Medical Image of the Week: Scimitar Syndrome

Figure 1. Axial CT in lung windows at the level of the right atrium shows a dilated anomalous vein (arrow) coursing in close proximity to the major fissure (star).

Figure 2. Coronal CT in soft tissue windows at the level of the right hemi diaphragm shows evidence of post surgical repair with the anomalous vein draining into the left atrium (arrow). Note the right hemi-diaphragm is elevated suggesting some degree of pulmonary hypoplasia.
A 38 year-old woman presented for evaluation of palpitations and chronic progressive dyspnea on exertion accompanied by chest tightness and fatigue. Chest radiograph was normal except for low lung volumes. An echocardiogram revealed normal left ventricular size and function with an ejection fraction of 60%, normal right ventricular size and function and moderate tricuspid insufficiency with an estimated right ventricular systolic pressure of 36 mm Hg plus central venous pressure and a mildly enlarged right atrium. Computed tomography (CT) of the heart with contrast showed normal coronary arteries, enlarged right atrium and partial anomalous pulmonary venous return from the right lung to the inferior vena cava (IVC) (Figure 1). Cardiac catheterization with selective angiography confirmed anomalous pulmonary venous drainage from the right upper and right lower lobe to the hepatic portion of the inferior vena cava with obstruction (8 mm Hg gradient between the anomalous vein and the right atrium). The calculated pulmonary to systemic flow ratio (Qp:Qs) was 1.3:1. The pulmonary vascular resistance was 7.6 Woods units and the mean pulmonary artery pressure was 24 mmHg. The diagnosis of Scimitar syndrome was made.

The patient underwent surgical repair of partial anomalous pulmonary drainage and pulmonary vein stenosis as well as ligation of an aortopulmonary collateral artery found intraoperatively. The anomalous pulmonary vein was divided from the IVC, the caval end was oversewn and the anomalous vein was anastomosed to the left atrium with a CorMatrix patch (Figure 2). The patient recovered from surgery, however, her symptoms continued and Doppler pattern on a follow-up transthoracic echocardiogram suggested residual obstruction at the site of anastomosis to the left atrium. Catheterization confirmed obstruction of the anastomosed vein at the level of insertion into the left atrium. The patient underwent repeat sternotomy and repair of pulmonary vein obstruction using a CorMatrix patch and sutureless reconstruction of the right-sided pulmonary vein obstruction. She recovered from this procedure with some improvement of her symptoms.

Scimitar syndrome is a rare congenital anomaly characterized by partial anomalous pulmonary venous connection of the right pulmonary veins to the IVC, anomalous systemic arterial supply to the right lung and a variable degree of right lung hypoplasia with or without sequestration (1). Partial anomalous pulmonary venous return (PAPVR) is an uncommon congenital anomaly and accounts for less than 1% of congenital heart lesions. Scimitar syndrome comprises 3-5% of PAPVR (2). The term refers to the distinctively shaped vein, which resembles a short, curved Turkish sword called a scimitar (Figure 2). Most cases present in infancy and childhood and diagnosis in adulthood is quite rare. In adults, the clinical presentation is variable and the lesion may be discovered incidentally or patients can present with dyspnea, pulmonary hypertension and recurrent right-sided lung infection (1). The treatment of isolated PAPVR in adults is controversial due to rare occurrence of the disease, complexity of surgical repair and risk of re-stenosis. Surgical repair involves re-implantation of the anomalous vein into the left atrium and can be a complex and difficult procedure,
however it can be accomplished with low morbidity and mortality at specialized centers (3). Thrombosis or stenosis of the scimitar vein is a serious complication of surgery. Our patient demonstrates some of the challenges in diagnosis and treatment and illustrates some of the post-operative complications of this rare disease.

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References