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REVIEW / DERLEME

Congenital pulmonary malformations

Konjenital pulmoner malformasyonlar

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ABSTRACT

There are many congenital anomalies of the lung, some of which have no clinical symptoms and are detected incidentally, while others, particularly in the neonatal and infant period, are recognized by their typical signs, symptoms, and radiological appearance. Some congenital lung anomalies are so important that they can cause the death of the patient if not diagnosed and treated early. Classification of congenital lung anomalies is difficult since these anomalies may be related to the airway, arterial and venous vascular system, pulmonary parenchyma, and primitive anterior intestinal anomalies from which the lung originates, and some anomalies may have several etiologic origins. In this review, all subgroups of congenital pulmonary malformations will be discussed.

Keywords: Bronchogenic cyst, congenital airway malformation, congenital lung diseases, congenital lobar emphysema, scimitar.

ÖZ

Akciğerin çok sayıda konjenital anomalisi mevcut olup bunlardan bazıları hiçbir klinik semptom vermez ve tesadüfen saptanırken, diğerleri, özellikle yenidoğan ve infant döneminde, tipik bulguları, semptomları ve radyolojik görünümleri ile tanınırlar. Bazı konjenital akciğer anomalileri, erken teşhis edilip tedavi edilmedikleri takdirde hastanın ölümüne neden olabilecek kadar öneme haizdirler. Konjenital akciğer anomalileri sınıflandırmasını yapmak bu anomalilerin havayoluna, arteriyel ve venöz vasküler sisteme, pulmoner parankime ve akciğerin köken aldığı primitif ön bağırsak anomalilerine bağlı olabilmesi hatta bazı anomalilerin birkaç etyolojik kökenden gelmesi nedeniyle oldukça güçlük arz etmektedir. Bu derlemede konjenital pulmoner malformasyonların tüm alt gruplarına değinilecektir.

Anahtar sözcükler: Bronkojenik kist, konjenital hava yolu malformasyonu, konjenital akciğer hastalıkları, konjenital lobar amfizem, scimitar.

The term congenital pulmonary malformation is an umbrella term to define various forms of disorders (Table 1). Congenital pulmonary malformation accounts for 5-18% of all congenital abnormalities, with an incidence of 30-42 cases per 100,000 individuals.[1] Although some anomalies do not cause clinical symptoms at birth, severe abnormalities may present with dyspnea and cyanosis. Prompt diagnosis and treatment are essential in patients with severe respiratory distress since these conditions may have a fatal outcome. However, many of these lesions have been detected in utero in recent years. Due to these abnormalities' complex nature and variations, the decision-making process can still be troublesome for clinicians. Thus, this review aims to summarize the etiology, clinical presentation, associated complications, diagnostic methods, and therapeutic strategies of the congenital pulmonary malformations.

HISTORY

The first congenital lung anomaly was a pulmonary cyst described by Fontanus in 1639. Huber described the first intralobar sequestration (ILS) in 1777. Subsequently, Koontz described the first congenital pulmonary cystic disease in 1925. The first surgical treatment was performed by Reinhoff in 1933 on a three-year-old child with a congenital cyst. This operation was followed by lobectomy by Gross and Lewis in 1943 in a patient with congenital lobar emphysema and pneumonectomy by Gross in 1946 for cystic lung disease.[2,3]

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Table 1. Congenital pulmonary malformations

Tracheal anomalies
Agenesis and atresia of the trachea
Congenital tracheal stenosis
Bronchial anomalies
Tracheal bronchus and diverticulum
Bronchial atresia
Tracheobronchomegaly
Bronchoesophageal fistula
Bronchobiliary fistula
Bronchopulmonary foregut malformations
Pulmonary sequestrations
Bronchogenic cysts
Anomalies of the lung
Pulmonary agenesis, aplasia/hypoplasia
Congenital lobar emphysema
Congenital cystic adenomatoid malformation
Congenital vascular anomalies
Pulmonary artery anomalies
Pulmonary artery agenesis
Supravalvular pulmonary artery stenosis
Pulmonary artery sling anomaly
Isolated pulmonary artery aneurysm
Pulmonary vein anomalies
Scimitar syndrome
Pulmonary arteriovenous malformations
Pulmonary varices
Lymphatic anomalies
Lympangiomas
Lympangiectasia
Lymphangiomatosis
Lymphatic dysplasia syndrome

EMBRYOLOGY

During gestation, the fetal lung undergoes various morphologic changes. These changes occur in five stages. In the third week of gestation, the respiratory system develops as the foregut endoderm prolapses into the splanchnic mesoderm. While the respiratory mucosa evolves from the endoderm, vascular structures, smooth muscles, cartilage tissue, and other connective tissue elements develop from the mesoderm. The stages of embryogenic formation of the lung are divided into five stages: embryonic, pseudo glandular, canalicular, saccular, and alveolar. In the embryonic and pseudo glandular stages, airways (tracheobronchial system) develop, while in the canalicular, saccular, and alveolar stages, alveolar structures for gas exchange develop.[4,5]

TRACHEAL ANOMALIES

Agenesis and atresia of the trachea

This anomaly can be defined as a partial or complete absence of the trachea below the level of the larynx. It is a rare anomaly that Payne described in 1900.[5] The three subtypes of this anomaly are described as follows. In type 1, the trachea originates from the esophagus. The distal trachea, including the carina, is normal. It constitutes 10-13% of all cases. In type 2, the trachea and carina are adherent to the esophagus, and there is no residual trachea. It constitutes 59-62% of all cases. In type 3, the right and left main bronchus originate from the esophagus, and there is no carina. It constitutes 22-31% of cases.

