

Case Report

Scimitar syndrome in an older adult

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Abstract

Scimitar syndrome is a rare congenital heart disease. It is divided into subgroups of infantile, adult, and multiple cardiac and extracardiac malformation. Most patients are diagnosed during infancy and occasionally in adolescence, but very few patients are older than 20 years of age, and only some cases have severe symptoms that require surgical correction. We report a case of a man 54 years of age who was diagnosed with asymptomatic scimitar syndrome with insignificant left-to-right shunt ($Q_p/Q_s = 1.51$) with a medical history of type 2 diabetes mellitus and hyperlipidemia. Related literature on scimitar syndrome, particularly on older adults, is also reviewed. Copyright © 2011 Elsevier Taiwan LLC and the Chinese Medical Association. All rights reserved.

Keywords: adult; asymptomatic; congenital heart disease; scimitar syndrome

1. Introduction

Scimitar syndrome is a congenital heart disease involving the pulmonary venous connection to the right heart in association with pulmonary abnormalities. The infantile form is usually cyanotic and requires surgical intervention; however, the adult form is usually asymptomatic and surgical repair is dependent on the severity of left–right shunt. We present a case of an adult incidentally diagnosed with scimitar syndrome.

2. Case report

A man 54 years of age with underlying type 2 diabetes mellitus and hyperlipidemia controlled well by metformin and simvastatin for many years presented with dizziness, nausea, and vomiting for a month, which became aggravated when

turning right in bed. He took antihistamines, benzodiazepine, and diphenidol with transient symptom relief. Vertigo and peripheral vasculopathy were diagnosed by an otorhinolaryngologist. A routine chest x-ray revealed an engorged vessel arising from the right heart border to the diaphragm. Right atrial hypertrophy was also noted. Under the impression of scimitar syndrome, the patient was referred to our hospital.

On consultation, there was no shortness of breath, cough, orthopnea, or hemoptysis, but the patient intermittently experienced mild dyspnea on exertion. He denied a history of recurrent pulmonary infection. Physical examination was unremarkable. Laboratory results revealed normal blood cell count (red blood cell count: 4.93 m/cumm, hemoglobin: 15.0 g/dL, hematocrit: 44%, mean cell volume: 90.0 fL, red blood cell distribution width: 13.3%, platelets 244,000/cumm, and mean corpuscular hemoglobin concentration: 33.4 g/dL). Arterial blood gas analysis showed PaO₂ 88 mm Hg, PCO₂ 46 mm Hg, HCO₃ 27 mm Hg, and pH 7.39 at room temperature, and venous blood gas analysis showed PO₂ 39 mm Hg, PCO₂ 53 mm Hg, HCO₃ 29 mm Hg, and pH 7.35 at room temperature. The A-a gradient was 4.2 mm Hg, while

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electrocardiography revealed an incomplete right-bundle branch block. The left ventricle ejection fraction (LVEF) was 52% and right ventricle ejection fraction 42%.

Chest computed tomography (CT) with arterial- and venous-phase angiogram reconstruction (Fig. 1) showed two engorged pulmonary veins returning to the inferior vena cava (IVC) at the diaphragm level, with a web-like material between the IVC

and pulmonary vein. An absence of a right middle lobe and hypoplasia of the lingular segment were observed, which lead to decreased lung volume. Other abnormalities such as bronchiectasis, right-side pulmonary sequestration, and anomalous systemic arterial supply were not observed.

Cardiac sonography revealed a moderately dilated right atrium, mildly dilated right ventricle (RV) [RV diastolic area:

