Extrapulmonary sequestration mimicking mediastinal cyst: Report of two identical cases

Ahmet Sami BAYRAM[1], Cengiz GEBİTEKİN[1], Murat BİÇER[2]

[1] Uludağ Üniversitesi Tıp Fakültesi, Göğüs Cerrahisi Anabilim Dalı,
[2] Uludağ Üniversitesi Tıp Fakültesi, Kalp Damar Cerrahisi Anabilim Dalı, Bursa.

ÖZET

Mediastinal kist gibi davranan ekstrapulmoner sekestrasyonlu iki olgu

Ekstrapulmoner sekestrasyon nadir olarak görülen ve genellikle yaşamın ilk altı ayında ortaya çıkan konjenital anomalidir. Sıklıkla sol akciğer alt lobu ile diyafragma arasında yer alır. Mediastinal yerleşim nadirdir. Biz bu yazımızda rastlantısal olarak tespit ettiğimiz ekstrapulmoner sekestrasyonlu iki erkek çocuk olgumuzu tanımladık. Bu tanı; anterior mediastinal kitleli tüm çocuklarda akılda tutulmalıdır.

Anahtar Kelimeler: *Sekestrasyon, mediasten, konjenital lezyonlar.*

SUMMARY

Extrapulmonary sequestration mimicking mediastinal cyst: Report of two identical cases

Ahmet Sami BAYRAM[1], Cengiz GEBİTEKİN[1], Murat BİÇER[2]

[1] Department of Chest Surgery, Faculty of Medicine, Uludağ University, Bursa, Turkey,
[2] Department of Cardiovascular Surgery, Faculty of Medicine, Uludağ University, Bursa, Turkey.

Extrapulmonary sequestration (EPS) is a rare congenital anomaly usually diagnosed during the first six months of life. It is mostly found between the left lower lobe of the lung and the diaphragm. Mediastinal presentation is extremely rare. We

Yazışma Adresi (Address for Correspondence):

Dr. Ahmet Sami BAYRAM, Uludağ Üniversitesi Tıp Fakültesi, Göğüs Cerrahisi Anabilim Dalı, 16059 BURSA - TURKEY

e-mail: asbayram2@yahoo.com

describe two identical cases of extrapulmonary sequestration mimicking mediastinal cystic mass in two boys. These emphasize the need to keep this diagnosis in mind when dealing with children presenting with anterior mediastinal mass.

Key Words: *Sequestration, mediastinum, congenital lesions.*

Pulmonary sequestration is a complex group of anomalies involving the anomalous connections of the pulmonary parenchyma and systemic vasculatures. Classic sequestration may be extralobar or intralobar according to its relationship to the normal lung parenchyma. Extrapulmonary sequestration is clearly congenital in origin and usually associated with chest wall deformities, cardiac anomalies etc. The blood supply is almost always from the thoracic aorta and venous return to the pulmonary veins. The aetiology of the disease is still unknown.

Extralobar sequestration is usually diagnosed in the left hemithorax between the left lower lobe and diaphragm, but subdiaphragmatic, mediastinal, intrapericardial and retroperitoneal locations have also been reported (1,2).

CASE REPORTS

Case 1

A seven-years-old boy presented with flu-like symptoms, anterior chest pain and fatigue of one-week duration. Chest X-ray revealed a mediastinal mass, Figure 1A. Computerized tomography (CT) confirmed an anterior cystic mass 6 x 3 cm in size, Figure 1B. The possible diagnosis was a thymic cyst. However, a median sternotomy revealed a mass receiving its arterial supplies from the arch of the aorta and the left pulmonary artery with its venous drainage to the left superior pulmonary vein. The complete removal of the mass was achieved. Operative findings and the histopathology (sections of the mass showed lung parenchymal tissue comprising dilated duct-like bronchioles and alveoli) confirmed the extrapulmonary sequestration. His postoperative recovery was uneventful.

Case 2

A routine preoperative chest X-ray on a 10-years old boy referred for pectus excavatum repair, Figure 2A, revealed a mediastinal shadow that was confirmed on CT to be a lobulated cystic mass in the anterior mediastinum, Figure 2B. Left anterior thoracotomy was performed and a lobulated cystic mass with its arterial supply from the left main pulmonary artery and its ve-

