Scimitar Syndrome and Associated Pulmonary Sequestration: Report of a Successfully Corrected Case

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Introduction

The scimitar syndrome is characterized by anomalous venous drainage from the right lung to the vena cava inferior. The appearance of this vein on chest X-ray is suggestive of a Turkish sword and is called the “scimitar” sign by Neill et al. (1960). The condition may be associated with anomalies of the right lung (absence of lung fissures, mirror image of the left lung, absence of lobe bronchi), anomalies of the pulmonary artery, and congenital heart defects (Mathey et al. 1968; Mende et al. 1973). The symptoms are similar to those of an atrial septal defect due to a left-to-right shunt, such as repeated respiratory infections. We present an uncommon case of scimitar syndrome combined with pulmonary sequestration; few cases of this anomaly have been reported in the literature (Alivizatos et al. 1985; Ohsaki 1983).

Case Report

A 6-month-old boy was transferred from Libya to our clinic because of recurrent pulmonary infections and effort dyspnoea. Chest X-ray led us to suspect atelectasis of the right lower lobe; a scimitar vein was not recognized at this point (Fig. 1). Bronchoscopy showed a normal bronchial tree without obstruction, but the middle lobe bronchus could not be identified. In echocardiography dilatation of both ventricles was found but no atrial or ventricular septum defect, and valve function was normal. The pulmonary veins were recognized only on the left side. Cardiac catheterization demonstrated a left-to-right shunt of 23% and atypical branching of the right pulmonary artery with absence of a pulmonary arterial supply to the right lower lobe (Fig. 2).

The venous phase of the pulmonary angiogram showed an atypical common pulmonary vein descending along the right side of the heart and draining nearly all the blood from the right lung into the vena cava inferior. Only a minor lung vein from the upper lobe seemed to drain into the left atrium (Fig. 3). The aortogram showed two systemic arteries arising from the abdominal aorta, supplying the right lower lobe of the lung (Fig. 4). The diagnosis of a scimitar vein and pulmonary sequestration was made. The pressure in the pulmonary artery was 35/12/
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Fig. 1. On the chest X-ray the scimitar vein is hardly distinguishable from the right cardiac border owing to dextrocardia.

Fig. 2. Pulmonary arteriogram showing absence of pulmonary artery supply to the right lower lobe.

Fig. 3. Venous phase of the pulmonary arteriogram showing the scimitar vein along the right cardiac border and a tortuous vein entering the left atrium.

18 mmHg, and systemic pressure was 75/30/50 mmHg. Figure 5 shows the entire anomaly.

At operation the right pleural space was found to be partly obliterated by adhesions. The right lung showed no interlobar segmentation. As seen in the angiogram, a large anomalous venous trunk descended in a sulcus of the lower lobe along the right heart and drained the veins from the entire right lung into the