

Scimitar Vein Anomaly with Total Right-Side Pulmonary Vein Return to Inferior Vena Cava Concomitant with Atresia of Upper Right-Side Pulmonary Vein and Secundum Atrial Septal Defect

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ABSTRACT

Scimitar vein is the partial anomalous pulmonary venous connection usually draining lower pulmonary lobe to the inferior vena cava or right atrium. We present a scimitar vein anomaly in a 34-year-old woman with the uncommon association of a secundum type of atrial septal defect and atretic right upper pulmonary vein. She presented with increasing dyspnea for 2 months, however, she was asymptomatic in the past 10 years despite the presence of atrial septal defect. We describe its diagnostic and surgical approach.

INTRODUCTION

Partial anomalous pulmonary venous return (PAPVR) is an uncommon congenital anomaly with an occurrence rate of 0.5%-1% of congenital disease [Kamler 2003]. PAPVR of part of or the entire right lung to the inferior vena cava (IVC) or the right atrium (RA) as scimitar vein (SV) is the main abnormality of the scimitar syndrome (SS) together with hypoplasia of the right lung and dextroposition of the heart [Casha 2003; Juraszek 2005; Lee 2007; Ruggieri 2003]. Also, other cardiac abnormalities such as atrial septal defect (ASD) can occur with PAPVR [Kamler 2003; Brown 2003]. In this case, we describe the PAPVC of the right lung to IVC with atresia of the right upper pulmonary vein and a secundum type ASD.

CASE PRESENTATION

A 34-year-old woman, with a 10-year history of ASD, was admitted with increasing dyspnea over the course of 2 months. In the primary evaluation we found high jugular vein pressure, normal S1, loud S2 and left side border systolic murmur II/VI. Chest x-ray showed cardiomegaly and prominent pulmonary artery. Transthoracic echocardiography

(TTE) revealed normal sized left ventricle with good function, severe right ventricle enlargement with good function, bicuspid aortic valve, mild tricuspid regurgitation, mild pulmonary hypertension (PHT) with systolic pulmonary artery pressures (PAP) estimated about 38 mm hg and evidence of left to right shunt through interatrial septum in doppler study with (Qp: Qs = 2.2/L). Transesophageal echocardiography (TEE) confirmed left to right shunt with large secundum type ASD (Figure 1), and abnormal drainage of right pulmonary vein. Since the exact abnormal drainage site of PAPVC was not found and the only abnormal turbulent flow was in IVC, a CT angiography was performed for her which revealed shunt related pulmonary hypertension, abnormal drainage of right lower pulmonary vein into IVC (Figure 2) and atretic right upper pulmonary vein (total right lung blood drainage to IVC) (Figure 3). Oximetry during cardiac catheterization confirmed abnormal left to right drainage into IVC. Oxygen saturations in the RA were stepped up to 90%, while those in the IVC below the kidney were 80%, which increased to 85% and 88% near the entrance into the RA. Surgical approach was considered for the patient. Through midsternotomy approach, cardiopulmonary

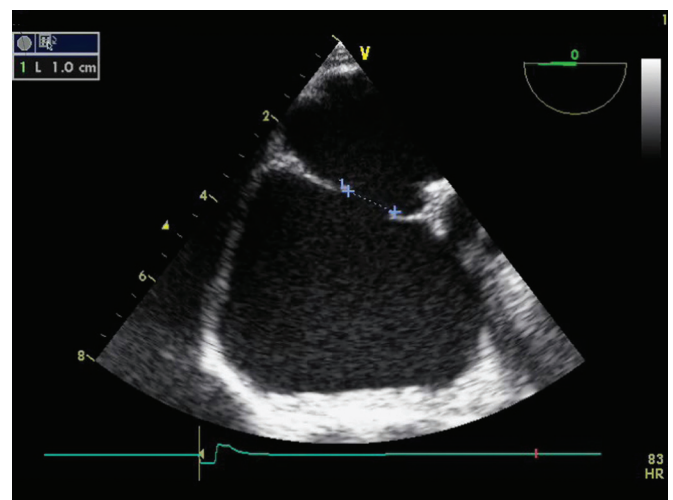


Figure 1. Transesophageal echocardiography; (0°, ME view) atrial septal defect (ASD) secundum.

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bypass (CPB) initiated by cannulation of ascending aorta and superior vena cava also left femoral vein for drainage of IVC. The patient was cooled to 18°C. After cardiac arrest, RA was opened and ASD enlarged by a total resection of fossa ovalis (FO). Hypothermic total circulatory arrest (TCA) was established and the site of abnormal vein drainage, that was just

below of the diaphragm, exposed. The entrance site baffled by a large fresh pericardial patch as described by Kouchoukos et al [2003] to the ASD. The patient's IVC and lower RA were repaired by another patch to prevent stenosis of IVC orifice. CPB was then established and the patient was weaned uneventfully. Intraoperative TEE showed no shunt or abnormal flow after correction. Patient was discharged on postoperative day 7 on warfarin and low dose diuretics.

Two weeks later, she was readmitted with increasing dyspnea over the course of 2 days. Lung perfusion scan showed no evidence of pulmonary emboli, and echocardiography revealed massive pericardial effusion. She underwent surgical pericardial window. During past two months she has remained in stable condition.

