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Embolization of the Systemic Arterial Supply via a Detachable Silicon Balloon in a Child with Scimitar Syndrome

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Abstract

Scimitar syndrome is a rare congenital disorder. It is characterized by partial or total abnormal venous drainage of the right lung into the inferior vena cava, which is often associated with anomalous systemic arterial supply to the right lung, congenital cardiac anomalies, hypoplasia of the right lung and bronchial anomalies. Symptoms depend on the degree of the shunt and severity of the associated anomalies, which determine the treatment. We present a 6-year-old boy who was diagnosed as having the adult form of scimitar syndrome during evaluation for recurrent pulmonary infections, and underwent embolization with a detachable silicon balloon of the anomalous systemic arterial supply from the abdominal aorta to the right lower lung lobe. Successful elective surgery was performed 6 months later, in which right pulmonary veins were directed to the left atrium using a Gore-Tex patch by creating an intra-atrial tunnel. The patient has been symptom-free period during 6 months of follow-up, which supports the idea that recurrent pulmonary infections can be eliminated by embolization of the anomalous arterial supply.

Key words: Scimitar syndrome—Embolization—Silicon balloon—Anomalous arterial supply—Pulmonary infection

Scimitar syndrome is a rare congenital disorder characterized by partial or total abnormal venous drainage of the right lung into the inferior vena cava. This malformation is often associated with anomalous systemic arterial supply to the right lung from the abdominal aorta, which may sometimes represent lung sequestration and is one cause of recurrent respiratory infections in patients with scimitar syndrome [1, 2]. Other congenital cardiac anomalies, especially septal defects, hypoplasia of the right lung, dextroposition of the heart and bronchial anomalies, may be associated in complex forms of the syndrome [1–3].

We report the embolization of the anomalous systemic arterial supply to the right lower lung lobe by a detachable silicon balloon in a 6-year-old boy with scimitar syndrome who was diagnosed during evaluation for recurrent pulmonary infections.

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Case Report

A 6-year-old boy was admitted to hospital with a history of recurrent respiratory infections. All physical findings were normal except for a grade 1/6 systolic ejection murmur at the left upper sternal border and slightly diminished lung sounds at the right lower region. The chest radiograph revealed an abnormal shadow along the right heart border (scimitar sign) (Fig. 1). To rule out any mass or infection, a computed tomography (CT) scan of the chest was performed which revealed the shadow as a large (12 mm) abnormal vascular structure at the lower lobe of the right lung; no consolidation signs or mass were detected. To delineate the vascular anatomy, contrast-enhanced magnetic resonance angiography (Ce-MRA) was performed. A four-phase dynamic breath-hold fast SPGR study was performed with a 1.5 T MR imaging unit (Echo speed; GE Medical Systems, Milwaukee, WI, USA) by placing the smart prep tracer in the right ventricle. Ce-MRA demonstrated an anomalous right pulmonary vein draining into the inferior vena cava and an abnormal arterial branch from the abdominal aorta (Fig. 2). Echocardiography showed slightly dilated right heart chambers and the presence of the abnormal pulmonary vein draining into the inferior vena cava below the diaphragm, but no associated intra-cardiac anomalies (Fig. 3).

Cardiac catheterization and angiocardiography was performed to delineate the exact anatomy and to detect the magnitude of the left-to-right shunt and pulmonary artery pressure. It revealed hypoplasia of the right inferior pulmonary artery, total abnormal pulmonary venous return of the right pulmonary veins into the inferior vena cava (Fig. 4A, B) and an aortopulmonary collateral vessel, 8 mm in diameter, originating from the abdominal aorta supplying the lower lobe of the right lung. The arterial supply from the abdominal aorta originated as a single main artery dividing into branches before crossing the diaphragm (Fig. 4C). The pulmonary artery pressure was 28/12 (mean 17) mmHg, aortic pressure was 105/75 (mean 90) mmHg, and the pressure in the aortopulmonary collateral was equal to that in the aorta. Oxygen saturation was 94% in the inferior vena cava, 80% in the superior vena cava, 92% in the right atrium, and 91% in the right ventricle and pulmonary artery. Simultaneous aortic saturation was 98% and the right anomalous pulmonary vein saturation was 99%. The pulmonary-to-systemic flow ratio was calculated as 1.91. No associated extra cardiovascular anomaly was detected.

