Arch Iranian Med 2009; 12 (1): 79 – 81

Case Report

Scimitar Syndrome

Soheila Khalilzadeh MD•*, Maryam Hassanzad MD*, Amir-Ali Khodayari MD*

Scimitar syndrome or congenital pulmonary venolobar syndrome is a rare anomaly most commonly consisting of partial pulmonary venous drainage into the hepatic portion of the inferior vena cava, right lung hypoplasia, dextroposition of the heart, and anomalous systemic arterial supply from aorta or one of its branches to the right lung. We report a four-year-old girl with recurrent pneumonia and failure to thrive, who was diagnosed as having scimitar syndrome.

Archives of Iranian Medicine, Volume 12, Number 1, 2009: 79 – 81.

Keywords: Abnormality • pulmonary • venous

Introduction

Scimitar syndrome or pulmonary venolobar syndrome is a rare, complex, and variable malformation of the right lung characterized by an abnormal right sided pulmonary drainage into the inferior vena cava, malformation of the right lung, abnormal arterial supply, and sometimes cardiac malformation.[1]

Despite the varying degrees of pulmonary hypoplasia and pulmonary artery hypertension, about half of the patients with scimitar syndrome are asymptomatic or mildly symptomatic when the diagnosis is made. Neonates have severe symptoms and worse prognosis while older children come to light because of recurrent respiratory infections, heart murmur, or an abnormal chest radiograph.[2]

We present a four-year-old girl with recurrent pneumonia and failure to thrive, who was diagnosed as having scimitar syndrome.

Authors' affiliation: *Chronic Respiratory Disease Center, National Research Institute of Tuberculosis and Lung Disease (NRITLD), Masih Daneshvari Hospital, Shaheed Beheshti University of Medical Sciences, Tehran, Iran.

•**Corresponding author and reprints:** Soheila Khalilzadeh MD, Chronic Respiratory Disease Center, NRITLD, Masih Daneshvari Hospital, Shaheed Beheshti University of Medical Sciences, Tehran, Iran.
P.O. Box: 19575/154
Tel: +98-212-297-0757, E-mail: soheilak@yahoo.com
Accepted for publication: 18 December 2007

Case Report

A four-year-old girl was referred to our hospital because of recurrent pneumonia and failure to thrive. Her examination was entirely normal except for right sided decreased lung sounds. There was no evidence of any external congenital anomaly.

Laboratory evaluation revealed normo-chromic normocytic anemia. The rest of the laboratory tests were normal.

The chest radiograph showed a small right hemithorax and an anomalous pulmonary vein on the right side (Figure 1).

According to the findings of chest radiography, the scimitar syndrome was suggested. For confirmation of the diagnosis computed tomography (CT) of the thorax was performed, which showed right sided volume loss of the lung parenchyma along with the shift of the heart and mediastinum to the right and dextroposition of the heart. Dilated vascularity was detected in the right lower lobe with homogeneous enhancement and bronchiectatic changes (Figures 2 and 3).

For detection of associated anomalies, echocardiography and abdominal ultrasonography were performed, which were

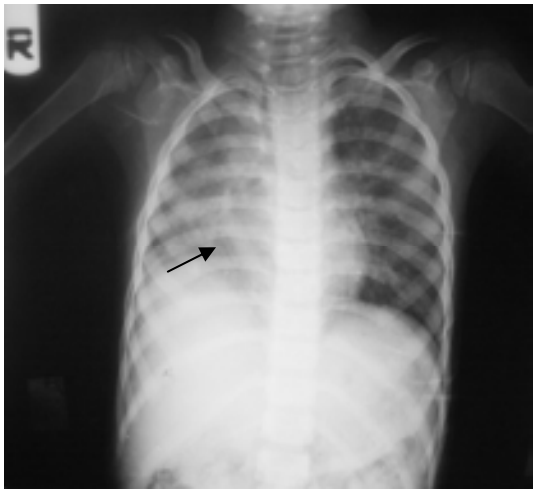

Figure 1. The chest radiograph of the patient shows a small right hemithorax and an anomalous pulmonary vein on the right side.

unremarkable.

The hypoplastic right lung and anomalous pulmonary vein confirmed the diagnosis of congenital pulmonary venolobar syndrome also known as scimitar syndrome.

Discussion

In addition to classic findings of scimitar syndrome, anomalies include hypoplastic or absent pulmonary artery, anomalous systemic arterial supply to the right lung from aorta or one of its branches and bronchopulmonary sequestration, absence of inferior vena cava, and accessory diaphragm.[2,3] The incidence of

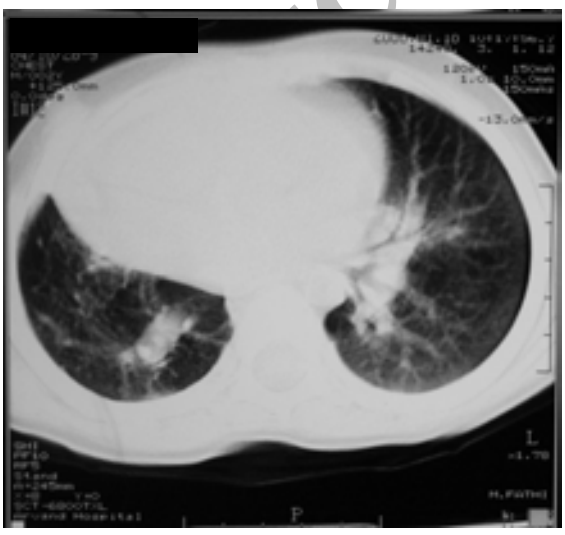

Figure 2. Tomogram of the thorax shows right sided volume loss of the lung parenchyma and bronchiectatic changes in the right lower lobe.