The typical clinical triad is severe respiratory distress, inability to cry, and failure to intubate the newborn. Mortality is very high. Intubation of the esophagus can be lifesaving. Associations with bronchopulmonary malformations and cardiac, vertebral, and gastrointestinal anomalies have been described. If extracorporeal membrane oxygenation (ECMO) can stabilize the patient, colon interposition or gastric conduits can be used to achieve gastroesophageal continuity. Currently, trachea reconstruction is not possible.[6-8]

Congenital tracheal stenosis

It is a rare clinical spectrum causing the narrowing of the large airways in infants and children. It may be associated with cardiovascular anomalies, such as vascular ring, pulmonary artery sling, double arch aorta, and aberrant right subclavian artery. Cantrell and Guild[9] defined three subtypes of this anomaly. In type 1, almost the entire trachea is stenotic, whereas in type 2, the entire or a portion of the trachea has funnel-shaped stenosis. In type 3, segmental stenosis is primarily caused by the underlying vascular ring.

Patients may present with symptoms such as dysphagia, stridor, and respiratory distress. Thoracic computed tomography (CT) or magnetic resonance imaging (MRI) is beneficial in demonstrating the abnormal anatomy and narrowing of the airway. Although bronchoscopy is still the gold standard for airway visualization, it should be remembered that it may cause bronchospasm in young children.

The prognosis depends on the symptoms and the possibility of surgical treatment. Partial tracheal resection is the most common surgical method for short-segment stenoses. In long-segment stenoses, slide tracheoplasty should be the preferred method. In cases where surgical correction is impossible,

palliative options, such as balloon dilatation, split posterior tracheoplasty, stenting, local steroid injection, electroresection, and cryotherapy, should be considered.[3,10]

BRONCHIAL ANOMALIES

Tracheal bronchus and diverticulum

The tracheal bronchus is a bronchial structure originating from the trachea and terminating in a segment or lobe with regular arterial and venous blood supply. It may arise from any level, most often from the lateral wall of the trachea. Patients are usually asymptomatic, and it is detected in routine examinations by bronchoscopy or thoracic imaging (CT or MRI). Apart from symptomatic cases where tracheal bronchus stenosis results in recurrent episodes of pneumonia, surgery is not indicated. Nevertheless, the tracheal diverticulum originates from the cervical or thoracic part of the trachea and terminates as a blind pouch or with a rudimentary lung. Surgical excision of the diverticulum should be considered in two scenarios: patients with frequent infections due to the accumulated secretions and patients who had undergone surgery for another reason.[11]

Bronchial atresia

Bronchial atresia is defined as the termination of a lobe or segment bronchus into the lung tissue as a blind pouch. It is most seen in the left upper lobe apicoposterior segment bronchus. The segment distal to the atresia is emphysematous due to air diffused through the Kohn interalveolar pores and Lambert bronchoalveolar ducts. Surgical indications are recurrent and severe infections, respiratory failure, and an increase in the size of the distal translucent lung segment over time.[3,12]

Tracheobronchomegaly (Mounier-Kuhn Syndrome)

Tracheobronchomegaly is an abnormal dilatation of the trachea and the main bronchus. A malfunction in the formation of connective tissue and smooth muscle in the trachea and bronchi causes tracheobronchomegaly. Patients often remain asymptomatic until the third decade. Recurrent lower respiratory tract infections and persistent cough may be observed. Diagnosis can be made by chest radiography or thorax CT. Surgery is not indicated. Treatment is mainly supportive, which includes prevention with vaccinations, treatment of infections with antibiotics, and elimination of secretions with mucolytics and pulmonary physiotherapy.[13,14]

Bronchoesophageal fistula

Congenital bronchoesophageal fistula is an extremely rare anomaly caused by incomplete separation of the trachea and esophagus during embryonic development. It is usually detected in adulthood unless accompanied by esophageal atresia. The fistula is located most often between the esophagus and the right lower lobe bronchus. Association of esophageal web, tracheal agenesis, pulmonary hypoplasia, and diaphragmatic hernia have been reported. Patients may develop recurrent lung infections and, subsequently, bronchiectasis. The barium esophagogram is the initial diagnostic modality of choice. Although aspiration of small amounts of barium sulfate into the airway on barium esophagography may be tolerated, aspiration of large amounts of barium sulfate into the lungs may result in acute inflammation, aspiration pneumonia, and acute respiratory failure. Depending on the fistula's location and the underlying condition's severity, these patients can be considered for either esophageal or bronchial stenting. An alternative surgical treatment option is the division or excision of the fistula. In cases with accompanying bronchiectasis, resection of the involved lung may also be considered.[15]

Bronchobiliary fistula

Congenital bronchobiliary fistula is also an infrequent condition. Green sputum with bile, dyspnea, and cough are typical findings. The fistula often opens into the proximal right primary or middle lobe bronchus. Diagnostic modalities include bronchoscopy, cholescintigraphy, and thoracic MRI. Resection of the fistula tract provides curative results. Procedures such as hepatic lobectomy or Roux-en-Y hepaticojejunostomy may be required when biliary drainage is not possible.[16]

Bronchopulmonary foregut malformations

Bronchopulmonary foregut malformations are rare anomalies in which the respiratory (bronchial) system has an abnormal connection with the stomach or esophagus. The right or left lower lobe is commonly affected. The diagnosis is usually made within the first months of life. Most patients require a lobectomy due to recurrent infections. Concomitant excision of the fistula and repair of the esophageal defect is the ideal surgical procedure. Lung tissue should be saved if there are no signs of infection.[3]

