Severe Pulmonary Hypertension in an 87-Year-Old Woman With Scimitar Syndrome

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Abstract

Scimitar syndrome is a rare congenital cardiovascular disorder characterized by terminal insertion of the right pulmonary vein into the inferior vena cava. Right lung hypoplasia, dextraposition of the heart, and anomalous systemic collaterals from the aorta to the right lung are also seen. Patients with scimitar syndrome typically present in infancy with pulmonary hypertension or heart failure, or as asymptomatic adults, in which scimitar is incidentally discovered during imaging studies. In this case, we describe an 87-year-old woman with scimitar syndrome who presented with severe pulmonary hypertension. The patient remained asymptomatic until 2014 when she presented with dyspnea on exertion. An echocardiogram showed a right ventricular systolic pressure (RVSP) of 120 mmHg–125 mmHg. She was started empirically on phosphodiesterase-5 inhibitor with improvement in symptoms and decreased RVSP. The patient, however, deteriorated and died of right heart failure 5 months after initiation of therapy. Few cases of adults with scimitar syndrome and pulmonary hypertension have been reported. This article highlights a rare case of scimitar syndrome in an elderly woman with transient improvement of pulmonary hypertension on empiric phosphodiesterase-5 inhibitors. To our knowledge, this patient represents the oldest case of scimitar with associated pulmonary hypertension.

Scimitar syndrome, a variant of partial anomalous pulmonary venous connection, is a rare congenital cardiovascular disorder that occurs in less than 1% of the general population. It is characterized by terminal insertion of the right pulmonary vein into the inferior vena cava and is associated with right lung hypoplasia, dextraposition of the heart, and anomalous systemic collaterals from the aorta to the right lung. On chest X-ray, the anomalous vein appears as a curved shadow that resembles a scimitar—a Turkish sword.

Scimitar syndrome typically presents during infancy or in adulthood. Infant presentation is associated with signs of heart failure and pulmonary hypertension, including hemoptysis, tachypnea, and poor growth. These young patients often require early surgical intervention and have a poor prognosis. In contrast, adults with scimitar are often diagnosed incidentally during imaging studies. Adults with unrepaired scimitar can be asymptomatic, but they may also have a history of recurrent pulmonary infections and exertional dyspnea; adult patients rarely require intervention. The infantile and adult forms of scimitar likely represent extremes on a continuum of disease presentation.

Case Description

We present a novel case of an 87-year-old woman with unrepaired scimitar syndrome who developed severe pulmonary hypertension late in life. The patient’s medical history is significant for atrial fibrillation, aortic insufficiency, stroke, and hypertension. Computed tomography (CT) of the chest was performed in 2004 because of an abnormal chest radiograph finding. To our knowledge, this was the first imaging study in this patient to detail the partial anomalous pulmonary venous return, congenital hypoplasia of the right lung, large pulmonary arteries, aberrant arterial supply...
to a portion of the right lower lobe, and dextrocardia consistent with scimitar syndrome (Fig. 1).

Mild pulmonary hypertension with a right ventricular systolic pressure (RVSP) of 60 mmHg was first noted during a 2009 echocardiogram performed for a transient ischemic attack. We have no characterization of the patient’s level of dyspnea or any record that she obtained pulmonary follow-up. In 2011, she had her first complication—pneumonia with an exudative pleural effusion. The pneumonia resolved with antibiotics and the pleural effusion was drained by a thoracentesis.

In 2014, the patient presented to her primary care physician with increasing dyspnea upon minimal exertion, cough, fatigue, and changes in her exercise capacity. An echocardiogram demonstrated an RVSP of 120 mmHg–125 mmHg with severe right ventricular enlargement, severe right atrial enlargement, and preserved left ventricular systolic function with moderate aortic stenosis. CT angiogram demonstrated persistent dilation of pulmonary arterial tree with no evidence of pulmonary embolism. Further hemodynamic evaluation, including heart catheterization, was recommended but declined by the patient. She was started on a phosphodiesterase-5 (PDE5) inhibitor, bronchodilators, and nocturnal oxygen.

