

Scimitar Syndrome: Original Image

Scimitar Sendromu

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A 6 month-old female infant admitted to our hospital because of respiratory distress and failure to thrive. She was born full term without any complications by spontaneous vaginal delivery, and prenatal history was unremarkable. Physical examination revealed weight 4,3 kg (<3%), temperature 38°C, tachypnea, tachycardia, subcostal retraction, reduced breath sounds in right lung, wheezing and crepitations in both lungs. Her cardiovascular examination revealed regular heart rate and rhythm. Heart murmur was not heard.

A chest radiograph showed the mild right lung hypoplasia, cardiomegaly, but not abnormal shadow (Turkish sword or scimitar sign) due to significant dextroposition of the heart (Figure 1). Echocardiography demonstrated right atrial and ventricular dilatation and small atrial septal defect. Computed tomography scan of the chest revealed dextroposition of the heart, right lung hypoplasia, enlargement of the main pulmonary artery due to partial anomalous pulmonary venous drainage into the inferior vena cava and hypoplastic



FIGURE 1: Chest radiography shows cardiomegaly and dextroposition of the heart.



FIGURE 2: Computed tomography scan of the chest shows hypoplastic right pulmonary artery, hypoplastic right lung, dextroposition of the heart, enlargement of the main pulmonary artery.



FIGURE 3: Computer tomography with bi-dimensional reconstruction shows abnormal pulmonary venous drainage of the right lung into the inferior vena cava (Turkish sword or scimitar sign).

right pulmonary artery (Figure 2). Multi-slice computed tomography with bi-dimensional recon-

struction confirmed the abnormal pulmonary venous drainage from the the right lung through a scimitar vein that drained into the inferior vena cava (Figure 3).

Scimitar syndrome is a rare congenital cardiopulmonary malformation characterized by hypoplasia of lung with hypoplasia of pulmonary artery and drainage of the right pulmonary veins into the inferior vena cava. The look of closely resembles that of a Turkish sword or scimitar.^{1,2} If the patient has recurrent pneumonia and right lung hypoplasia, the clinician should think of the possibility of Scimitar syndrome. Age and clinical presentation vary widely and determined by the degree of pulmonary hypoplasia and presence of significant left to right shunt.³ Furthermore, early diagnosis of Scimitar syndrome is essential because it can cause heart failure and pulmonary hypertension in infancy.

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