Pulmonary sequestrations

Bronchopulmonary sequestration is a mass of abnormal lung tissue commonly of embryonic origin

from the caudal foregut that does not have any anatomical connection to the tracheobronchial tree. Pulmonary sequestration is vascularized by an aberrant systemic artery, most frequently arising from the descending thoracic or abdominal aorta. Venous return is usually to pulmonary veins and rarely to systemic veins. It often occurs in the basal parts of the lungs. There are two forms: extralobar sequestration (ELS) and ILS. Compared to ILS, which is incorporated into regular lung tissue, ELS is typically separated from the normal lung parenchyma by its visceral pleura. Intralobar sequestration accounts for three-fourths of the cases.[17] Since sequestration originates from the caudal diverticulum of the caudal primitive foregut, it may be associated with other congenital lung anomalies of the primitive foregut origin.[18] Ultrasonography can diagnose in the antenatal period at 16-24 weeks of gestation. Ultrasonography shows a solid, well-circumscribed echogenic mass, mediastinal shifting, polyhydramnios, and hydropic changes. Postnatal CT is highly sensitive and specific for both types of anomalies. Barium esophagography is necessary in cases where esophageal connection is suspected. The optimal approach is segmental excision of the sequestration if ILS is not infected. Whereas in infected cases, segmentectomy or lobectomy should be performed. Treatment of ELS is also surgical resection. Early identification, dissection, and ligation of the systemic artery or branches arising from the aorta, typically from below the diaphragm, is crucial in both circumstances. Video-assisted thoracoscopic surgery and robotic techniques have been used more frequently in the era of minimally invasive surgery with excellent results (Figures 1, 2).[17,19,20]

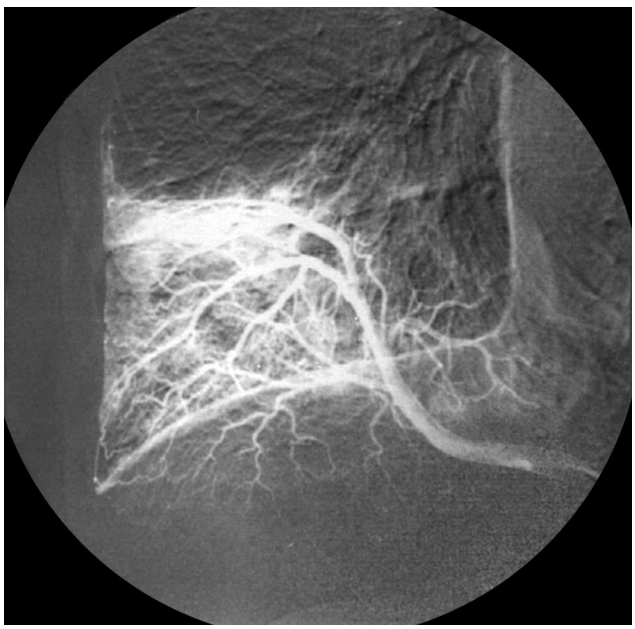

Figure 1. Aortography of a patient with extralobar pulmonary sequestration. It appears to be supplied by a thick artery arising from the abdominal aorta.

Bronchogenic cysts

Bronchogenic cysts are foregut-derived cystic malformations of the respiratory tract. They constitute approximately 60% of cysts developing in the mediastinum. The usual localization of the cyst is the right paratracheal region. They occur more commonly in males. They are usually asymptomatic and detected incidentally. Intraparenchymal cysts are more likely to be infected and may cause cough, purulent sputum, fever, and hemoptysis if infected. Thorax CT is the investigation of choice. A fluid-dense, rounded, or oval lesion with smooth margins might be seen on CT imaging. Magnetic resonance imaging is superior to a CT scan for identifying anatomic relationships. The presence of an air-fluid level suggests that the cyst is infected. The differential diagnosis should consider lymphadenopathy, pulmonary sequestration, teratoma, hemangioma, neurogenic tumors, foregut and pericardial cysts, lung abscesses, and hydatid cysts. However, the definitive diagnosis is established primarily by histopathological examination of the surgical specimen.

The treatment of patients with bronchogenic cysts is surgical resection. Excision of the cyst results in symptom relief, complication prevention, and establishment of a definitive diagnosis. Although treating asymptomatic bronchogenic cysts remains controversial, most authors support this strategy to prevent complications such as perforation, hemorrhage, growth, infection, and malignant degeneration.[21,22] Due to the possibility of recurrence following incomplete surgical excision, the resection must be complete. Needle aspiration alone may result in recurrence. Long-term results are excellent when complete resection is performed.[23,24] There has been a recent rise in the popularity of video- or robot-assisted resection techniques for the resection of bronchogenic cysts with excellent results (Figure 3).[21]

ANOMALIES OF THE LUNG

Pulmonary agenesis, aplasia, and hypoplasia

Pulmonary agenesis is the complete absence of the carina, main bronchus, lung, and pulmonary vasculature on the affected side. It is a very rare

