Our Cases with Scimitar Syndrome

Scimitar Sendromlu Olgularımız

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ABSTRACT Scimitar syndrome is a rare congenital cardiopulmonary system abnormality with an incidence of 0.001-0.003% that is characterized mostly with anomalous pulmonary venous drainage from right lung to the inferior vena cava instead of the left atrium, hypoplasia of the right lung and right pulmonary artery with dextroposition of the heart, and anomalous systemic arterial blood supply to the right lung. There also may be anomalies of the bronchial supply to the right lung; therefore, sequestration is frequently present. We present three cases with Scimitar syndrome, two of them were recognized while investigating for recurrent pulmonary infection and one of them was determined incidentally during catheterization. Two cases with recurrent pulmonary infection were researched for an opacity localized at the inferior zone of the right lung on the plain chest radiography. The other case was recognized during the angiographic study of descending aorta for aort coarctation when an abnormal artery supplying the inferior zone of the right lung was seen.

Key Words: Scimitar syndrome; heart defects; congenital; child

ÖZET Scimitar sendromu, yüz binde 1-3 oranında görülen seyrek bir konjenital anomali olup, sağ akciğer kökenli bir pulmoner venin sol atriyum yerine sıklıkla vena kava inferiora açılması ile birlikte bazen sağ akciğer ve sağ pulmoner arterin hipoplazisi, kalbin dekstropozisyonu ve sağ akciğerin bir bölümünün sistemik arteriyel kanlanması özelliklerini taşır. Sağ akciğerde anormal bronşiyal dallanma nedeniyle sekestrasyon oluşumu sıklıkla vardır. Bu yazıda ikisi tekrarlayan akciğer enfeksiyonu nedeni ile araştırılırken, diğeri ise kalp kateterizasyonu sırasında, Scimitar sendromu tanısı alan 3 olgudan söz edilecektir. Olguların ikisinde tekrarlayan akciğer enfeksiyonu nedeniyle çekilen akciğer grafilerinde sağ akciğer alt zonunda opasite artışı saptanması üzerine Scimitar sendromundan şüphelenilip yapılan incelemelerde, diğerinde ise aort koarktasyonu için uygulanan kalp kateterizasyonunda inen aortaya yapılan enjeksiyonda aortadan köken alan bir arterin sağ akciğer alt lobunu beslediğinin gözlenmesi üzerine yapılan anjiyografik incelemede tanı konmuştur.

Anahtar Kelimeler: Scimitar sendromu; kalp kusurları; doğumsal; çocuk

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Scimitar syndrome, also known as venopulmonary syndrome is a rare pulmonary venous return anomaly.[1,2] In this pathology, one or more pulmonary veins of right lung, usually drain to inferior vena cava (IVC). Moreover, hypoplasia of the lower lobe of right lung, dextroposition of heart, hypoplasia of right pulmonary artery and the systemic arterial circulation of right lung are often seen.[1-4]

Atrial septal defect (ASD), ventricular septal defect (VSD), coarctation of aorta and anomalies of arcus aorta that can be seen together are the main

factors to determine the prognosis of the disease with pulmonary hypoplasia.[2,4-6] In this report, we have presented three cases of Scimitar syndrome from 2352 patients admitted to Dr. Sami Ulus Child's Hospital between May 2002 and January 2009 and cardiac catheterization has been performed.

CASE REPORTS

CASE 1

A six months old female patient was admitted because of her third recurrent pulmonary infection. In the physical examination, rales were heard at the both of the lung's inferior zones. Scimitar syndrome was suspected because of increased opacity at the inferior zone of the right lung at chest X-Ray (Figure 1). Echocardiographahy showed absence of a right pulmonary vein and left atrial isomerism. The computerized thorax tomography revealed that the inferior pulmonary vein of the right lung opened through the vena cava inferior instead of the left atrium and lower of the right lung was hypoplastic. The cardiac catheterization and angiography showed that the right pulmonary artery was hypoplastic, the lower part of the right lung was supplied by an artery that roots from the descendent aorta and a part of right pulmonary venous blood was draining into the junction of the right atrium and IVC.

