SCIMITAR SYNDROME WITH ABSENCE OF THE RIGHT PULMONARY ARTERY*

A Case Report

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SUMMARY: Saltık İL,Eroğlu AG, Öztunç F, Sarıoğlu A. (Division of Pediatric Cardiology, İstanbul University Institute of Cardiology, İstanbul, Turkey). Scimitar syndrome with absence of the right pulmonary artery: a case report. Turk J Pediatr 1999; 41: 399-402.

We report a two-year-old female child with scimitar syndrome associated with absent pulmonary artery but with normal pulmonary artery pressure although several collaterals originated from the abdominal aorta to the right lung. To our knowledge, this is the fifth case with an absent pulmonary artery. Our case also had microphthalmia, an association not previously described in scimitar syndrome.

Key words: scimitar syndrome, right pulmonary artery.

Scimitar syndrome is a congenital anomaly that consists mainly of total or partial anomalous venous drainage of the right lung to the inferior vena cava. It is often associated with hypoplasia of the right lung, dextroposition of the heart, anomalous systemic vascular supply of the right lung from the aorta and bronchial anomalies. The name "scimitar" is derived from the radiological shadow of the abnormal right-sided pulmonary vein along the right border of the heart, resembling a Turkish sword. In about half of the cases, the size of the right pulmonary artery is minimally reduced, but in some cases the reduction can be severe. Only four cases have as yet been described in which the right pulmonary artery is absent[1-3]. All these patients had pulmonary hypertension.

Additional forms of cardiovascular abnormalities have been described in approximately 25 percent of cases with scimitar syndrome. A variety of extracardiac abnormalities have been frequently associated with the syndrome.

We present a case in which the right pulmonary artery was absent, but in whom there was a normal pulmonary artery pressure although several collaterals originated from the abdominal aorta to the right lung. Our case also had microphthalmia, an association not previously described in scimitar syndrome.

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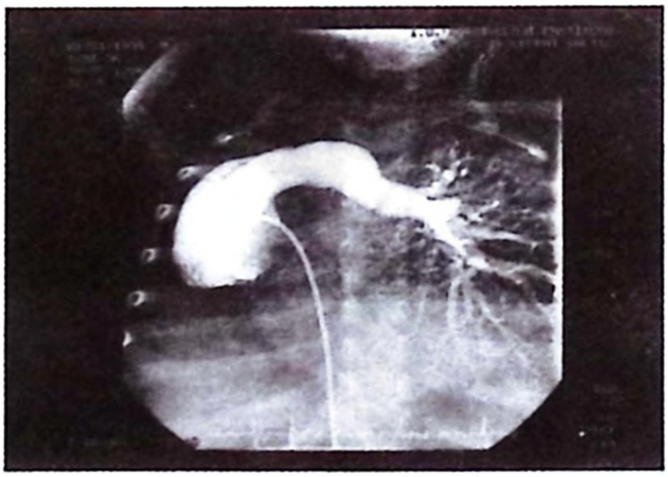
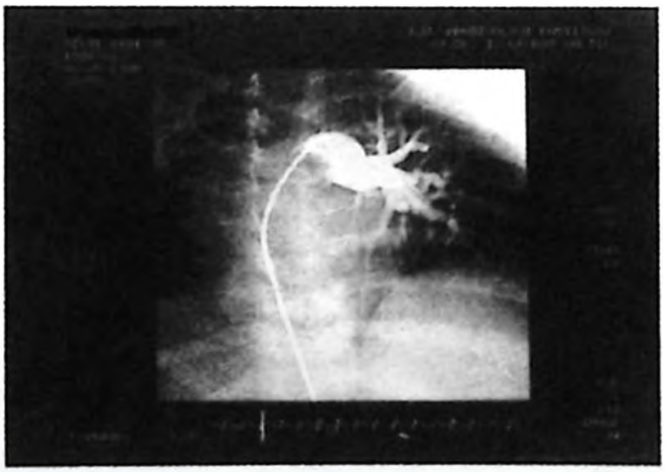
Case Report

A two-year-old girl presented with a past history of frequent respiratory infections. The physical examination was normal except for microphthalmia of the left eye and palpable cardiac activity which could be felt best over the right chest. Chest roentgenogram showed hypoplasia of the right lung with dextroposition of the heart and mediastinal shift to the right. The electrocardiogram was normal. The echocardiogram was normal except for an undetectable right pulmonary artery and dextroposition of the heart. Computerized tomography scans showed dextroposition of the heart, hypoplasia of the right lung and absence of the right pulmonary artery. Cardiac catheterization revealed dextroposition of the heart and normal right ventricular and pulmonary arterial pressures (30/0 mmHg and 30/10 mmHg, respectively). There was no right pulmonary artery (Fig. 1) and arterial supply to the right lung was via several collateral vessels from the abdominal aorta (Fig. 2). Venous drainage of the right lung was to the inferior vena cava.