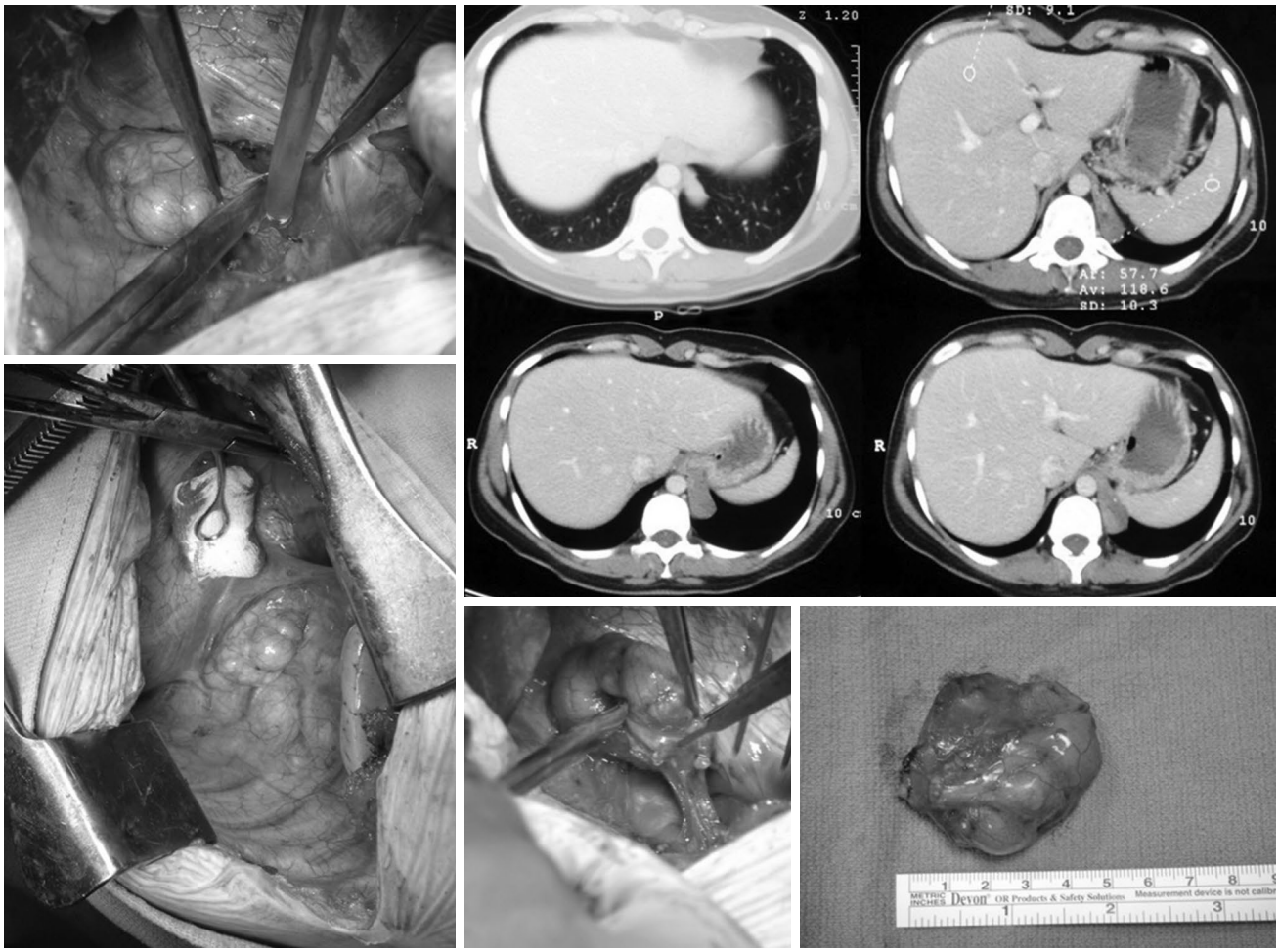

Figure 2. Thorax computed tomography of a patient with extralobar pulmonary sequestration located in the left lower zone, gross appearance of sequestration from the aorta dissection of the ascending artery and view of the extracted pathologic specimen.

anomaly and is often associated with cardiac anomalies. Imaging modalities detect a shift of the mediastinum through the affected side. Transthoracic echocardiography or angiography should be added to the diagnostic workup in cases where additional cardiac anomalies are suspected. The mortality rate is pretty high, and there is no specific treatment.[25]

Pulmonary aplasia is the presence of the carina and main bronchus without any vascularization and parenchyma. Resection of the diseased bronchial stump may be indicated due to the risk of recurrent lung infection caused by the accumulation of secretion in the blind pouch. On the other hand, pulmonary hypoplasia is defined as the underdevelopment of the bronchial tissue in combination with a reduced number of alveoli. Hypoxemia, acidosis, and hypercarbia may develop after birth in severe cases. There is no specific surgical treatment option. Optimal treatment is sedation, paralysis, high-frequency ventilation, and supportive treatment with ECMO when necessary.[3]

Congenital lobar emphysema

Congenital lobar emphysema constitutes almost half of all congenital lung anomalies. It is described as excessive distension of histologically normal and mature lung parenchyma due to disruption of bronchial continuity caused by various internal or external causes. The enlarged lung often compresses the adjacent lung and shifts the mediastinum through the opposite side. Typical findings include dyspnea, intercostal and supraclavicular retractions, wheezing, cyanosis, and feeding failure. Characteristic imaging findings include emphysematous parenchyma compressing the adjacent lung, causing atelectasis, flattening of the diaphragm on the affected side,

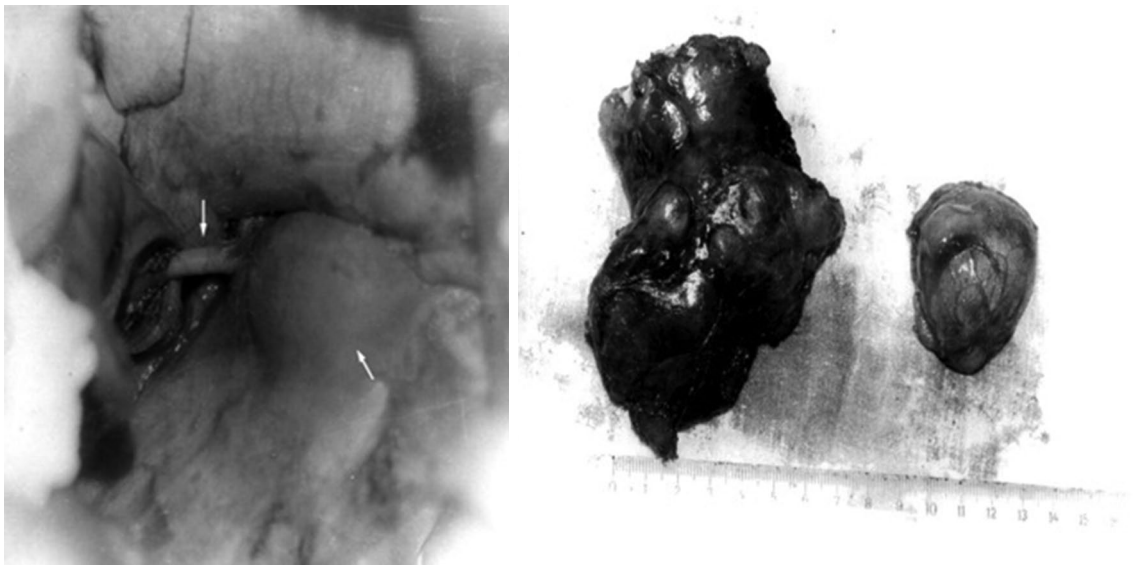

Figure 3. When a thoracotomy is performed, the bronchogenic cyst and extrapleural; it was surprising to find an intralobar sequestration located in the lower lobe, fed by a thick vessel arising from the abdominal aorta. Right lower lobectomy and bronchial cyst specimens.

and mediastinal shift towards the unaffected side.[25,26] Thorax CT is performed to confirm the chest X-ray findings and exclude mediastinal masses or vascular anomalies. In the differential diagnosis, parenchymal lung cysts, pneumothorax, and congenital diaphragmatic hernia should be considered.