CASE 2

12 months old male patient admitted because of his fifth pulmonary infection. A transcatheter coil occlusion of patent ductus arteriosus (PDA) was performed to him six months ago. In physical examination, a grade 1/6 systolic murmur was heard at the right second intercostal area. Scimitar syndrome was suspected because of nonhomogen opacity in the lower zone of right lung on chest X ray. Lower right pulmonary vein was not be able to displayed in the echocardiographic study. Cardiac catheterization and angiography studies showed that combined right upper and middle lobe pulmonary veins drain into the left atrium (Figure 2). The infradiaphragmatic vascular stractures originated from descendent aorta were shown to perfuse lower lobe of right lung at descendan aorta injection.

CASE 3

3.5 months old male patient was referred with a diagnosis of coarctation of aorta and ASD. On physical examination, breath sounds were decreased over the right lower lung on auscultation. A grade 2/6 systolic murmur was heard at the left second intercostal area and his back. Blood pressure was decreased at the lower extremities, increased at the upper extremities. His femoral pulses were weak.

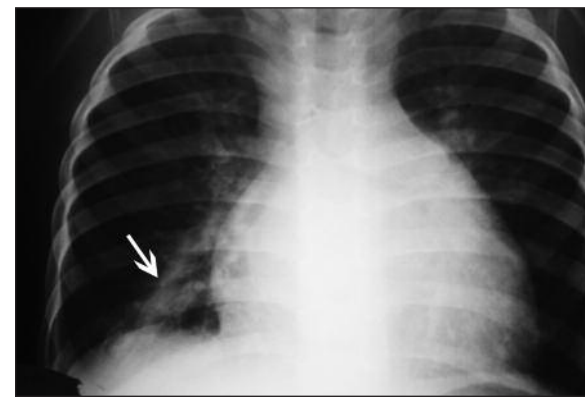

FIGURE 1: Chest X-ray of the case 1 showing Scimitar sign and increased opacity at the inferior zone of the right lung (White arrow).

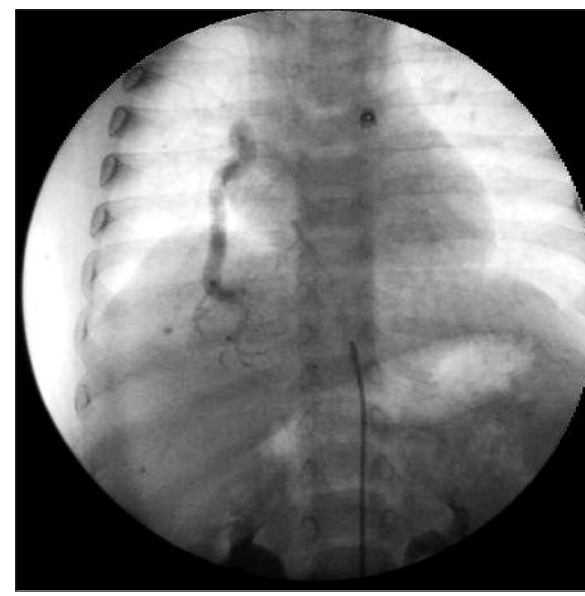

FIGURE 2: Scimitar sign is seen caused by pulmonary vein of the right lung in the venous return phase of right pulmonary artery injection.

The dextroposition of heart was seen on telecardiogram. In the echocardiographic study, coarctation of aorta was detected and 44 mmHg pressure gradient was measured at this localisation. The catheterization study revealed a small artery originated from infradiaphragmatic part of the aorta supplying the right lower lung (Figure 3), hypoplasia of right pulmonary artery and anomalous drainage of right pulmonary veins to the suprahepatic portion of IVC. Aortic coarctation was seen as a tortuous stucture. In the coarctation area, gradient decreased from 44 mmHg to 13 mmHg after baloon coarctation angioplasty.