Because of the recurrent pulmonary infections, selective embolization of the main collateral artery from the abdominal aorta to the lower lobe of the right lung was planned. After obtaining an abdominal aortogram, a 7 Fr guiding catheter was advanced into the anomalous artery and a test occlusion balloon (Boston Scientific, USA) (TOB) was introduced through the guiding catheter. The TOB was inflated to check whether the main artery could totally be occluded and to find out whether any other branches were present that supplied the right lower lobe. After confirmation of the absence of any other abnormal vascular supply to the right

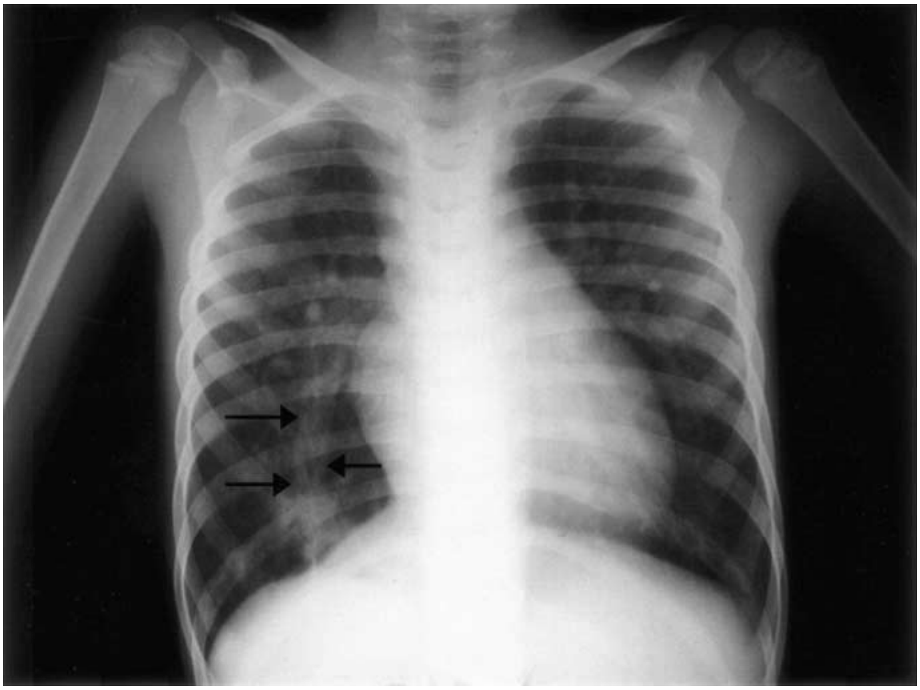

Fig. 1. Chest radiograph showing the scimitar sign along the right heart border (arrows).

Fig. 2. Magnetic resonance angiography demonstrating an anomalous right pulmonary vein draining into the inferior vena cava (arrows) and an abnormal arterial branch from the abdominal aorta (arrowheads).

lower lobe and also the total occlusion of the main artery, a 9.9 × 24.5 mm detachable silicon balloon (Boston Scientific, USA) (DSB) was introduced through the guiding catheter following the withdrawal of the TOB. The DSB was inflated with a 50%/50% mixture of contrast medium and saline solution at the proximal part of the anomalous main artery and was released after confirmation of its position and the total occlusion of the anomalous main artery by contrast injection through the guiding catheter. No passage of contrast was seen beyond the DSB after the

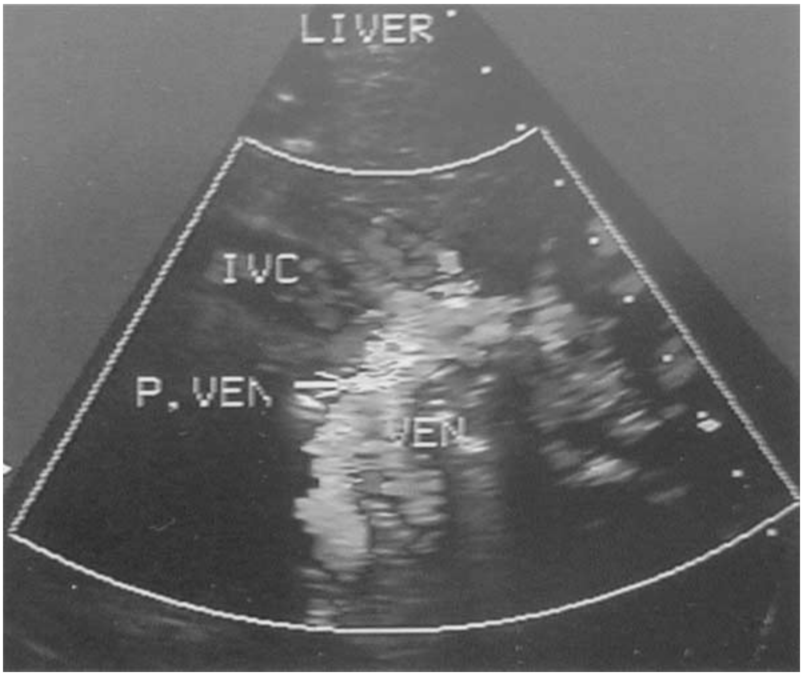

Fig. 3. Echocardiography demonstrating the abnormal pulmonary vein draining into the inferior vena cava below the diaphragm.

procedure (Fig. 5). No significant pressure differences were detected in the pulmonary artery.