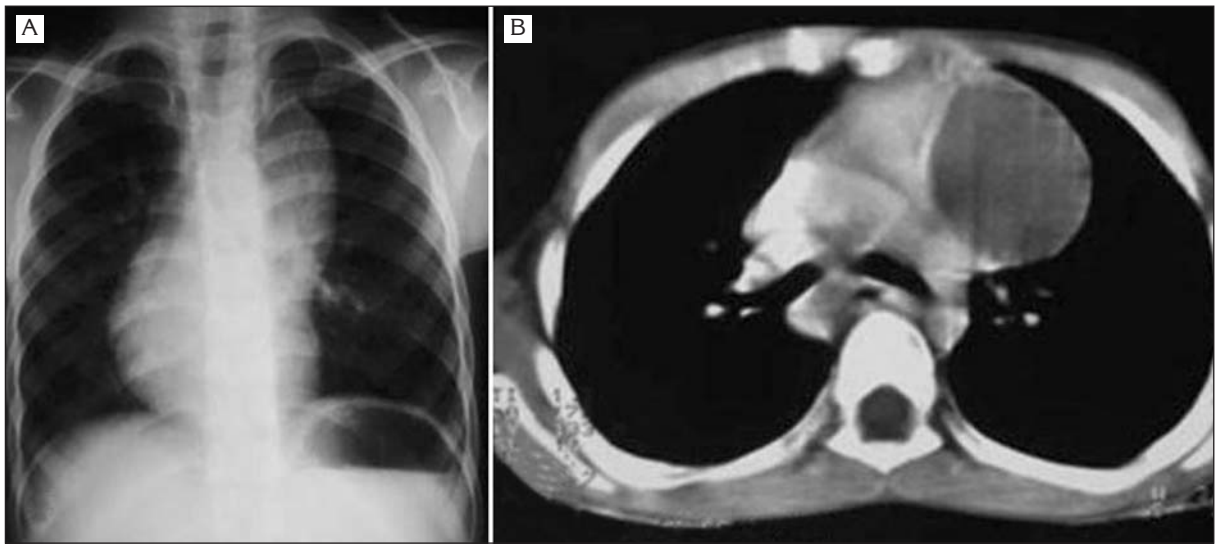

Figure 1. A: The anterior mediastinal mass displacing the mediastinum to the right on frontal chest X-ray of case 1. B: CT appearance of mass in case 1.

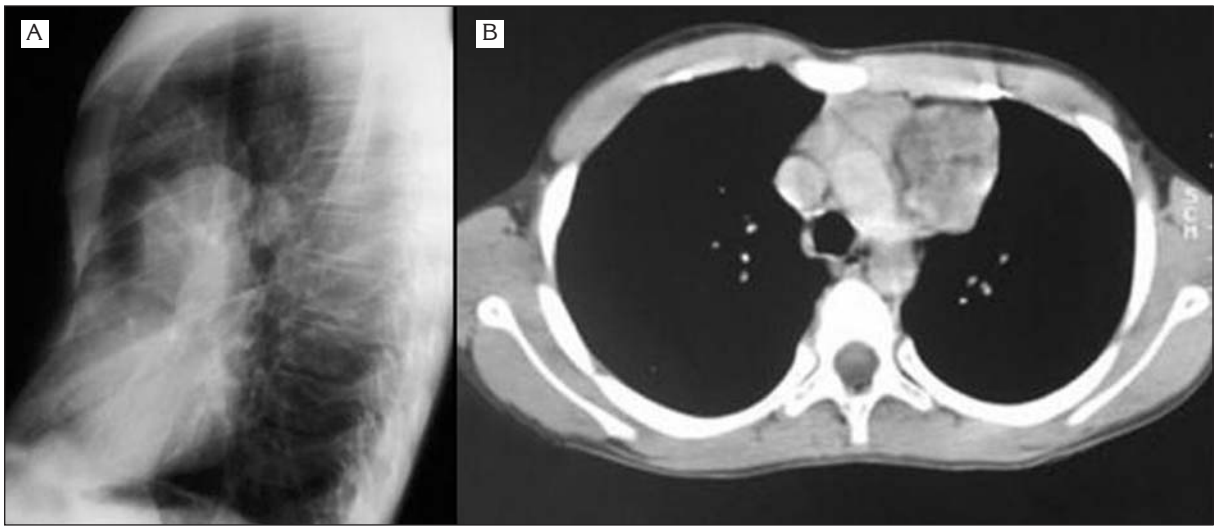

Figure 2. A: Mediastinal mass and pectus excavatum on lateral chest X-ray of case 2. B: CT appearance of mass in case 2.

nous return into the left superior pulmonary vein was removed. Subsequent histopathology (sections of the mass showed lung parenchymal tissue comprising dilated duct-like bronchioles and alveoli. Focally, they formed cysts of bronchogenic origin filled with a large amount of inspissated mucin) confirmed an EPS. The postoperative recovery was uneventful. The repair of pectus excavatum was performed three months after the surgery.

