatment. Total excision is efficient in those with mediastinal cysts; however, BCs located intraparenchymally require wedge resection, segmentectomy, or lobectomy. Despite being a benign anomaly, they may recur if not completely resected [10]. Lobectomy can be performed if atelectasia or pneumonia is frequent around the cyst. In case of small cysts, anatomical segmentectomy is proper. Three of our patients with an intraparenchymal lesion received a cystectomy and capitonage. Two lesions located mediastinally were resected through right thoracotomy.

Pulmonary Sequestration

PS accounts for 0.15–6.4% of all the congenital pulmonary malformations [11]. PSs are abnormal pulmonary tissue that may be located inside or outside of the lungs. Vascularization of the lesion, which has no direct contact with the tracheobronchial system, is provided by systemic arteries. Depending on its relation with the pleura, there are two types of PS: intralobar and extralobar. Those that arise before the visceral pleura is formed are termed intralobar pulmonary sequestration (ILPS), and those that arise after the visceral pleura is formed are termed extralobar pulmonary sequestration (ELPS). Both types have been shown to have an embryonic origin. Stocker and Malczak hypothesize infection as the etiology of intralobar sequestration [12]. This hypothesis is supported by the common symptom of recurrent pneumonia and the presence of patients with purulent bronchiectasis. On the other hand, detection of intralobar sequestration, not accounted for by infections, indicates a congenital etiology. In both types, blood flow is provided by the aorta or its

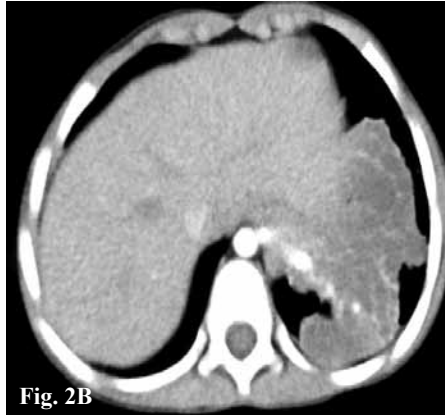


Fig. 2A — Radiograph of the chest of a 7-year-old patient with coexisting ILPS and CCAM type I in the left lower lobe.

Fig. 2B — CT of the chest of the same patient with coexistent ILPS and CCAM type I.

branches. Generally, the venous circulation of ELPS is through systemic veins (v. azygos, v. hemiazygos, vena cava), while in ILPS, it is through pulmonary veins [11]. ILPS accounts for $\frac{3}{4}$ and ELPS for $\frac{1}{4}$ of all pulmonary sequestration cases. All of our patients with pulmonary sequestration had ILPS.

The vascularization of ILPS is usually provided by an aberrant branch arising from either the thoracic aorta or abdominal aorta. Intralobar sequestration is rare in other congenital anomalies. There may be a shunt from left to left, and patients may suffer congestive heart failure. Although it is not related to the bronchial system, if infected, it may discharge into the bronchus. Nearly 60% of these cysts are located in the lower left lobe and 40% in the lower right lobe. Upper lobe involvement and bilateral involvement are very rare. Although the segment with sequestration can be removed, in most of the reported cases, lobectomy was performed. In our series, 3 patients with cysts in the left lower lobe and 3 patients with cysts in the right lower lobe underwent lobectomy. ELPS is adjacent to the left diaphragm in 90% of cases. It is usually located under the left lower lobe, and, in very rare cases, under or in the diaphragm. Its systemic artery usually arises from the abdominal aorta. It is four times more common in males than in females. The presence of intralobar and extralobar sequestrations in the same patient is rare. ELPS is frequently accompanied by diaphragmatic hernia among other congenital anomalies. Because it has its own pleural

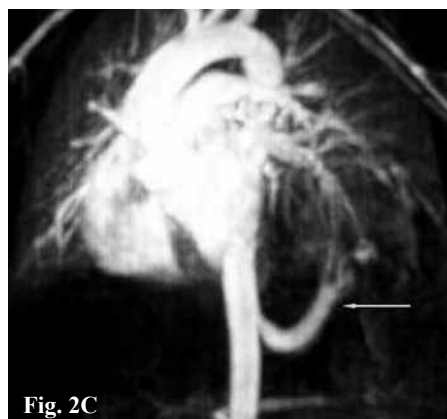
sack, the risk of infection is lower, and, unless infected, it remains asymptomatic. The treatment is the resection of the sequestered lobe.

The detection rate of ELPS is higher in the prenatal and neonatal periods, while ILPS is usually detected in childhood. In neonates and infants, sequestration is usually accompanied by recurrent lower lobe pneumonia. In childhood, most patients are asymptomatic. ELPS is usually large enough to cause respiratory distress in the neonatal period. ILPS tends to be smaller and is often incidentally detected in children and adolescents after an infection [9]. Three of our patients applied with recurrent lower lobe pneumonia.