Elective repair of the anomalous pulmonary venous return of the right lung was scheduled. Successful surgical repair was performed 6 months after the embolization of the anomalous systemic arterial supply to the right lower lung lobe, in which right pulmonary veins were directed to the left atrium using a Gore-Tex patch by creating an intra-atrial tunnel. While waiting for surgery the patient did not have any respiratory infections.

Discussion

Scimitar syndrome is a rare, complex congenital malformation with variable presentation. The main feature is the abnormal pulmonary venous drainage into the inferior vena cava. It was given its name because a crescentic shadow of vascular density along the right border of the cardiac silhouette that is characteristic of the syndrome resembles a Turkish sword on chest radiography, but it is not seen in all patients [4–6]. The syndrome is also frequently characterized by varying degrees of hypoplasia of the right pulmonary artery and is associated with right lung bronchial anomalies. Dextroposition is often present as a result of the hypoplasia of the right lung and the anomalous systemic arterial supply to the right lower lobe from the subdiaphragmatic aorta or its main branches [7–9]. Approximately 25–30% of patients have associated congenital heart diseases, especially atrial septal defects [3, 8, 9]. About one-third of cases appear in infancy and childhood. Clinical manifestations, which are usually not prominent, and the prognosis depend on the severity of the pulmonary hypoplasia, the presence and severity of the cardiac abnormality and the degree of pulmonary artery pressure [8–10]. Three types of the syndrome have been described: (1) The *adult form*, characterized by a small shunt from right pulmonary veins to the inferior vena cava and intracardiac defects. This is usually well tolerated and has a good prognosis. (2) The *infantile form*, characterized by a large shunt from abnormal systemic arterial supply to the lower part of the right lung. This is symptomatic in early infancy with respiratory distress and heart failure, and has a poor prognosis. (3) A third form characterized by predominant multiple cardiac and extracardiac malformations. In such cases scimitar syndrome is of minor concern [11]. Although cardiac catheterization and angiography is the method for determination of cardiac anomalies and quantitative measurements of the left-to-right shunting, velocity-encoded cine MR (VEC-MR) may be used as a noninvasive method to determine the congenital heart disease and quantification of shunting [12].

The term ‘‘adult form’’ is debatable; it merely means that the patient is asymptomatic in infancy, as in our case. It is detected either incidentally or when a child is evaluated for dyspnea or recurrent pulmonary infections [8, 9]. In our case, recurrent pulmonary infections were the only complaint and there was no associated intracardiac anomaly.

Treatment of scimitar syndrome is controversial. There is no consensus on the treatment procedure because the syndrome is too rare for pediatric cardiologists, cardiovascular surgeons or radiologists to be familiar with it [1]. An indication for surgery is the presence of associated severe cardiac malformation [5]. For the infantile form of scimitar syndrome associated with an abnormal systemic arterial supply to the lower part of the right lung and pulmonary hypertension, a two-stage approach has been suggested: initially, ligation of the systemic arteries with or without lobectomy, and then rerouting of the abnormal pulmonary venous return of the right lung and, if present, correction of the intracardiac defects later [2, 13]. Recently, coil embolization of the anomalous systemic arterial supply has been reported (a) as an alternative to surgical ligation in symptomatic infants before cardiac surgery [2, 7, 14], or (b) to control infection in patients with pulmonary sequestration [15]. If coil embolization is preferred, coils made of titanium or platinum should be used as both are excellent for registering future MR images. Although most patients with the adult form of scimitar syndrome have not undergone surgery, some authors suggest that surgical correction, rerouting the pulmonary venous flow to the left atrium, should be performed if the left-to-right shunt is significant (Qp/Qs greater than 1.5) [8, 9]. Additionally some patients with recurrent pulmonary infections or hemoptysis may require resection

