

Images in Cardiovascular Disease



An Unusual Case of Scimitar Syndrome with Drainage to the Right Atrium

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Conflict of Interest

The authors have no financial conflicts of interest.

Author Contributions

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A 37-year-old woman presented with exertional dyspnea. She had a history of open-heart surgery due to an unknown atrial septal defect (ASD) at the age of 10 years. Chest X-ray showed mild cardiomegaly and an enlarged right atrium (RA) with prominent pulmonary vessels (**Figure 1**). Transthoracic echocardiography (TTE) showed right atrial and ventricular enlargement (**Figure 2A and B**) and a Qp/Qs of 3.8. In a modified apical 4-chamber view with inferior tilting, a defect mimicking a primum ASD was observed (**Figure 2C, Movies 1 and 2**). In the apical 2-chamber view, this defect was noted to connect to a conduit with multiple tributaries. The conduit was curved and had a “scimitar” shape (**Figure 2D, Movies 3 and 4**). Transesophageal echocardiography revealed no residual ASD. The scimitar vein drained into the RA, coursing behind the interatrial groove (**Figure 2E and F**). On computed tomography, the right upper, middle, and lower pulmonary veins coalesced into the scimitar vein behind the interatrial groove and drained into the RA (**Figure 3**). The patient was referred to a tertiary hospital where she underwent surgery.

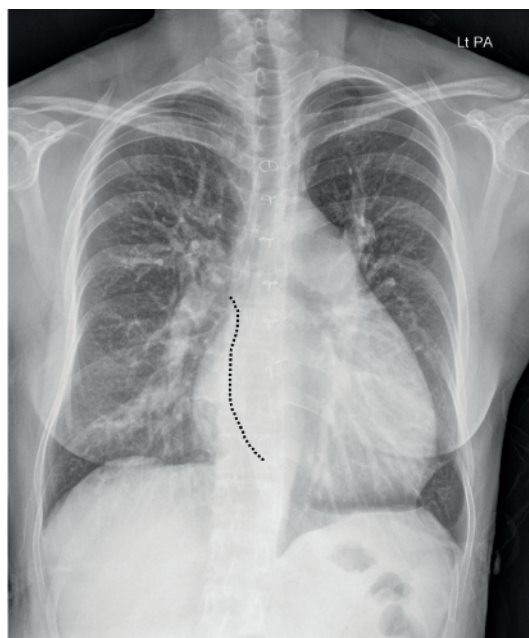


Figure 1. Postero-anterior chest X-ray shows mild cardiomegaly, and an enlarged right atrium with prominent pulmonary vessels. The scimitar vein (dotted line), which is not well detected by visual inspection from this view, courses behind the junction of the superior vena cava and the right atrium before travelling down to the interatrial groove and draining to the right atrium.