In the prenatal period, diagnosis can be established by ultrasonography (USG) in the 4th-6th months. Prenatal MRI can show a well demarcated and defined mass with high density. On a direct thoracic radiograph, PS is classically observed as a basal mass with three corners or an oval shape, located in the posterior. Cystic pulmonary tissue containing air or air-fluid may be seen. Bronchiectasis, atelectasis, mediastinal shift, and a marked hilus on the same side are other commonly seen radiological findings. CT provides information on the location of the sequestration and the pulmonary parenchyma. Colored Doppler USG and MRI are superior to CT in reflecting the systemic arterial circulation. Differential diagnosis of PS involves tumoral lesions such as CCAM, other congenital pulmonary malformations, diaphragmatic hernia, Wilms tumor,

Fig. 2C — MRI angiography of the patient with coexistent ILPS and CCAM type I revealing the anomalous artery arising from the thoracic aorta.

Fig. 2D — The intraoperative appearance of the anomalous artery arising from the thoracic aorta.



neuroblastoma, and teratoma and accessory spleen [9].

Some authors advocate conservative treatment in most patients with PS and recommend surgery only when complications arise. Severe complications such as fungal infection, tuberculosis, fatal hemoptysis, hemothorax, cardiovascular problems, benign tumors, and even malign degeneration associated with PS have been reported [13]. Thus, the treatment of choice for PS is surgical resection. During resection, the aberrant artery should be ligated. Otherwise, the artery that shifts into or under the diaphragm may lead to uncontrollable hemorrhage. Another important point is to clearly define the venous circulation in order to avoid pulmonary infraction.

Congenital Lobar Emphysema

Congenital lobar emphysema was first reported in 1932 by Nelson [14]. It is usually characterized by hyperinflammation in one lobe. The left upper lobe (50%) and the right middle lobe (30%) are most often involved, followed by the right upper lobe. More than one lobe may be affected. However, involvement of lower lobes is rare. The incidence in male children is three times higher than in female children. Of the four patients with CLE in our series, two were male and two was female. The lesions were located in the left upper lobe in two patients, in the right middle lobe in one patient and in the right upper lobe in one patient.

The etiology for CLE is not known for almost half of patients. Entrapment of air due to the valve effect of the dysplastic bronchial cartilage, mucous plaques in the bronchus, aberrant veins compressing the bronchus, and infections causing bronchial disorders are some of the known etiologies [15]. In addition, one etiology of CLE involves polyalveolar lobe formation by numerous alveoli with normal size. No destruction occurs at the alveolar wall. However, the alveolar count is 3-5 times higher than that of normal parenchyma. The incidence rate of coexisting CLE and congenital heart failure is 20%. Anomalies such as renal agenesis, renal cyst, pectus excavatum, diaphragmatic hernia, and extremity anomalies may also coexist [16].

Most of the patients are symptomatic in the neonatal period. Myers has classified CLE into 3 clinical types: CLE type I, symptomatic in infants, CLE type II in adolescents, and CLE type 3 in asymptomatic patients with incidental diagnosis [17]. Types 2 and 3 are rare. The onset of the symptoms is usually in the first week in half of the patients and within 6 months in the other half. The number of reported cases with CLE detected in the adulthood is very limited. The most common symptom is respiratory distress. CLE is the most common etiology for Neonatal Respiratory Distress Syndrome and is usually seen in the first six months of life. Three of the four CLE patients in our series were under six months of age, and the other was three years old.

On physical examination, hyperresonance, reduced respiratory sounds, and deviation of the trachea to the opposite side are detected in the involved side. Compression of healthy

pulmonary tissue results in dyspnea, cyanosis, reduced venous circulation, hypertension, and eventually cardiac arrest. The diaphragm may shift downwards bilaterally. Direct thoracic radiography is usually sufficient in establishing a diagnosis of CLE. In suspected cases, CT aids in diagnosis [9]. On thoracic radiograph and CT, hyperlucency, collapsed adjacent lobe, and mediastinal shift as well as hernia of the hyperinflated lobe to the other side are observed. On CT, tense and thinned veins are also observed in the emphysematous lobe.

The differential diagnosis includes pneumothorax, pulmonary hypoplasia, pneumatocele and endobronchial mass, and CCAM. Congenital diaphragmatic hernia and foreign body aspiration should also be kept in mind. In many cases, tension of the lobe (excessive air) is mistaken for pneumothorax, and a thoracostomy was performed. This will not lead to clinical relief. Rather, it deteriorates the present clinical picture.

In infants with severe respiratory symptoms, pulmonary resection is needed to avoid morbidity and mortality. The recommended treatment is lobectomy. However, some authors have reported that, in asymptomatic patients or in patients with minimal symptoms, conservative treatment can be applied [18]. Infants with severe respiratory distress may need urgent thoracotomy or lobectomy. Intraoperatively, until the thorax is penetrated, excessive expansion of the lungs should be avoided in order to prevent an increase in mediastinal shift and compressive shock. After the thorax is opened, the emphysematous lobe should be taken outside the chest and lobectomy should start.