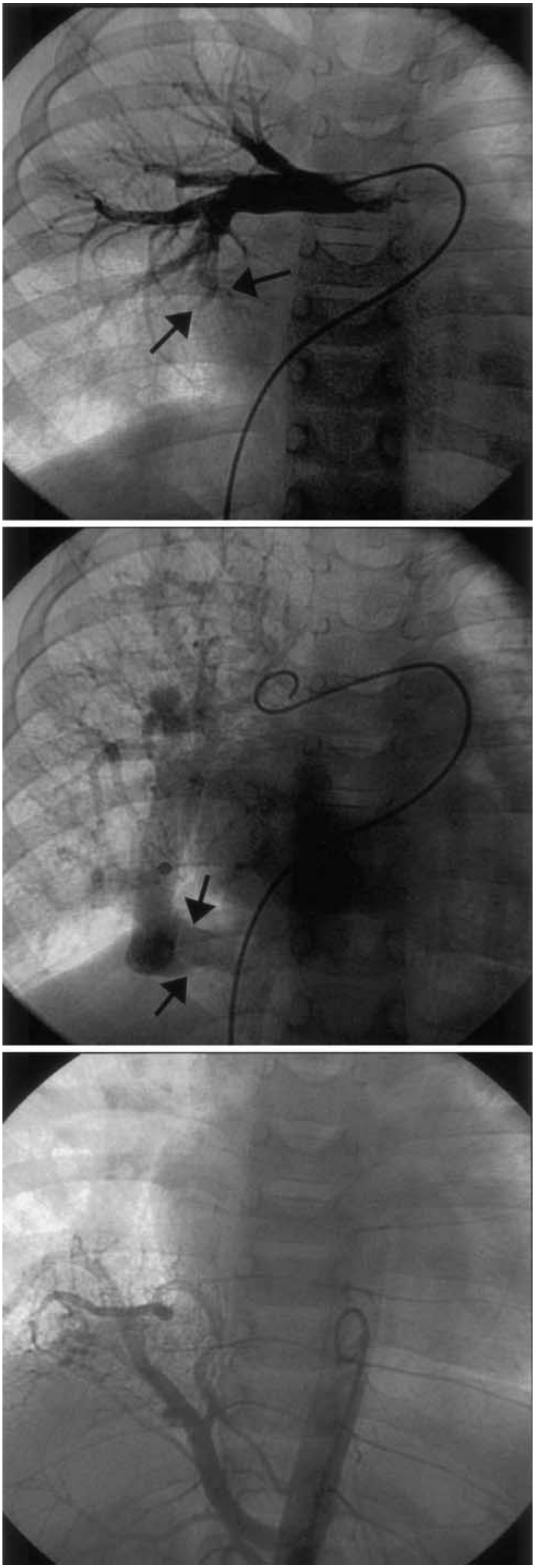

Fig. 4. Pulmonary angiography demonstrating **A** hypoplasia of the right inferior pulmonary artery (arrows) and **B** total abnormal pulmonary venous return of the right pulmonary veins (arrows) into the inferior vena cava below the diaphragm. **C** Abdominal aortography demonstrating the abnormal arterial supply from the abdominal aorta originating as a single main artery dividing into branches before crossing the diaphragm.

of the lower part of the lung supplied by the abnormal systemic arterial vessel. Also lobectomy or pneumonectomy may be necessary during surgery in case of either difficulty mobilizing the anomalous pulmonary vein or postoperative complications, mostly thrombosis of the anastomosis [1, 2, 5, 8]. In the multicenter study reported by Dupius et al., long-term follow-up of the adult form of scimitar syndrome without pulmonary hypertension showed that this condition was usually well tolerated [1]. The main indication for surgery was the elimination of the shunt. Because of recurrent pulmonary infections and significant left-to-right shunt a two-stage approach was planned in our case.

As the first stage, we preferred DSB occlusion of the systemic arterial supply instead of surgical ligation. A single DSB was placed into the proximal portion of the arterial collateral before branching to avoid multiple embolizations of the distal branches. Coil embolization may be performed for each anomalous arterial branch with one or more coils, followed by embolization of the main anomalous arterial branch. The embolization of the main branch may be performed by forming an endoskeleton with high-radial-force coils, followed by many platinum-fibered coils to achieve cross-sectional occlusion. In such a procedure the number of coils that will be required can not be ascertained, and additional liquid embolization agents, such as Histoacryl or Avacryl, may be necessary in case of insufficient occlusion. In our case, by occluding the anomalous main systemic artery with a DSB before branching, the time and cost of the procedure were reduced. It was more cost-effective than coil occlusion of the branches. In our case no pulmonary infection has been observed during a 6 month follow-up period. We believe that recurrent pulmonary infections can be eliminated by embolization of the anomalous arterial supply. Also this approach may help to reduce the complication rate and shorten the time needed for surgical repair of the anomalous pulmonary venous return.

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Fig. 5. Aortography after occlusion shows **A** position of the silicon balloon and **B** no passage of contrast to the distal part of the DSB (arrows) in the anomalous branches.

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