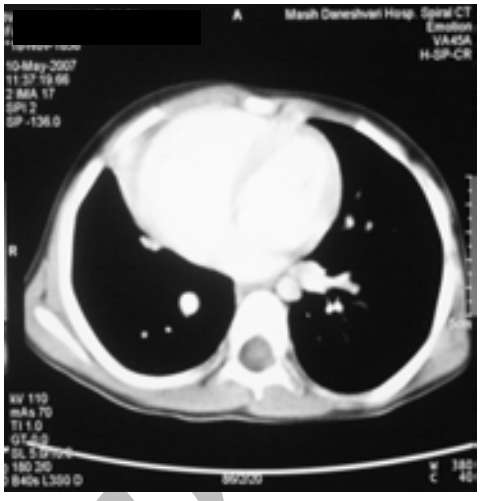

Figure 3. The spiral tomogram of the thorax shows shift of the heart and mediastinum to the right and dextroposition of the heart.

the associated congenital cardiovascular abnormalities is about 36% in the pediatric age group and 75% among the neonates. These abnormalities include atrial septal defect, ventricular septal defect, coarctation of the aorta, abnormalities of the aortic arch, and abnormal relationship of the pulmonary arteries and bronchi. Swyer-James syndrome and lung hypoplasia should be considered in the differential diagnosis of this syndrome.[4]

The defining characteristic of scimitar syndrome is the partial anomalous pulmonary venous return. Usually this anomalous venous return is to the hepatic portion of the inferior vena cava, but it may also be to the portal vein, to a hepatic vein, or to the right atrium. The name comes from this anomalous pulmonary vein, which may be visible on chest radiograph as a curvilinear shadow just above the right diaphragm said to resemble a "scimitar" or Persian sword, “shimshir.”[3]

The characteristic scimitar sign is represented in Figure 1.[1]

The diagnosis is usually established on chest radiograph that shows dextroposition of the heart and hypoplasia of the right lung.[5] According to the different articles CT and magnetic resonance imaging (MRI) show anatomic findings of scimitar syndrome. For detecting cardiac and pulmonary anomalies, CT angiography is recommended.

Scimitar syndrome has a variable

presentation based on the age at which the diagnosis is made. Infants typically present with the main symptoms of severe respiratory insufficiency and cardiac failure.[6] Pulmonary hypertension is a commonly associated problem. In older children and adults the diagnosis of scimitar syndrome is often made incidentally in patiants who undergo chest radiography for diverse reasons.[7] Recurrent respiratory infections and heart murmur may be the mode of presentation.[3]

Our patient was presented with recurrent pulmonary infections without cardiac involvement. Her weight and height were below 5% of growth chart. In this case, the failure to thrive could be explained by recurrent pulmonary infections, which showed improvement in follow-up visits.

The age of patients at the time of diagnosis of scimitar syndrome is usually between neonatal period till adulthood. Our patient was diagnosed at the age of four years. The age at the time of detection of scimitar syndrome and the presence of associated anomalies is important in predicting the outcome. In general, infants presenting with heart failure have more associated anomalies and their prognosis is much worse.[5] The prognosis for older children is better, either with or without surgery. Treatment for symptomatic scimitar syndrome consists of surgical repair. Surgical repair seldom results in normal blood flow to the right lung but abolishes left-to-right shunt. Postoperative pulmonary venous obstruction is prevalent, especially in infants.[8] Therefore, the therapeutic approach for patients with scimitar syndrome, respiratory manifestations, and onset beyond the neonatal period, should be conservative.[6]

In our patient, according to the clinical and radiologic findings, the diagnosis of scimitar syndrome was made. Due to improvement in clinical symptoms of pneumonia, she is receiving supportive care and cardiac follow-up. In case of persistent pulmonary symptoms and failure to thrive, she would be a candidate for surgical treatment.

The triad of respiratory distress, right lung hypoplasia, and dextroposition of the heart should alert the clinician to the possibility of this syndrome.[1]

References

1 Canter CE, Martin TC, Spray TL, Weldon CS, Strauss AW. Scimitar syndrome in childhood. *Am J Cardiol*. 1986; **58:** 652 – 654.

2 Oakley D, Naik D, Verel D, Rajan S. Scimitar vein syndrome: report of nine new cases. *Am Heart J*. 1984; **107:** 596 – 598.

3 Rokade ML, Rananavare RV, Shetty DS, Saifi S. Scimitar syndrome. *Indian J Pediatr*. 2005; **72:** 245 – 247.

4 Gikonyo DK, Tandon R, Lucas RV Jr, Edwards JE. Scimitar syndrome in neonates: report of four cases and review of the literature. *Pediatr Cardiol*. 1986; **6:** 193 – 197.

5 Huddleston CB, Exil V, Canter CE, Mendeloff EN. Scimitar syndrome presenting in infancy. *Ann Thorac Surg*. 1999; **67:** 154 – 159.

6 Lluna-Gonzalez J, Barrios-Fontoba JE, Cavalle-Garrido T, Gutierrez-San-Roman C, Malo-Concepcion P, Carrasco-Moreno JI, et al. Scimitar syndrome: series of 12 cases. *Cir Pediatr*. 1995; **8:** 2 – 6.

7 Gao YA, Burrows PE, Benson LN, Rabinovitch M, Freedom RM. Scimitar syndrome in infancy. *J Am Coll Cardiol*. 1993; **22:** 873 – 882.

8 Najm HK, Williams WG, Coles JG, Rebeyka IM, Freedom RM. Scimitar Syndrome, 20 years experience and results of repair. *J Thorac Cardiovasc Surg*. 1996; **112:** 1161 – 1169.