Following 1 month of treatment, the patient’s symptoms improved: Her basic natriuretic peptide (BNP) levels decreased (1762 pg/mL to 618 pg/mL), and her 6-minute walking distance remained stable (255 m to 243 m). Three months after starting PDE5 inhibitor, the patient experienced worsening dyspnea and lower extremity edema and was started on a loop diuretic. During this time, her BNP levels ranged from 269 pg/mL–778 pg/mL, and an additional 6-minute walking test showed a sizable decrease in distance (109 m). A repeat echocardiogram, however, demonstrated an improvement in pulmonary hypertension with an RVSP of 65 mmHg–70 mmHg. Unfortunately, the patient deteriorated and died of right heart failure 5 months after initiation of therapy.

**Discussion**

The authors suspect that this woman had a significant left-to-right shunt through the scimitar vein resulting in overcirculation of the pulmonary vasculature and increased pulmonary vascular resistance. This left-to-right shunt is the result of recirculation of oxygenated pulmonary venous blood through the pulmonary vasculature. The fraction of blood being shunted depends on the proportion of anomalous drainage compared with the total pulmonary venous return. This fraction is influenced by the number and size of anomalous pulmonary veins, pulmonary segment from which the scimitar vein originates as a result of differences in distribution of blood flow to each segment, relative resistance of the pulmonary veins, and compliance of the respective receiving chambers.8
PULMONARY HYPERTENSION IN LATE SCIMITAR SYNDROME

The patient’s condition most closely resembles the World Health Organization’s Group 1 classification of pulmonary arterial hypertension (PAH), which is associated with congenital heart disease. This case, however, is unusual in that the initial presentation of PAH was both late and severe.

Possible contributing factors to the patient’s pulmonary hypertension include aortic stenosis, aortic insufficiency, paroxysmal atrial fibrillation, and hypertension leading to increased left ventricular end diastolic pressures, diastolic heart failure, and consequent pulmonary hypertension. Because of the absence of invasive hemodynamic testing, the exact cause of her pulmonary hypertension is unknown. Treatment with a PDE5 inhibitor was, therefore, empiric and not without risk. The decision to treat her PAH was undertaken only after extensive discussions with the patient and her family, who desired a minimally invasive and palliative approach.

The patient initially improved with treatment, but then succumbed to the natural progression of her advanced pulmonary hypertension. It is unknown whether the PDE5 inhibitor contributed to worsening pulmonary venous congestion secondary to a decrease in pulmonary vascular resistance. Although several cases of scimitar with pulmonary hypertension in adults have been described in the literature, no case has reported the severity of pulmonary hypertension seen in this patient.3,7,9

Scimitar syndrome is a rare condition that is generally asymptomatic in adults. However, when adults with scimitar present with PAH, it can result in debilitating symptoms and a poor prognosis, similar to the infantile form of scimitar.7 We propose that identifying causes of pulmonary hypertension in adult scimitar may allow physicians to identify and prevent PAH in adults with asymptomatic scimitar syndrome. Additionally, optimal treatment of this condition is unknown.2 Although PDE5 inhibitors are a standard therapy in managing PAH, their use in relation to infantile scimitar has been published in the literature only once.6,13

This case suggests the need to include a cardiologist within the treatment team, the importance of right-heart catheterization to better understand the causes, the need to consider other treatment modalities besides pulmonary vasodilators (such as angiotensin-converting enzyme inhibitors and diuretics), and the need to involve an experienced adult congenital team member in complex multimorbidity cases.2,9

Conclusion

In conclusion, we have presented a rare case of scimitar syndrome with severe pulmonary hypertension in an elderly woman with symptomatic and hemodynamic improvement, albeit transient on empiric PDE5 inhibitor. To our knowledge, this patient is the oldest case of scimitar syndrome with associated pulmonary hypertension in the literature.

References


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