The severity of symptoms identifies the appropriate treatment option. Surgical treatment is not recommended for asymptomatic or mildly symptomatic patients as pulmonary function is generally preserved and may even return to normal in infancy. Pulmonary resection (lobectomy or segmentectomy) should be considered in patients with moderate to severe symptoms (Figure 4).[3,26]

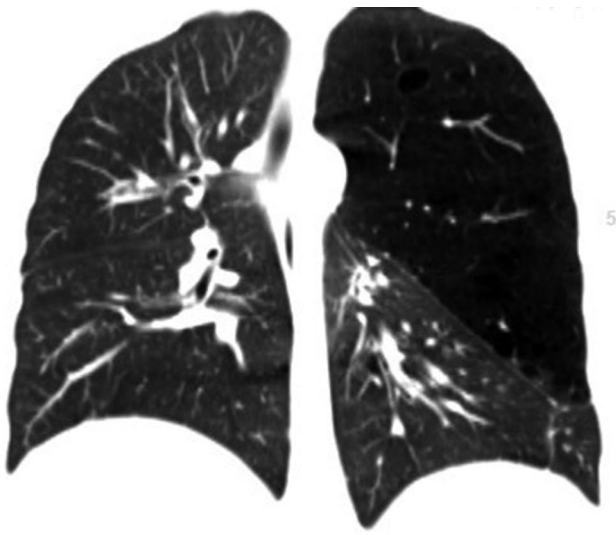

Figure 4. Congenital lobar empysema-thorax computed tomography image.

Congenital cystic adenomatoid malformation (congenital pulmonary airway malformation)

Congenital cystic adenomatoid malformations (CCAMs) of the lung are rare congenital cystic lung lesions that arise from excessive proliferation of tubular bronchial structures without normal alveolar development. It is also called congenital pulmonary airway malformation and represents 25% of congenital lung malformations and 95% of congenital lung lesions. Vascular development is typically normal, and the affected area is connected to a regular airway.[27,28] It is generally a solitary lesion affecting a single lobe. Stocker classified this anomaly into five subtypes. Type 0 involves all lobes of the lung, which is incompatible with life. Type 1 constitutes 60-70% of all cases with CCAM and contains single or multiple cysts, typically larger than 2 cm. Type 2 constitutes 15-20% of all cases, and the cysts are smaller than 2 cm in diameter. Type 3 represents the least common type (5-10%), and the cyst diameter is less than 0.5 cm. Solid lesions are typical of this kind. Type 4 cysts are typically large, peripherally located, and thin-walled.[29,30] They can present with tension pneumothorax.

Congenital cystic adenomatoid malformations may show signs of respiratory distress in the neonatal period or be unrecognized for years. The severity of symptoms is related to the extent of the lesion. The typical presentation of CCAM in postnatal life is progressive respiratory distress, including tachypnea, grunting, retraction, and cyanosis, and in adults, it usually presents as repeated chest infections. Fetal ultrasonography can be diagnostic in the prenatal period, and thoracic CT may be used postnatally.[31,32]

The treatment strategy is controversial. Around 36-97% of patients with CCAM are asymptomatic at birth and remain so afterward. While some clinicians recommend an elective surgical resection for these children, others prefer a conservative management strategy. Many surgeons still prefer the resection of the lesion, even in asymptomatic newborns, considering the risk of infection and malignant transformation. It is still debatable whether asymptomatic patients should receive surgical treatment. The results of the CONNECT (Collaborative Neonatal Network for the first European CCAM Trial) consortium, which is a randomized controlled trial comparing conservative and surgical management of patients with an asymptomatic CCAM, are expected soon and will probably guide clinicians in deciding the optimal treatment strategy.[33]

Currently, surgical resection is recommended in symptomatic patients. Lobectomy is considered the standard resection option. Results of sublobar resections, such as segmentectomy or wedge, are also satisfactory in selected cases. In recent years, successful results have been reported in resections performed with minimally invasive methods such as video-assisted thoracoscopic surgery or robotic techniques (Figure 5).[34,35]

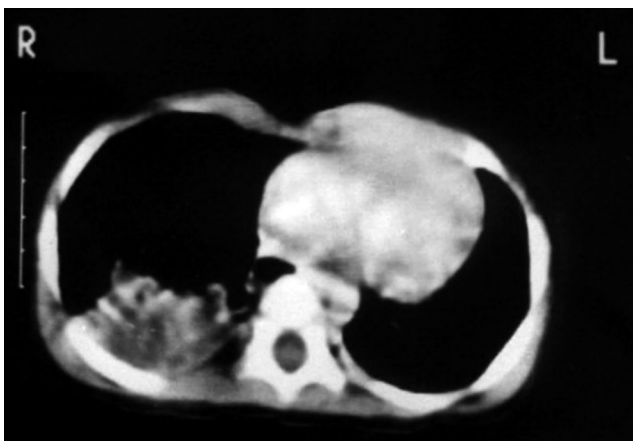

Figure 5. Congenital cystic adenomatoid malformation (congenital pulmonary airway malformation).

