Case Report

Breathless at the Point of a Sword

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Abstract

Context: Scimitar syndrome is a congenital anomaly of pulmonary venous return where right pulmonary artery drains into right side other heart, instead of the left side, causing pulmonary hypertension resulting in shortness of breath, recurrent lower respiratory tract infections, chest pain, and fatigue. Early diagnosis and surgical intervention would correct this congenital anomaly reducing morbidity and complications in otherwise healthy young patients. Case Report: We present a case of an 18-year-old female who presented with exertional shortness of breath, fatigue, and recurrent lower respiratory tract infections. She had unremarkable physical examination but chest x-ray showed an abnormal opacity next to right heart border. Computed tomography (CT) chest was performed that showed possible scimitar syndrome. Transesophageal echocardiogram (TEE) and right heart catheterization (RHC) confirmed the diagnosis. Conclusion: Scimitar syndrome is a very rare congenital anomaly of pulmonary venous return. It is usually diagnosed in early childhood but the diagnosis may be delayed until later in adulthood. The consequences are pulmonary hypertension, right-sided heart failure, and frequent pulmonary infections resulting in increased morbidity, mortality, and frequent doctor visits for otherwise healthy young patients.

Keywords: Multidetector computed tomography (MDCT) chest, pulmonary venous return, scimitar, scimitar syndrome

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Introduction

Pulmonary venous return usually consists of four pulmonary veins with two veins from the right lung and two veins from the left lung all draining into the left atrium. Various forms of anomalous pulmonary venous return could complicate the situation. Scimitar syndrome is a rare congenital anomaly where single right pulmonary vein drains the pulmonary blood into the inferior vena cava (IVC) or right atrium. This anomaly is usually diagnosed in childhood and is surgically corrected. In a few cases, diagnosis could be delayed until adulthood resulting in long-term complications.

Due to increased preload, the right side of the heart is overloaded resulting in pulmonary hypertension in the long run and right-sided heart failure. It also results in recurrent respiratory infections and the patients could present with respiratory symptoms including chest pain, exertional dyspnea, cough, and congestion. In case of delayed diagnosis, complications of this condition with resultant increased economic burden of frequent visits to urgent care, emergency department (ED), and doctor’s office culminating in multiple necessary and unnecessary medical interventions. As “eyes can’t see what mind does not know,” it is important for primary care physicians and urgent care providers to have the knowledge of this rare anomaly while dealing with otherwise healthy young patients with common respiratory signs and symptoms.

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**Case Presentation**

Our patient was an 18-year-old female who presented with worsening dyspnea on exertion and fatigue for the last 6 months. Her past medical history was significant for frequent respiratory tract infections in her childhood which were treated as upper respiratory tract infections with antibiotics and she was discharged from the hospital. The reason for these frequent infections was never been thoroughly investigated. On examination, she was hemodynamically stable with blood pressure of 130/80, heart rate of 86, and respiratory rate of 20. Her cardiac and respiratory examinations were normal. Her chest x-ray showed a curvilinear opacity adjacent to her right heart border. Bronchovascular markings were prominent on the right side [Figure 1]. Electrocardiogram (EKG) showed right axis deviation and right ventricular hypertrophy. The finding of the x-ray raised the suspicion of the admitting physician to order computed tomography (CT) chest that revealed dilated curvilinear pulmonary vein, which appeared to drain into the IVC. The diagnosis of scimitar syndrome (partial anomalous pulmonary venous return) was suspected [Figures 2 and 3]. TEE showed single anomalous pulmonary vein entering at the junction of right atrium and IVC [Figure 4].

RHC showed significant left to right shunt (Qp/Qs ratio was 3.25) and elevated pulmonary artery pressure at 40 mmHg [Figure 5]. She was diagnosed with scimitar syndrome, and she was scheduled for repair of her partial anomalous pulmonary venous return.

**Discussion**

Scimitar syndrome, first described by Chassinat in 1836, is a variant of partial anomalous pulmonary venous drainage (PAPVD). It is a rare complex congenital anomaly characterized by an abnormal pulmonary venous return of the right lung into the IVC or right atrium. It is usually diagnosed in childhood, but it may be delayed until later in life when it is discovered.
incidentally.\[1\] It is often associated with not only other cardiac defects including atrial septal defect, patent ductus arteriosus,\[2\] and hypoplasia of the left heart and aorta\[3\] but also lung anomalies such as congenital right pulmonary hypoplasia and anomalous blood supply of the right pulmonary lower lobe from the infradiaphragmatic segment of the aorta.\[4\] It is a very rare disorder with an incidence of 1/100,000 live births. Median age of diagnosis is 7 months.\[5\] Clinical features vary and depend upon the age of presentation and underlying complications of heart failure, pulmonary hypertension, or repeated pulmonary infections due to these complications. Infants usually present with more severe disease and have a poorer prognosis as compared to children and adults with this condition.\[5,6\] Infants with this condition present with tachycardia, failure to thrive, poor feeding, lethargy, and cyanosis. Adult patients present with fatigue, exertional dyspnea, and recurrent lower respiratory tract infections,\[3\] although children with this condition have a higher incidence of respiratory complications.\[7\] Diagnosis requires a careful history, physical examination, and appropriate imaging studies. Physical examination may demonstrate a prominent precordial bulge (in patients with large left to right shunt), a systolic murmur or diastolic rumble. Aortic and pulmonary components of the S2 are usually fixed split. Chest x-ray usually demonstrates scimitar-shaped “Turkish sword sign”\[8\] or “scimitar sign”.\[9\] “Scimitar sign” [Figure 1] is also present in anomalous unilateral single pulmonary vein (AUSPV) that needs to be differentiated from the scimitar syndrome due to different prognosis.\[10\] CT chest could further aid in diagnosis with typical features of anomalous pulmonary venous return. Echocardiography and magnetic resonance imaging also help to delineate the anatomy and direction of flow. Although right heart catheterization (RHC) is the gold standard confirmatory test for the diagnosis (which could also be used for therapeutic occlusion of small anomalous venous connections) but noninvasive multidetector CT (MDCT), angiography, and magnetic resonance (MR) angiography have recently been proven to be better modalities to further assess the anatomical details of the anomalous vasculature and guide the surgical repair while decreasing the risks associated with invasive heart catheterization.\[11\] It identifies the course of pulmonary venous drainage, and also measures the degree of left to right shunt, pulmonary vascular resistance, ventricular pressures, and cardiac output. The need for treatment depends on the presence of symptoms, amount of left to right shunt, and pulmonary vascular resistance.\[12\] Asymptomatic patients with small shunt do not necessarily need any treatment as the small shunt does not have any clinical effects and most of these patients have normal life expectancy without surgical intervention. The treatment consists of surgical intervention, which is the definitive treatment and is needed for large, hemodynamically significant left to right shunts (Qp/Qs ratio >1.5) resulting in symptoms of pulmonary hypertension or heart failure.\[13\]

Conclusion

Early diagnosis and prompt surgical intervention of this rare congenital anomaly would result in significant improvement in the quality of life of otherwise healthy young patients and could also prevent the economic burden of frequent doctor visits and unnecessary medical interventions.

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Conflicts of interest

There are no conflicts of interest.

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