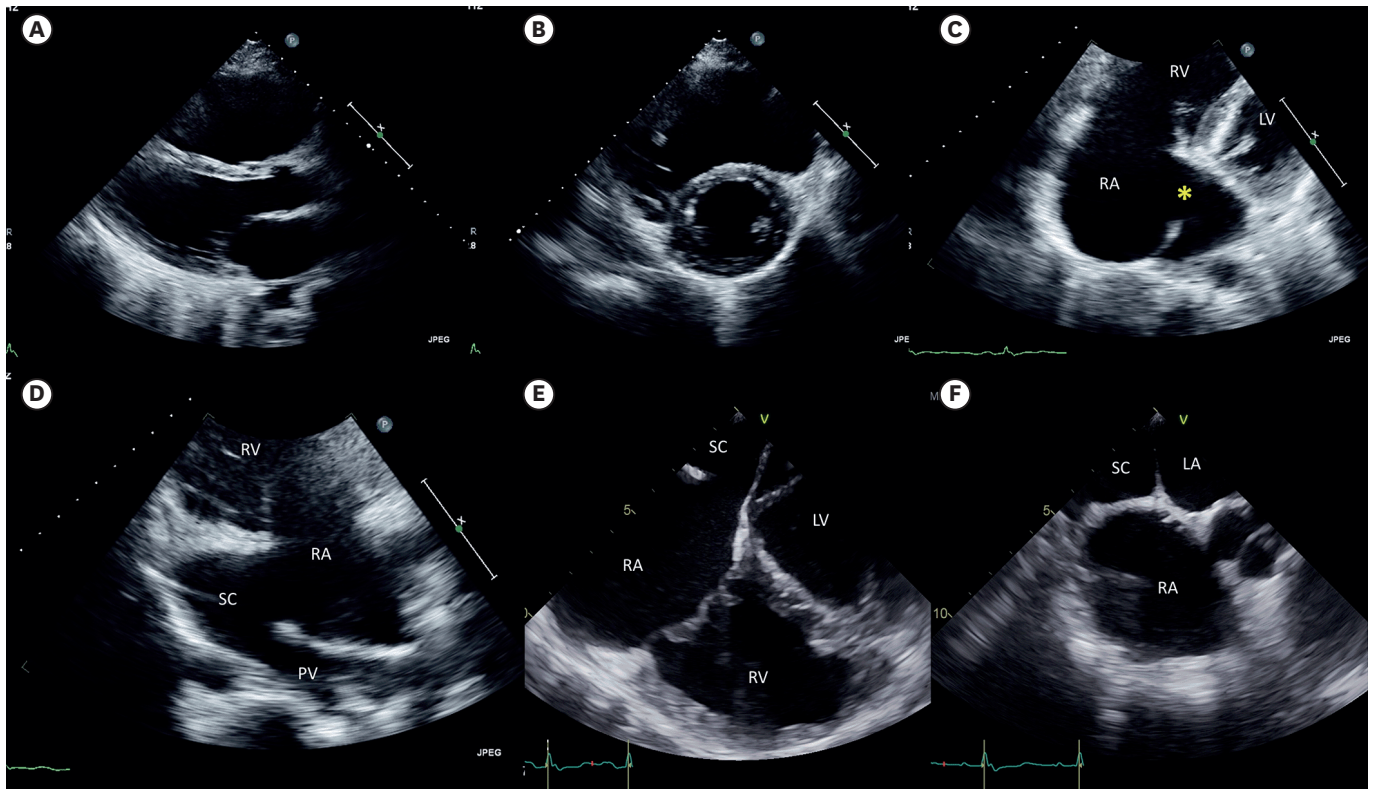


Figure 2. (A, B) Transthoracic echocardiography shows the RV and RA are enlarged. (C) Modified apical 4-chamber view with inferior tilting shows a large defect (yellow asterisk) mimicking primum ASD. (D) Apical 2-chamber view reveals the defect to be connected to a scimitar-shaped conduit (SC). (E, F) Transesophageal echocardiography shows SC drains into RA, coursing behind the interatrial groove. ASD: atrial septal defect, LV: left ventricle, RA: right atrium, RV: right ventricle, PV: pulmonary vein, SC: scimitar vein.

Scimitar syndrome is characterized by anomalous pulmonary drainage in which part of or even the entire right lung is drained by the right pulmonary veins into the inferior vena cava (IVC).¹ It is extremely uncommon in scimitar syndrome for the right pulmonary veins to drain anomalously to a site other than the IVC.² Multiplane echo imaging with other imaging modalities is needed for correct diagnosis.¹ In one study, blunting of the right side of the left atrium (LA) was observed by TTE in all patients with total unilateral right-sided anomalous pulmonary venous drainage.³ Conversely, the current case showed the scimitar vein coursing between the interatrial groove and noted a characteristic finding of echocardiographic “scimitar sign” (Figure 2D, Movie 3 and 4) which was not described in previous studies. Pulmonary hypertension in scimitar syndrome is responsible for the symptoms.⁴ Surgical repair such as re-routing the scimitar vein into the LA and direct scimitar vein ‘re-implantation’ into the LA is indicated in symptomatic patients, and is safe and effective.⁵

SUPPLEMENTARY MATERIALS

Movie 1

Modified apical 4-chamber view with inferior tilting via transthoracic echocardiography reveals a defect mimicking a primum atrial septal defect.