CONGENITAL VASCULAR ANOMALIES OF THE LUNG

Congenital vascular anomalies of the lung are relatively rare. They can be classified into three primary groups: abnormalities affecting the pulmonary arteries, those affecting the pulmonary veins, and those affecting the lymphatic vascular system. Notably, most anomalies are associated with congenital heart disease, and the clinical course may vary according to the underlying disease. While some patients may present with severe cardiorespiratory symptoms soon after birth, others may remain asymptomatic until adulthood.[36]

PULMONARY ARTERY ANOMALIES

Pulmonary artery agenesis

The agenesis of the pulmonary artery is defined as the unilateral absence of the proximal segment of the pulmonary artery. The unilateral form may occur as an isolated anomaly or associated with other cardiac or vascular anomalies. Lesions on the right side tend to occur more frequently. They are often associated with patent ductus arteriosus, whereas lesions on the left side are typically associated with tetralogy of Fallot or right arcus aorta. Blood supply of the affected lung is primarily through bronchial arteries and branches of the intercostal, internal thoracic, subclavian, and innominate arteries.[36]

The development of pulmonary hypertension (PH) determines the clinical course and prognosis of the disease. Patients who are diagnosed in adulthood typically do not have PH. These patients experience recurrent lower respiratory tract infections and hemoptysis, eventually resulting in bronchiectasis of the affected lung. The characteristic imaging findings on thoracic CT and MRI are hypoplasia of the pulmonary artery, shifting of the heart and mediastinum to the affected side, and hyperinflation of the contralateral lung.[36,37]

The therapy aims to restore the recirculation of the distal pulmonary artery before the onset of pulmonary hypoplasia. A modified Blalock-Taussig shunt could be performed to restore hypoplastic pulmonary artery flow, followed by an interposition of the pulmonary artery to the pulmonary root using autologous or prosthetic material. During adulthood, lungs are usually hypoplastic with bronchiectasis and systemic collateral vascularization. In the case of

hemoptysis, embolization is typically the first course of action. However, if symptoms cannot be controlled, pneumonectomy may be the only viable treatment option available (Figure 6).[38]

Supravalvular pulmonary artery stenosis

Pulmonary artery stenosis can either be encountered as an isolated lesion or in association with other congenital cardiac anomalies. Pulmonary valvular stenosis and ventricular septal defect are the most associated anomalies.

While patients with mild stenosis may not experience symptoms, patients with moderate to severe stenosis typically show signs of systemic venous congestion. Thoracic CT angiography can be used to visualize stenosis and right ventricular dilatation. Thoracic MRI can provide additional information about the amount and direction of abnormal flow caused by stenosis.[36,39] For multiple peripheral stenotic lesions, balloon angioplasty is the recommended treatment. However, patch arterioplasty or graft replacement is needed for central and bifurcation stenoses to ensure proper pulmonary flow.[40]

Pulmonary artery sling anomaly

Pulmonary artery sling is a specific type of compressive anatomical vascular anomaly in which the left pulmonary artery arises from the right pulmonary artery instead of arising from the main pulmonary artery and runs posteriorly between the esophagus and trachea. This anomaly is also accompanied by congenital tracheal stenosis in almost half of the patients. In this scenario, the stenosis is caused by a complete cartilage ring, which is referred to as the "ring-sling complex."[41]

Symptoms are nonspecific, are due to the abnormal anatomic course of the left pulmonary artery coursing between the esophagus and trachea, and include respiratory distress, stridor, and cyanosis. The presence of symptoms is an indication for surgery. Bronchoscopy is recommended to evaluate the extent of the tracheal ring anomaly. Thoracic CT angiography provides additional detailed anatomy.[42,43]

Surgical treatment of patients with mild tracheal narrowing without a complete tracheal ring consists of transection and reimplantation of the left pulmonary artery to the main pulmonary artery anterior to the trachea. In the presence of a complete tracheal ring, simultaneous correction should be performed with good outcomes. Tracheal stenosis repair techniques include resection with end-to-end anastomosis, patch tracheoplasty, tracheal autograft, and slide tracheoplasty. While short-segment tracheal stenosis

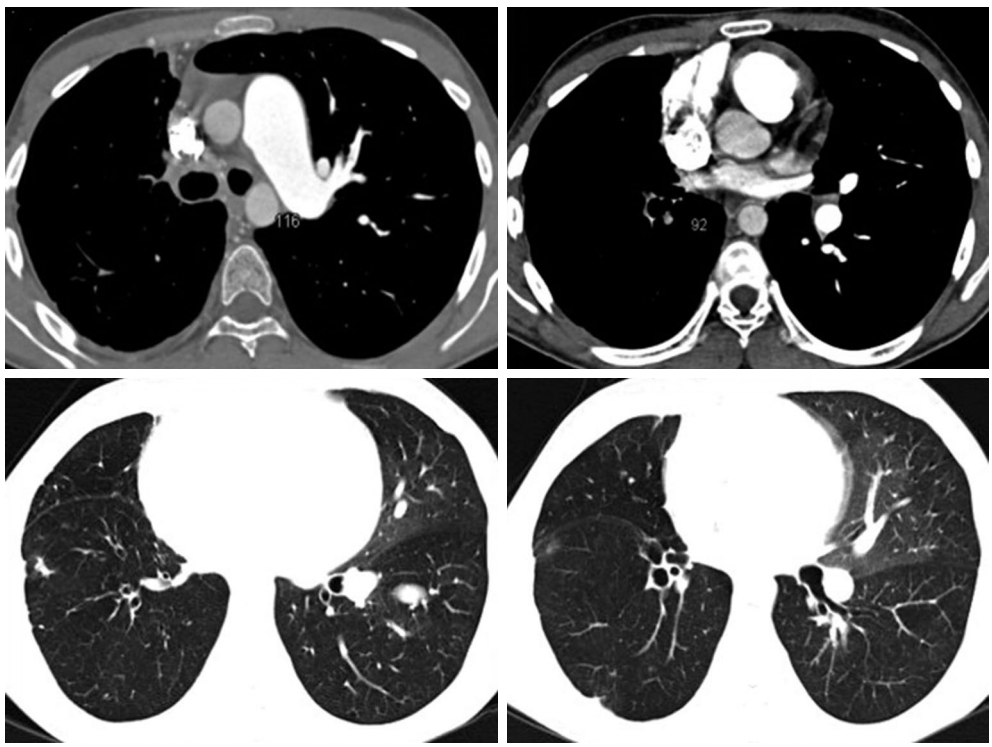

Figure 6. Pulmonary artery agenesis-thorax computed tomography image.