DISCUSSION

Scimitar syndrome is a rare form of partial anomalous pulmonary venous return. It has first been reported in 1836 and was published as the name "Scimitar syndrome" by Neil and friends in 1960.[2] It accounts for 0.5-1% of the congenital heart diseases and its overall incidence is 1-3/100 000.[6] The three of our 2352 (0.12%) patients undergoing diagnostic and/or interventional cardiac catheterization were diagnosed as Scimitar syndrome between May 2002 and January 2009. This syndrome has a wide spectrum of symptoms and findings a number of which are related to the accompanied anomalies.[5] Although infants are often diagnosed with symptoms of recurrent pulmonary infections, respiratory failure or heart failure, adult patients mostly take the diagnosis incidentally or sometimes while investigation of recurrent haemoptysis or bronchopulmonary infections.[3-5] Abnormal pulmonary vein of the right lung usually opens into vena cava inferior and rarely can drain into the coroner sinus or into the left atrium as in one of our patients.[7]

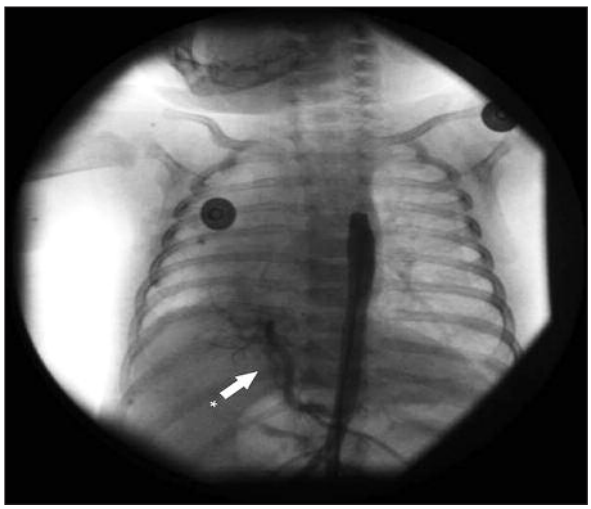

FIGURE 3: The artery originated from descendan aorta and perfusing the right lung in third case (White arrow).

We detected left atrial isomerism and hypoplasia of right pulmonary artery in first case; PDA in second case; ASD, coarctation of aorta, hypoplasia of right pulmonary artery and dextroposition of heart in third case. In fact, a dextroposition as a result of hypoplasia of the lower lobe of right lung is detected in Scimitar syndrome, although some authors esignate this pathology as dextrocardia.[2,3,6]

Recurrent pulmonary infections are expected depending on the degree of the right pulmonary hypoplasia.[2,3] After all, the two of our cases were diagnosed while investigating the cause of recurrent pulmonary infections.

Detailed evaluation is substantial for the management of Scimitar syndrome's diagnosis and treatment. A right sided pulmonary venous drainage into the inferior vena cava causes a characteristic sign, curved vascular shadow along the right heart border on the chest radiograph, called "Scimitar sign".[7,8] Computerized tomography and magnetic resonance imaging help to show detailed vascular and paranchimal structures of mediastinum,[4] whereas bronchoscopy and bronchography are useful for investigating pulmonary components.[4] Echocardiographic study is a valuable method to detect pulmonary vein drainage and additional cardiac anomalies. However, angiography and cardiac catheterization are the indispensable methods to know detailed vascular structures, pulmonary artery pressure, calculation of the ratio of pulmonary and systemic flows and planning treatment. However, the value of oxymetric study is limited owing to poor stream of pulmonary vein and participating of the high- oxygen saturated blood flow from renal veins to vena cava inferior.[9]

Scimitar syndrome is often detected incidentally, sometimes it is a rare cardiopulmonary anom-

aly that can be overlooked.[4] While clinical follow-up is often enough in asemptomatic cases, surgical treatment is necessary in cases with recurrent pulmonary infections and respiratory or heart failure.[1,5]

In conclusion, it will be useful to keep this anomaly in mind in children who have recurrent pulmonary infections and must be investigated for other congenital heart diseases.

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