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Isolated Left-sided Scimitar Vein Connecting All Left Pulmonary Veins to the Right Inferior Vena Cava

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Abstract. When the common pulmonary vein fails to develop, the embryonic connections of the pulmonary veins to one or more of the systemic veins almost always persist. Anomalous pulmonary venous connections to the inferior vena cava (IVC) are typically characterized by hypoplasia of the involved pulmonary veins and pulmonary artery, as well as abnormal parenchyma of the involved lung. Such cases have been described as "scimitar syndrome." We report the case of a young female patient in whom all the left pulmonary veins converged into a common vessel that drained into the IVC but who had a normal left pulmonary artery and left lung. Surgical intervention was successful, and our patient is still alive.

Keywords: Congenital heart disease — Pulmonary vein anomaly — Scimitar vein — Inferior vena cava

The scimitar syndrome [1] typically is characterized by anomalous right pulmonary venous drainage into the inferior vena cava (IVC), anomalous right pulmonary arterial supply from the abdominal aorta, hypoplasia and dysplasia of the right lung. hypoplasia of the right pulmonary artery, and secondary mesocardia or dextrocardia. Scimitar syndrome gets its name from the chest radiograph appearance of the curvilinear anomalous pulmonary vein that casts a shadow resembling a curved sword or scimitar. We report an exceedingly rare and clinically important variant in a female patient who had a left-sided scimitar vein that drained into a right-sided IVC, but without any other features of the scimitar syndrome.

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Case Report

At the age of 3 years, this well-developed young girl had been found to have a murmur and mild intolerance to exercise. Cardiology evaluation at age 9 in 1979 included physical examination, chest radiograph, electrocardiogram, and echocardiogram and revealed an ostium secundum atrial septal defect (ASD). She returned at the age of 10 years for elective surgical closure of the ASD. Her initial postoperative course was complicated by chest bleeding and bilateral pleural effusions. She then developed post-pericardiotomy syndrome that resolved with antiinflammatory medication. Postoperative echocardiogram at 1 year after the operation showed showed complete closure of the ASD.

She was well until the age of 22 years when she developed mildly decreased exercise tolerance. A chest radiograph showed mild cardiomegaly. Cardiac catheterization at a referring institution revealed a significant atrial level shunt. It was presumed that the surgical suturing of the ASD had dehisced. Subsequent evaluation by echocardiography at our institution showed that the atrial septum was intact. It was also found that all the left pulmonary veins joined together in a confluence behind the left atrium and then drained below the diaphragm into the right-sided inferior vena cava (IVC) (Fig. 1A and 1B). There was also marked right ventricular dilatation. Cardiac catheterization and angiocardiography confirmed this diagnosis and also revealed a right upper pulmonary vein draining anomalously into the cranial portion of the right superior vena cava (SVC).

Soon after catheterization, she underwent a surgical repair consisting of side-to-side anastomosis of the left pulmonary venous confluence to the left atrium with ligation and division of the distal portion of the anomalous pulmonary venous connection. Her recovery was uneventful.

Cardiac MRI 6 years after the anastomosis of the left pulmonary veins to the left atrium demonstrated wide patency of the anastomosis of the left pulmonary venous confluence to the left atrium (Fig. 1C). The MRI also showed the uncorrected right upper pulmonary vein draining into the SVC. There was mild right ventricular dilatation and normal ventricular function. Ten years after her second operation, she remains asymptomatic.

Discussion

Despite the presence of an abnormal venous confluence draining the entire left lung to the right

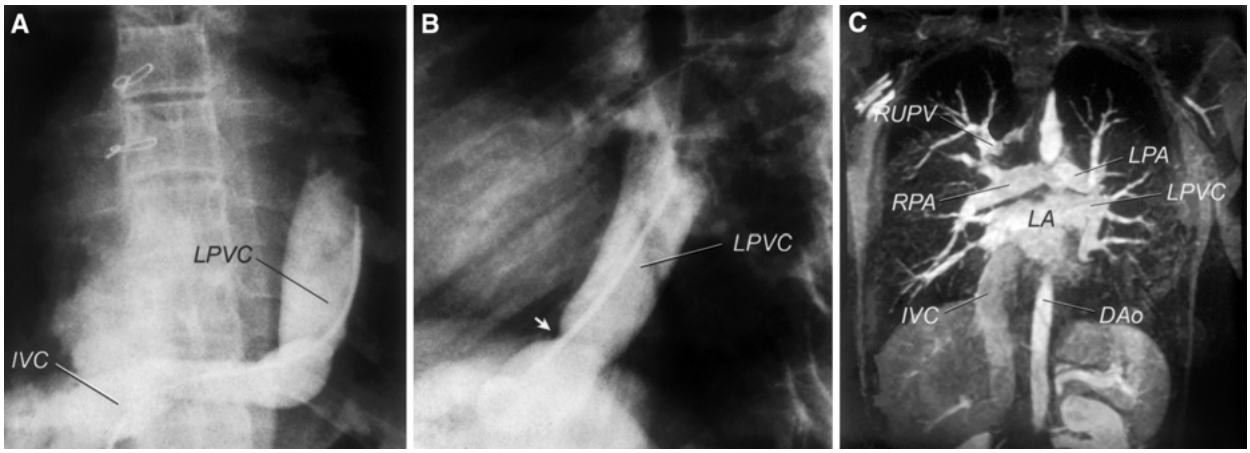

Fig. 1. **A** Anteroposterior projection of selective injection of anomalously connected left pulmonary veins. **B** Lateral projection of same injection showing mild constriction at the level of the diaphragm (*arrow*). *IVC*, inferior vena cava; *LPVC*, left pulmonary venous confluence. **C** Gadolinium-enhanced MR angiogram showing normal left pulmonary artery (*LPA*), normal left lung, and patency of the surgical anastomosis between the left pulmonary venous confluence (*LPVC*) and the left atrium (*LA*). *DAo*, descending aorta; *IVC*, inferior vena cava; *RPA*, right pulmonary artery; *RUPV*, right upper pulmonary vein.

IVC—i.e., the presence of a scimitar vein—this case did not have the other characteristics typical of scimitar syndrome [1]. The ipsilateral lung was not abnormal. The other vascular malformations usually observed in the complete forms of scimitar syndrome—namely, hypoplasia of the ipsilateral pulmonary artery and an anomalous arterial supply of the involved lung by a systemic artery originating below the diaphragm—were also absent. In addition, the patient was free of symptoms indicative of lung dysfunction or significant exercise intolerance while half of the pulmonary venous return was entering the IVC.

To our knowledge, isolated left-sided scimitar vein has not been previously reported in the English-language literature. This case underscores the need for careful assessment of the pulmonary venous connections in all patients with secundum ASD.

References

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