(less than 30%) can be repaired with resection and primary end-to-end anastomosis, slide tracheoplasty is the recommended approach in the presence of long-segment stenosis.[44,45]

Isolated pulmonary artery aneurysm

Pulmonary artery aneurysm is a rare and vital anomaly congenital in roughly 40% of cases. The remaining cases are secondary due to PH, infections (syphilis, tuberculosis, and fungal infections), vasculitis (Behçet's disease and Hughes Stovin syndrome), infective endocarditis, arteriosclerosis, penetrating trauma, and catheter-related injuries.[46] Aneurysms can develop in the main pulmonary artery, lobe, or segmental arteries. Isolated aneurysms are usually asymptomatic and appear as a mass around the pulmonary artery on chest imaging.

The treatment management strategies are variable and are based on the underlying etiology and associated comorbidities. Conservative treatment options include medical management of the underlying disease (immunosuppression in vasculitis-related aneurysms) and management of PH. Surgical techniques such as graft replacement, patch repair, primary resection-reconstruction, and pulmonary resection have been described in the literature but carry a high morbidity and mortality risk, particularly in patients with PH. Peripheral lesions can be treated with embolization without specific guidelines regarding their use (Figure 7).[47,48]

PULMONARY VEIN ANOMALIES

Scimitar syndrome

Scimitar syndrome is a rare congenital pulmonary anomaly consisting of an anomalous vein that drains part or all of the right lung blood flow into the systemic venous circulation, usually the inferior vena cava. It may also associated with an ipsilateral pulmonary hypoplasia, right pulmonary artery hypoplasia, pulmonary sequestration, and, occasionally, an anomalous systemic arterial supply from the descending aorta to the hypoplastic lung. In certain instances, the abnormal venous drainage vein extends along the right side of the heart on chest radiographs in a curved sword-like fashion (Turkish sword) and forms the characteristic image of this syndrome.[49,50]

The adult type constitutes patients who are asymptomatic, have recurrent pulmonary infections, or are diagnosed with a typical scimitar pattern on routine chest radiography. At the same time, the infantile type comprises patients who exhibit symptoms at birth and develop heart failure and PH

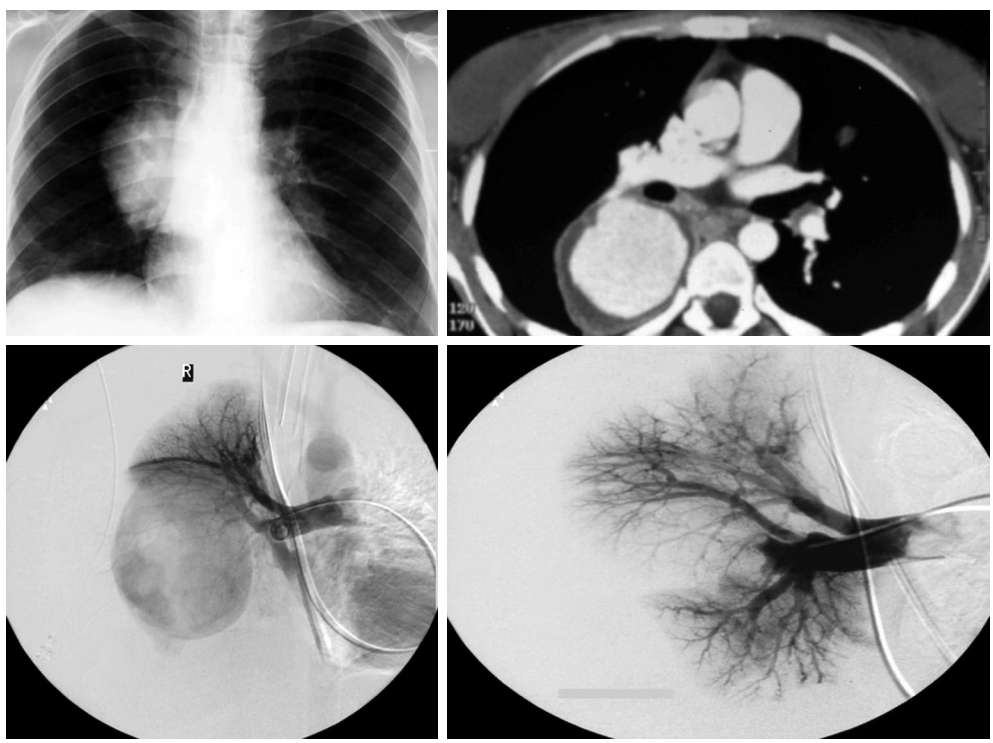

Figure 7. Endovascular treatment of Behçet's disease-related aneurysm.

around the age of one due to increased left-to-right shunt. The prognosis is relatively poor in the infantile type due to associated cardiac anomalies.[51,52]

Treatment indications comprise the development of congestive heart failure, PH, and a pulmonary/systemic flow ratio over 1.5. Surgical treatment options include restoration of the abnormal pulmonary vein flow to the left atrium either through direct anastomosis or via intracardiac patch, interruption of the arterial supply to the sequestered lung lobe, coil embolization of aortopulmonary collaterals, and pulmonary resection (lobectomy or pneumonectomy) in cases with recurrent pulmonary infections and hemorrhage (Figure 8).[53,54]

Pulmonary arteriovenous malformations

Pulmonary arteriovenous malformations are described as a direct connection between the pulmonary artery and vein branches. It can be congenital in one-third of cases as a part of hereditary hemorrhagic telangiectasia or acquired due to trauma, infections (schistosomiasis and actinomycosis), cancer, cirrhosis, mitral stenosis, and physiologic changes due to pregnancy. Typical symptoms include exertional dyspnea, hemoptysis, chest pain, cyanosis, and epistaxis. Cyanosis is typically predominant in childhood.[3]

Diagnostic methods include contrast echocardiography, lung perfusion scintigraphy, contrast-enhanced thorax CT and pulmonary angiography. If left untreated, complications such as intrapleural rupture, massive hemoptysis, paradox embolism and stroke, and cerebral abscess may occur.