DISCUSSION

PHT is common in infantile form of SS [Dupuis 1993]. In the adult form of this disease, pulmonary artery pressures are normal in 77% of patients and may elevate slightly in the remaining 23% [Dupuis 1992]. Left-to-right shunt of more than 50% is able to induce dyspnea, chest infections and PHT in these patients [Walles 2002]. Association of SS with PHT and ASD is one of the indications for surgical repair [Brown 2003] and in compliance to this evidence, surgical treatment was considered for our patient. Since 1950, when the first surgery for repair of SS was performed, several operative approaches have been performed and classic baffling is one of the most popular of them. Classic baffling entails construction of a long intra-atrial baffle from the entry point of the scimitar vein into the inferior vena cava

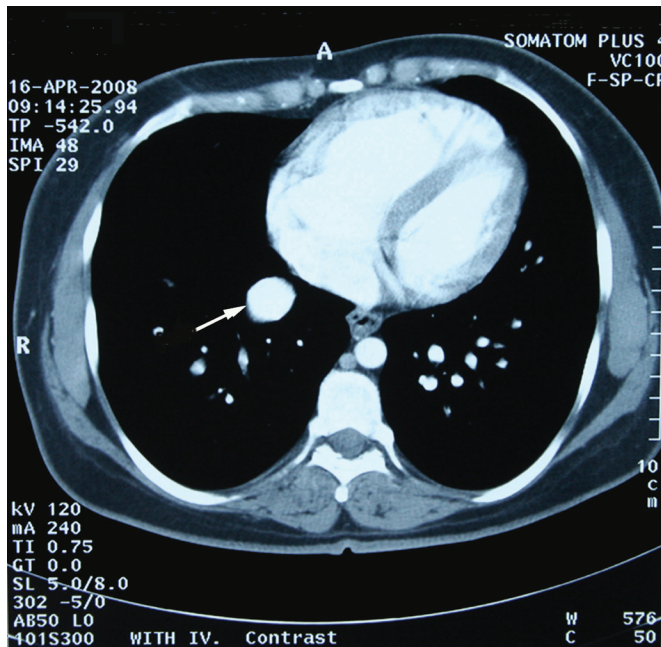
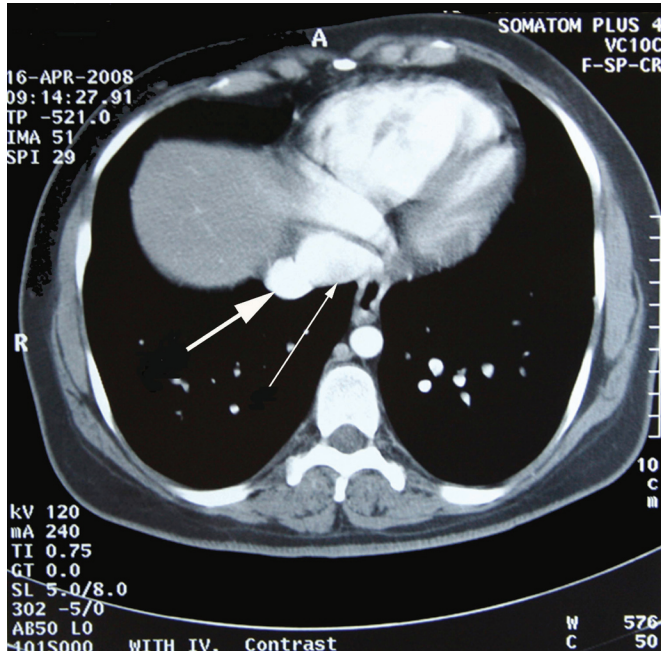


Figure 2. A, Axial image of thoracic computed tomography (CT) angiography shows dilated right inferior pulmonary vein (thick arrow) that merges with inferior vena cava (IVC) (thin arrow). B, Diaphragm level of CT angiography reveals dilated right cardiac chambers due to long-standing atrial septal defect (ASD).

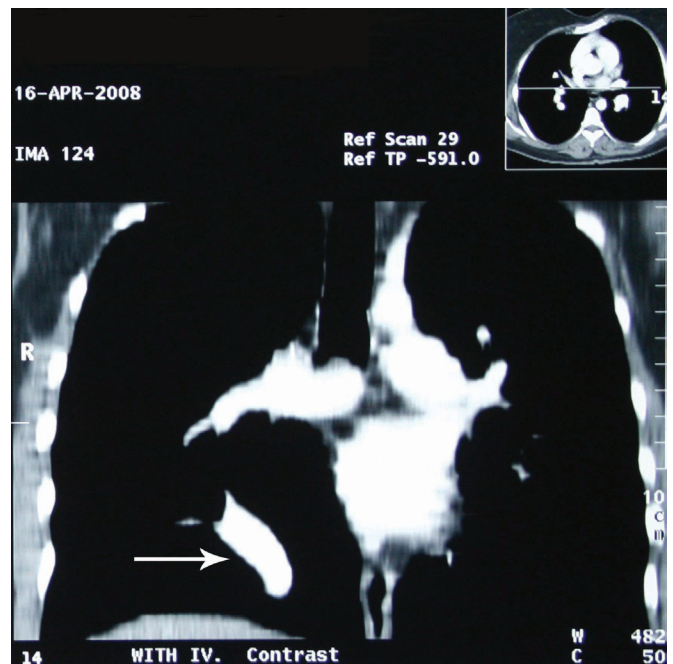


Figure 3. Coronal reconstructed image of the abnormal scimitar vein (arrow). The right upper pulmonary vein is absent.

to the left atrium through an ASD [Brown 2003]. There are certain other techniques for correction of SV. In one alternative approach, the anomalous pulmonary vein can be disconnect from IVC and reimplanted higher on RA with connection via a baffle to left atrium. In another approach, the anomalous vein can be implanted on to LA wall directly or by an interposition tube graft [Kouchoukos 2003]. On the other hand, surgical repair can be performed entirely during TCA, like in the case of small infants, or it can be performed partly during TCA in older patients [Kouchoukos 2003]. In brief, surgical approaches to SV have varied according to the patients' age and anomaly characterization in each case. We believe classic baffling using a fresh pericardial patch is still one of the best repairment approaches for this type of SV.

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