DISCUSSION

Pulmonary sequestration; first described by Rektorzik in 1861, pulmonary sequestration is defined as a mass of nonfunctional pulmonary tissue that lacks a normal communication with the tracheobronchial tree, is a rare congenital anomaly comprising 0.15-6.4% of all congenital pulmonary malformations (2,3). It is characterized by a mass of non-functioning lung tissue that lacks a tracheobronchial communication and is supplied by one or more anomalous systemic arteries from the thoracic or abdominal aorta or its major branches. Although the intralobar sequestration is diagnosed in 75% of cases, EPS is diagnosed in only 25% of cases, in which the abnormal tissue has its own pleura, arterial supply from the thoracic aorta or rarely from the pulmonary artery and venous return into systemic veins (4). Whilst intralobar sequestration shows no preference for one or the other side of the thorax, 80% of the EPS occur predominantly between the lower lobe and the diaphragm. In addition, subdiaphragmatic, pericardial, mediastinal and retroperitoneal locations have also been reported (1,2). Savic et al. (2) reported on 133 well-documented cases of sequestration and only 4% were found in the mediastinum. Sixty-five percent of the cases are associated with other congenital abnormalities such as diaphragmatic hernias, pectus excavatum, foregut duplication cysts of mixed bronchogenic and esophageal type and congenital heart defects (5,6).

The EPS is more common in males and almost two thirds of cases are symptomatic. Symptoms are usually caused by left to right shunting, anatomic impingements or associated anatomic defects and comprise chronic cough, recurrent pneumonia, chest pain, respiratory distress and haemoptysis. One of our cases presented with flu-like symptoms and anterior chest pain whilst the other was an incidental finding prior to his pectus excavatum repair. Interestingly these patients were diagnosed with anterior mediastinal masses and, due to their site, cystic teratoma, tymic cyst, cystic adenoid malformation were considered a likely diagnosis. They are clinically important lesions and failure to diagnose pulmonary sequestrations can have devastating intra-

operative consequences. There are reports of several patients who have bled to death when the feeding vessels were not properly ligated (7).

However, establishing a preoperative diagnosis can be difficult. Non-invasive procedures such as US, preferably spiral CT and MRI might demonstrate the blood supply but failure to demonstrate the vascular connections with these methods does not exclude the diagnosis (8). A definitive diagnosis can be best established by angiography/aortography, which usually reveals the anomalous systemic vessels supplying the sequestration (8). In contrast to usual blood supply, the branch from the left pulmonary artery was the feeding artery of the sequestered lung in our one child and the thoracic aorta in the other. Gamillscheg et al. found a similar unusual artery originating form the subclavian artery and venous return into subclavian vein (9). The pulmonary venous drainage is as important since the anomalous vein may drain not only the sequestrated area but also a major part of the adjacent lung. Haemorrhagic pulmonary infarction caused by ligation of the anomalous vein has been reported (10).

Although various investigations are useful in diagnosing this condition, a high index of suspicion remains a crucial element in diagnosis and based on these two case reports, the need to include pulmonary sequestration into the list of differential diagnosis of an anterior mediastinal cystic mass is once again reiterated.

REFERENCES

1. *Felker RE, Tonkin ILD. Imaging of pulmonary sequestration. Am J Roentgenol 1990; 23: 220-3.*
2. *Savic B, Birtel FJ, Tholen W, et al. Lung sequestration: Report of seven cases. Thorax 1979; 34: 96-101.*
3. *Lupinski RW, Agasthian T, Lim CH, et al. Extrapulmonary sequestration simulates posterior neurogenic tumor. Ann Thoracic Surg 2004; 77: 2203-4.*
4. *Gamillscheg A, Beitzke A, Smolle-Jütnerr FM, et al. Extralobar sequestration with unusual arterial supply and venous drainage. Pediatr Cardiol 1996; 17: 57-9.*
5. *Stocker JT. Sequestration of the lung. Semin Diagn Pathol 1986; 3: 106-21.*
6. *Kim KW, Kim WS, Cheon JE, et al. Complex bronchopulmonary foregut malformation: Extralobar pulmonary sequestration associated with a duplication cyst of mixed bronchogenic and oesophageal type. Pediatr Radiol 2001; 31: 265-8.*
7. *Flye MW, Conely M, Silver D. Spectrum of pulmonary sequestration. Ann Thorac Surg 1976; 22: 478-82.*
8. *John PR, Beasley SW, Mayne V. Pulmonary sequestration and related congenital disorders. A clinico-radiological review of 41 cases. Pediatr Radiol 1989; 20: 4-9.*
9. *Schwartz MZ, Ramachandran P. Congenital malformations of the lung mediastinum-A quarter century of experience from a single institution. J Pediatr Surg 1997; 32: 44-7.*
10. *Clements BS, Warner J, Shinebourne EA. Congenital bronchopulmonary vascular malformation: Clinical application of a simple approach in 25 cases. Thorax 1987; 42: 402-16.*