Congenital Cystic Adenomatoid Malformation

The incidence rate of CCAM varies between 1/4000 and 1/35,000 [19]. It is a congenital anomaly associated with disrupted embryogenesis and characterized by anastomosing proliferated terminal bronchioles of different volumes as well as cystic and solid structures. Its etiopathogenesis has not been fully described. It may be accompanied by cardiac, renal, and chromosomal disorders. The pulmonary segment affected by CCAM is not functional. However, because it is highly vascularized, functional areas receive less blood. Therefore, a right-left shunt and hypoxia are observed. With the development of mediastinal shift, mortality associated with left ventricle failure may occur.

CCAM is usually unilateral and located in one lobe only. Miller et al., have reported equal rates of upper and lower lobe involvement and lower rates for middle lobe involvement [20]. Hacham et al., on the other hand, reported a higher incidence rate for left lower lobe involvement by CCAM [21]. The patients with bilobar or bilateral lesions usually have a poorer prognosis due to hypoplasia in the residual lung. In three of our patients with type 1 CCAM, the lesions were located in the left lower lobe, in the right lower lobe and in the right upper and middle lobes. Nevertheless, the lesion in the patient with coexistent CCAM and ILPS was located in

the left lower lobe.

Bale has classified CCAM into three groups; cystic, intermediate, and solid, while Stoker classified CCAM into Type 1, Type 2, and Type 3, and then added Type 0 and Type 4 [22]. Type 1 and Type 4 are at increased risk of malignant transformation. Reports of patients with CCAM who developed rhabdomyosarcoma exist [16]. Type 1 is the most common form and has the best prognosis. Type 2 is usually accompanied by other congenital anomalies. Type 3 usually results in death at birth.

CCAM may present with pulmonary hypoplasia, fetal death, severe fetal hydrops, and intrauterine retardation. Half of the pediatric or adult patients of CCAM present with symptoms of pneumonia. In newborns, inflammation of the lesion area is rare, whereas in adults, it is more common [23]. Despite antibiotic treatment, no radiological improvements are observed, and symptoms tend to recur. CCAM is very rare in adults. Entrapment of air within the lesion leads to a mediastinal shift. CCAM is the second most common reason for neonatal distress syndrome.

The diagnosis of CCAM can usually be established by USG in the prenatal period. Matsuoka et al. have recommended MRI to aid in the diagnosis of atypical patients with multiple anomalies in the prenatal period [24]. The presence of fetal hydrops and cysts < 5 mm and with a solid appearance denote poor prognosis [25]. Hemodynamic changes due to cardiac compression have been shown to cause hydrops. Despite the ability to identify anomalies using USG, thoracic radiography of most newborns is normal. In the postnatal period, lesions are easily found on CT. Through imaging modalities, abnormal air is observed in CCAM type 1, air-fluid level in type 2, and fluid-filled cysts with solid structure in type 3. Definitive diagnosis, however, can be established through histopathological evaluation.

CCAM is most frequently mistaken for congenital diaphragmatic hernia. Nevertheless, it can be differentiated from a diaphragmatic hernia by observing the normal gas in the diaphragm or abdomen. Other differential diagnoses include intrapulmonary bronchogenic cyst, pulmonary sequestration (PS), congenital lobar emphysema, cystic bronchiectasis, pneumotocele, and necrotizing pneumonia. Intrapulmonary bronchogenic cysts can be differentiated because they are solitary lesions with wall cartilages and have no connection with alveoli. In PS, the artery running from the systemic circulation is observed within the involved lobe. In lobar emphysema, there are no alveoli between the cysts. A pneumotocele complex does not contain epithelial or stromal components. In children, it may be difficult to differentiate CCAM from necrotizing pneumonia that progresses with recurrent infections. The presence of recurrent infection, hyperextension of the affected lobe, and the absence of air bronchograms in the same location is suggestive of CCAM. Richard et al. reported that 50% of the patients with extra-lobar sequestration also have accompanying CCAM [26].

Patients with CCAM should be treated surgically even if they are asymptomatic because of the risk of malignancy. Lobectomy is the treatment of choice. If the lesion is located in one segment only, segmentectomy will suffice. However, pneumonectomy should be avoided. Patients who undergo surgical treatment have an optimal prognosis.

In conclusion, congenital cystic pulmonary anomalies may lead to life-threatening respiratory distress in infants. They may progress with recurrent pneumonia in children and adults and have potential for malignancy. Surgical treatment is required to eliminate symptoms, prevent complications, and establish a histopathological diagnosis.

Conflict interest statement The authors declare that they have no conflict of interest to the publication of this article.

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