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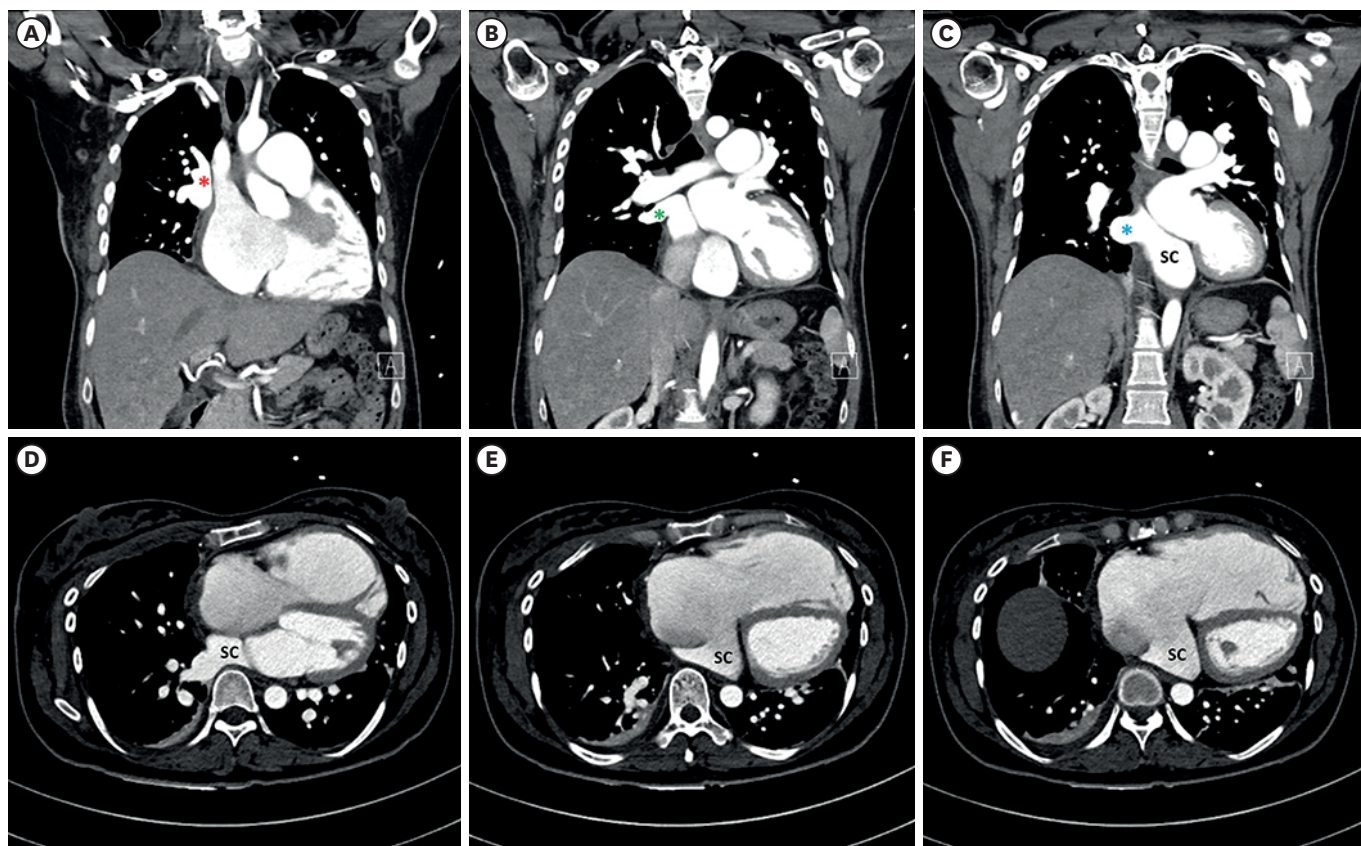


Figure 3. (A-C) Computed tomography with coronal planes (from the shallow to the deep plane), the right upper (red asterisk), middle (green asterisk), and lower pulmonary veins (blue asterisk) coalesce into the SC. (D-F) Cut-by-cut transverse planes reveal that the scimitar vein courses behind the interatrial groove and drains into the right atrium. SC: scimitar vein.

Movie 2

Color Doppler echocardiography of **Movie 1** reveals blood flow through the defect from the left to the right atrium.

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Movie 3

Apical 2-chamber view via transthoracic echocardiography reveals a curved “scimitar” vein behind the right atrium.

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Movie 4

Color Doppler echocardiography of **Movie 3** reveals the scimitar vein with blood flow from the pulmonary veins to the right atrium.

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