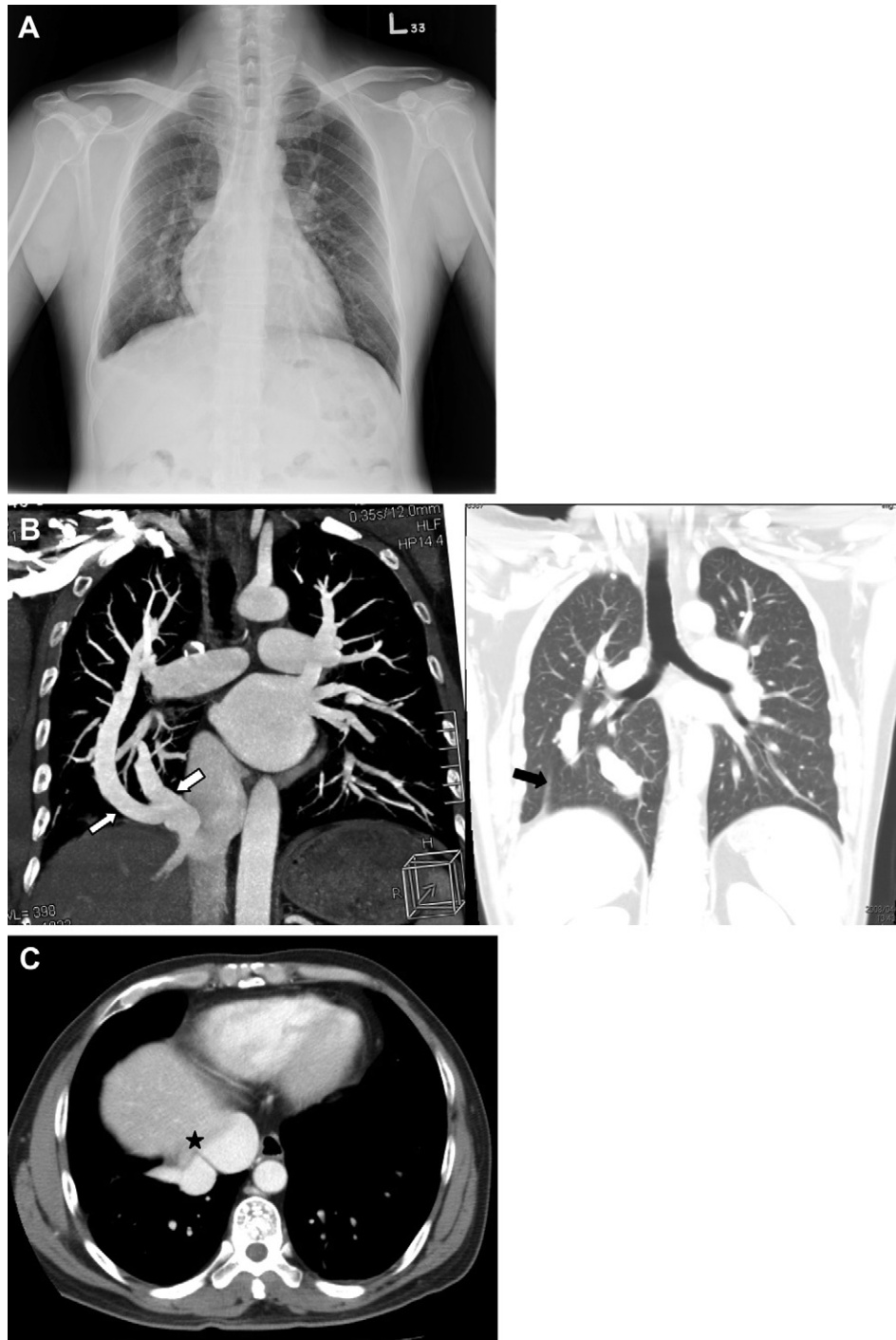


Fig. 1. (A) Chest x-ray; (B) the double scimitar vein drained into the suprarenic inferior vena cava (IVC) with two convergent pulmonary veins (white arrow). Chest computed tomography (CT) coronal view with lung window showed an absence of right minor fissure and lack of right middle lobe. Only the right major fissure was observed (black arrow); (C) Chest CT cross-section where the scimitar vein joined the IVC, with a web-like material over the drainage site (black star).

35.4 cm², RV systolic area: 19.7 cm²],¹ normal left ventricular chamber size, wall thickness, systolic function (LVEF = 63%), and no septal defects. The tricuspid valve was prolapsed, while the mitral valve was thickened, with mild mitral regurgitation, tricuspid regurgitation, and pulmonary regurgitation. Pulmonary artery flow was increased, with a peak velocity of 1.19 m/s. There was also an abnormal return of the right pulmonary vein draining into the right atrium from the subhepatic region by transesophageal echography. Pulmonary artery systolic pressure was 27 mm Hg (<30 mm Hg) and mean pulmonary artery pressure was 18 mm Hg (<25 mm Hg).² Left-to-right shunt was calculated, with pulmonary artery flow 5.74 m/s (l/min), aortic flow 3.82 l/min, and Qp/Qs = 1.51.

Because there was no obvious left-to-right shunt by sonography and neither pulmonary hypertension nor atrial septal defects were observed, invasive cardiac catheterization was not performed. The patient also refused cardiac catheterization or further surgical intervention.

