Scimitar Syndrome Found at Age 36

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Introduction

A rare congenital heart condition in which anomalous venous return from part or the entire right lung inappropriately goes to the inferior vena cava, Scimitar Syndrome can result in right lung hypoplasia, pulmonary hypertension, shunting, and other complications. The condition only accounts for 3% to 6% of the partial anomalous venous connections, but even rarer is the diagnosis well into adulthood. In this case report will be described a case found incidentally in a 36-year-old female.

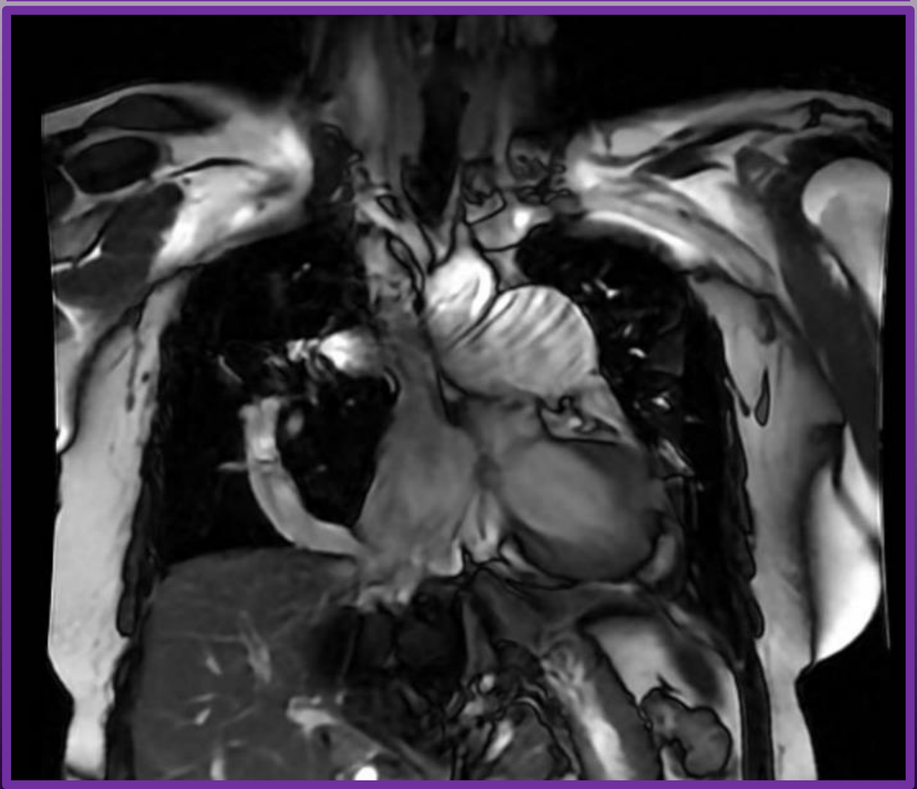
Case Summary

The patient is a 36-year-old female with a past medical history of migraines, depression, scoliosis requiring surgical correction, and a benign heart murmur who presented to the emergency department for right flank pain, nausea, vomiting, urinary frequency and urgency. She was admitted for pyelonephritis requiring antibiotics. A CT abdomen and pelvis without contrast did not show signs of pyelonephritis, but did show a "prominent pulmonary venous structure" that was unable to be completely evaluated. After this hospitalization, it was recommended that she follow up with pulmonology.

Preceding this admission, she had a transthoracic echocardiogram showing left ventricle ejection fraction of 60-65%, and normal diastolic function. Her right atrium was mildly dilated, right ventricle was mildly dilated and septal flattening was present. Pulmonary artery pressure was approximated at 35 to 45 mmHG.

Follow-up with pulmonology revealed that her only symptom was shortness of breath with cardio-type exercises. Her spirometry was significant for decreased FVC and FEV1 with normal ratio and mildly decreased volumes suggestive of mild restriction. Her 6-minute walk study was within normal limits with no desaturations noted. She was referred to interventional cardiology for right heart catheterization with saturation measurements.

This revealed pulmonary artery pressures of 44mm HG during systole and 17 mm HG during diastole, with mean of 29mm Hg. Wedge mean pressure was 10 mm Hg. The oxygen saturations were as follows: High IVC 60.6, Low IVC 60.6, SVC 70.1, RA 86.9%, RV 84.7%, PA 83.1%. The saturation at the area of anomalous pulmonary venous return to the IVC was greater than 95%. A CT angiogram as well as surgical referral were placed due to the significance of this shunt, Qp/Qs 1.83. A cardiac MRI was ordered and the patient's next steps are to follow up with surgery. This CT Angiogram and MRI confirmed the presence of partial anomalous pulmonary venous return of the right upper and lower pulmonary veins, which join into a common vein and drain into the IVC just below its junction with the right atrium.



Cardiac MRI showing the titular "Scimitar Vein" of the right lung draining into the inferior vena cava. The right pulmonary artery does not run completely contiguous, but rather divides into smaller branches before reconstituting into the vein.

Discussion

This patient is currently awaiting appointments with cardiothoracic surgery at a larger, tertiary care center. Surgical treatments typically involve correction of the abnormal venous return, though right lung resection may be required in severe case. This patient will most likely undergo direct reimplantation of the anomalous scimitar vein directly to the left atrium. Data from a multicenter study recommended this approach to other surgical options due to lower rates of stenosis or occlusion, though there is still risk of stenosis or occlusion requiring cardiac catheterization or a second operation.

Conclusion

The severity of scimitar syndrome may depend on the severity of the associated congenital heart disease as well as the age of presentation. In this case, this patient had very mild symptoms despite having the impressive imaging findings seen here. It is important to keep rare etiologies in mind when a patient presents with pulmonary hypertension and shortness of breath, and right heart catheterization should always be considered.

References

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