Patients with small solitary lesions can be followed up for a while. The development of symptoms or the existence of hereditary hemorrhagic telangiectasia are indications for treatment. Subpleural malformations can be treated adequately with wedge resection. In bilateral, multiple, or diffuse lesions, transcatheter embolization with coils, balloons, or an Amplatzer® atrial septal occlude device should be preferred.[55-57]

Pulmonary carices

Pulmonary varices are aneurysmal dilatation of the pulmonary veins. Pulmonary venous hypertension and inflammation are the underlying causes of this condition. Congenital varices, however, result from the dilation of embryologic venous drainage channels and are associated with

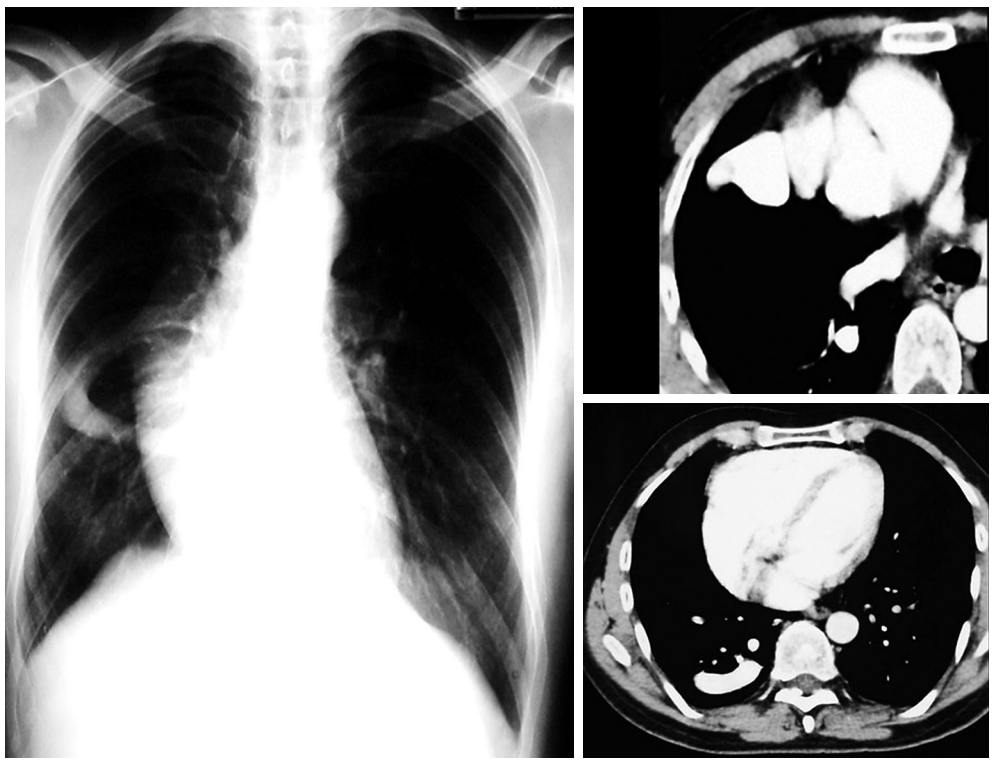

Figure 8. In another case of adult Scimitar syndrome, we found that the scimitar vein, it is observed to be slightly more transverse than normal.

mitral valve disease or other congenital heart diseases in most cases.[58]

Patients are usually asymptomatic. A definitive diagnosis is made by angiography. Varices remain asymptomatic for years and rarely may lead to hemoptysis or hemothorax if they rupture into the bronchus or pleural cavity. Congenital pulmonary varices do not progress, do not usually cause symptoms or complications, and typically do not require treatment. However, it is essential to differentiate pulmonary varices from pulmonary arteriovenous malformation, which requires treatment. When accompanied by a cardiac anomaly, the varices usually regress with treating the underlying anomaly. Surgical resection should only be considered when complications such as hemoptysis and hemothorax arise.[59,60]

PULMONARY LYMPHATIC ANOMALIES

Lymphatic anomalies can be classified as lymphangiomas, lymphangiectasia, lymphangiomatosis, and lymphatic dysplasia syndrome. Lymphangioma is a localized proliferation of lymphatic tissue, lymphangiectasia is the presence of dilated lymphatics, and lymphangiomatosis is the presence of multiple lymphangiomas. Lymphatic dysplasia syndrome consists of primary lymphedema syndrome, congenital chylothorax, and yellow nail syndrome.[61,62]

Thoracic lymphangiomas are usually seen on thoracic CT imaging as well-circumscribed, multiloculated, and heterogeneous masses that arise from the thoracic wall or mediastinum. Although their growth rate is relatively slow, they may cause symptoms by compressing the surrounding structures over time. Surgical excision plays a dual role and serves a diagnostic and therapeutic purpose.[63]

Pulmonary lymphangiectasia can be primary or secondary. The primary form occurs due to dilatation of the lymphatic channels in the lung. It frequently affects newborns and typically has a fatal outcome. The secondary form can appear at almost any age due to underlying pulmonary venous obstruction. Almost half of patients with pulmonary lymphangiectasia have an underlying congenital heart disease. Venous obstruction causes lymphatic obstruction, which usually results in chylothorax. Histologic examination is necessary for definitive diagnosis. Differentiation from lymphangiomyomatosis is challenging even with immunohistochemical examination. Treatment is usually supportive. Medications include corticosteroids, octreotide, and antiplasmin therapy. Fibrin glues or pleurodesis with autologous blood can be used in cases with refractory chylothorax. Surgery is rarely indicated in isolated lesions. High-frequency oscillatory ventilation or ECMO may be necessary in cases with persistent PH. Unfortunately, the condition frequently progresses and has a fatal outcome.[62,64]

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