3. Discussion

Scimitar syndrome is a rare congenital heart disease first reported in 1836 by Cooper³ and named by Neill in 1960.⁴ Its prevalence is around 1–3 per 100,000 live births.⁵ Anomalies include total or partial anomalous pulmonary venous connection (PAPVC) of the right lung to the IVC. In the infantile form, anomalous pulmonary venous return is often combined with congenital heart deformities, and the syndrome is usually diagnosed before 1 year of age. It presents with severe respiratory insufficiency, pulmonary hypertension, and cardiac failure, and it has a poor prognosis.⁶

The adult form is usually asymptomatic and diagnosed after 1 year of age with less marked hemodynamic changes. Dupuis⁷ showed a left-to-right shunt in fewer than 50% of patients with slightly elevated pulmonary artery pressures. These patients were able to lead a normal life without surgical correction.⁷

However, a review of all the cases in the literature revealed that the percentage of adult patients with an overt left-to-right shunt may be underestimated.⁷ A comparison of adult and infantile forms of scimitar syndrome is shown in Table 1.^{6–10} Patients usually develop fatigue, exertional dyspnea, chest infections, and pulmonary hypertension if the left-to-right shunt is more than 50% and Qp/Qs is more than 1.5–2:1. Nevertheless, the adult form usually presents without pulmonary hypertension, even when mild pulmonary hypertension or a left-to-right shunt is present.

There are two forms of “scimitar vein”: (1) partial or complete right lower pulmonary vein drainage to the IVC, and (2) both the right upper and lower pulmonary veins drain into the IVC (“double scimitar vein”). In our case, the second form was present, forming one vein draining into the IVC with a web formation at the entrance.

In addition to chest x-rays, right heart catheterization, cardiac sonography, transesophageal and transcatheter echography, and newer methods such as three-dimensional (3D) CT reconstruction and magnetic resonance imaging (MRI) have been reported to confirm the presence of scimitar syndrome.

Table 1
Characteristics of infantile and adult forms of scimitar syndrome.

	Adult	Infantile
Clinical manifestation		
Dyspnea on exertion	<50%	>50%
Shortness of breath	<50%	>50%
Cyanosis	<50%	>50%
Respiratory failure	<50%	>50%
Heart failure	<50%	>50%
Primary abnormality^{6,7}		
Hypoplasia of right lung	Yes	Yes
Bronchial segmentation	Yes	Yes
Bronchiectasis	Yes	Yes
Lung sequestration ⁸		
Co-abnormality^{9,10}		
Dextrocardia	Few	Often
Atrial septal dissection	Few	Often
Other cardiac deformity	Few	Often
Hemodynamic study⁷		
Left-to-right shunt	<50%	>50%
Pulmonary hypertension %		
Mild	23%	Few
Moderate to severe	Few	Many

Noninvasive examinations, including transcatheter and transesophageal echography, have limitations in estimating the hemodynamic status of the heart, and they have biased estimations of cardiac flow by the Doppler method. Gadolinium-enhanced 3D MR angiography has been reported to be able to calculate left-to-right shunt using phase-contrast MRI of the ascending aorta, main pulmonary artery, and anomalous pulmonary vein.^{11,12} However, the results approximate those of Doppler scans, and comprehensive comparisons are required to achieve the results of catheterization. Retrograde cardiac catheterization remains the unbiased measure for hemodynamic status of left-to-right shunt if surgery is expected.

Indications of surgical correction include the presence of a left-to-right shunt exceeding 50% and lung sequestration with recurrent right-sided chest infections.¹³ Surgical correction aims to direct blood flow from the scimitar vein to the left atrium. Some options include: (1) creating a long baffle from the orifice of the scimitar vein within the inferior vena cava to the atrial septal defect,¹⁴ (2) division with reimplantation of the scimitar vein into the right atrium with an intra-atrial baffle,¹⁵ (3) direct anastomosis of the divided scimitar vein to the left atrium,¹³ and (4) partitioning the IVC into anterior and posterior compartments with a pericardial baffle.¹⁶ A recent multicenter study suggested that intra-atrial baffle repair seems to have a better prognosis than reimplantation of the scimitar vein into the left atrium,¹⁷ with lower incidences of postoperative complications and better patency rates. Despite this, the long-term prognosis after surgery remains disappointing. Causes of failure include stenosis of the intra-atrial baffle, thrombosis of the anastomosis leading to right lung infarction, pulmonary hypertension, and hemoptysis.¹⁸

In conclusion, We have reported an asymptomatic adult with scimitar syndrome and borderline Qp/Qs. Corrective

surgery was not performed because of the absence of symptoms and patient refusal. Further follow-up to monitor complications such as to left-to-right shunting is warranted.

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