(a)



(b)

Fig. 1a: Anteroposterior projection of right ventricle angiogram and b: 30° left anterior oblique and 20° cranial projection of pulmonary angiogram demonstrate large main and left pulmonary arteries and absent right pulmonary atery.

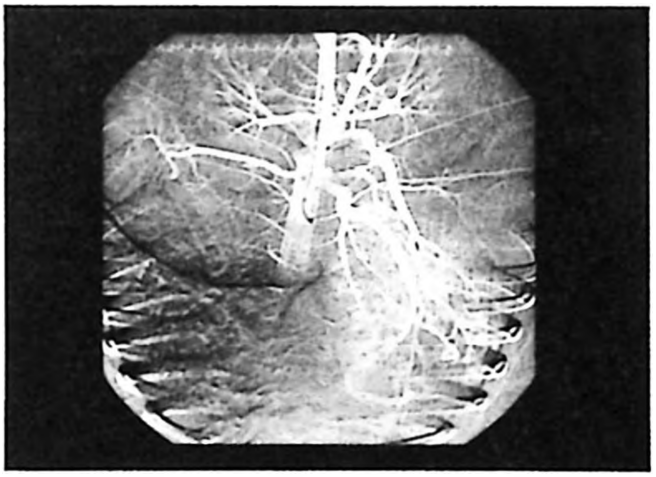


Fig. 2: Anteroposterior projection of aortogram demonstrates systemic arteries supplying the right lung.

Because of the relative well-being and young age of the child, surgical intervention was not planned for the near future. Medical treatment consisted of daily chest physiotherapy and early oral antibiotic therapy at the first signs of respiratory infections. At follow-up 36 months later, the child remained asymptomatic.

Discussion

Patients with scimitar syndrome are typically asymptomatic and thus, the actual prevalence of this rare syndrome is difficult to determine. The diagnosis is usually suspected as a result of incidental discovery of the scimitar vein on chest radiograph. Symptoms, when present, most often include the following: recurrent pulmonary infection secondary to alteration of the tracheobronchial architecture, congestive heart failure due to left-to-right shunt and associated cardiac malformations[1,4-6]. A small subset of patients, however, present with cardiac failure early in infancy[1,5,6]. All of them have extreme hypoplasia of the right pulmonary artery and a large left-to-right shunt from systemic collaterals via the right lung to the right atrium. Cardiac catheterization demonstrates pulmonary hypertension in most of them. Absence of the right pulmonary artery in scimitar syndrome has previously been described in only four cases[1-3]. All these patients had pulmonary hypertension and were critically ill.

The mechanisms for pulmonary hypertension in patients with scimitar syndrome have not been clearly delineated; there are several hypotheses[1,5-7]. It has been proposed that, in scimitar syndrome with extreme hypoplastic or even absent pulmonary arteries and large systemic collaterals, the left lung has to handle both systemic venous return and shunt volume and probably reacts abnormally to this increased volume load[1]. Absence of the right pulmonary artery without pulmonary hypertension despite several collaterals from the abdominal aorta,

as in our case, does not support tihs hypothesis. Another theory is that a reduction of the pumonary vascular bed may induce a persistence of fetal circulation[1]. More studies will be needed in order to detect the true origin of pulmonary hypertension.

Cardiovascular abnormalities, most commonly atrial septal defect, ventricular septal defect, coarctation of the aorta, patent ductus arteriosus, tetralogy of Fallot, atrioventricular septal defect, double outlet right ventricle, hypoplastic left heart syndrome and pulmonary valvular stenosis, may be present as well. Additional extracardiac anomalies that may coexist include accessory or eventrated diaphragm[5], horseshoe lung[5,8], hemivertebrae[8], scoliosis[6,8], meningomyelocele[5], double ureter[8], duplication of the uterus[2], Hirschsprung's disease[8], cleft palate[8] and hypospadias[8]. Our case had microphthalmia, an association not previously decribed in scimitar syndrome.

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