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The double scimitar sign

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Abstract

We present striking and unique imaging features of a child with scimitar syndrome, demonstrating a rare 'double sword appearance' across multiple imaging modalities including chest radiography, angiography and computed tomography.

Scimitar syndrome is characterised by partial or hemi-anomalous drainage of right-sided pulmonary veins into the inferior vena cava, hypoplastic right lung, dextroversion of the heart, anomalous systemic arterial collateral to right lung, and may be associated with intracardiac defects like atrial septal defect. The pulmonary vein forms a distinctive shadow on the X ray which resembles a curved Turkish sword, and hence the name "Scimitar" syndrome.

A 3-year-old girl child diagnosed to have Scimitar syndrome had undergone surgical baffling of right sided pulmonary veins to left atrium at another institute. However, on follow-up, she was found to have a large interatrial shunt (possible patch dehiscence) and subsequently referred to our institute one year later. Evaluation with echocardiogram, CT angiogram and cardiac catheterisation showed hemi-anomalous drainage of right-sided pulmonary veins to the Inferior

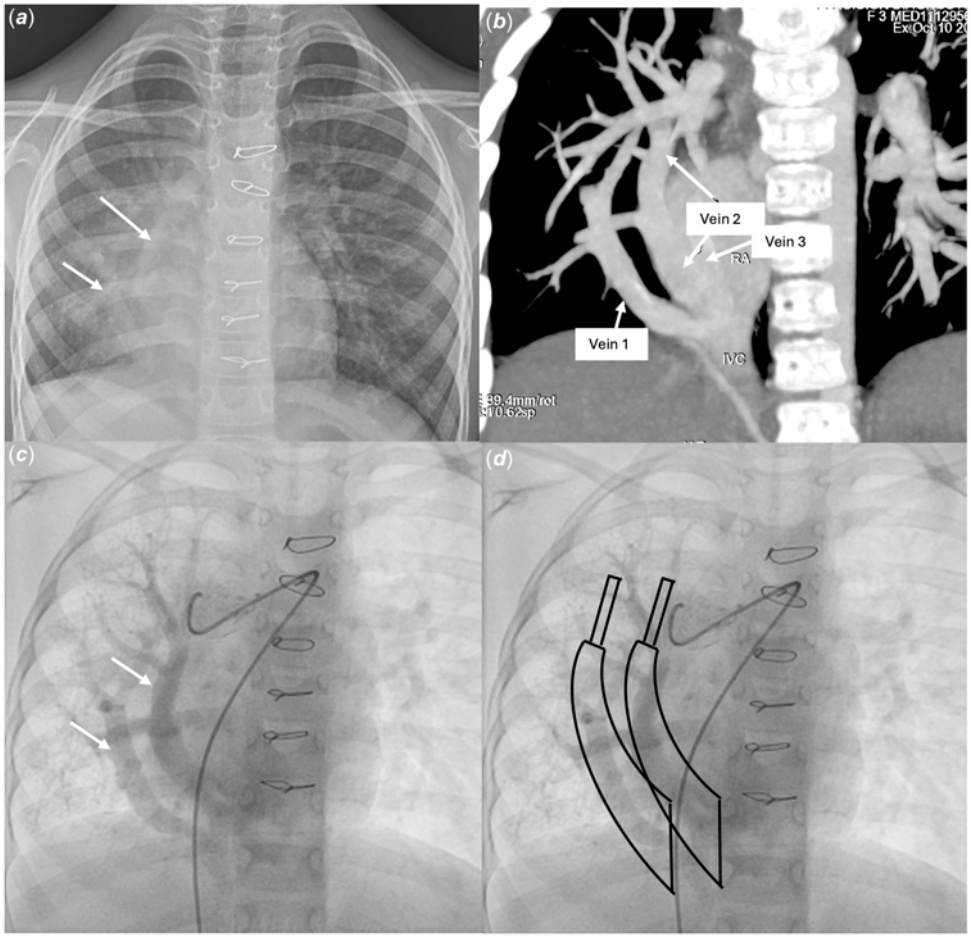


Figure 1. (A) Chest X ray of the child showing dextroversion of the heart and double scimitar sign. The two scimitar veins are marked with arrows. (B) Coronal view of CT angiogram showing three pulmonary veins from the right lung draining anomalously into the inferior vena cava. The two larger veins have the shape of Turkish sword or scimitar on the X ray and conventional angiogram. (C) Levophase of right pulmonary artery angiogram showing anomalous pulmonary venous return from right lung into inferior vena cava. The two large veins form a distinct double scimitar sign. (D) The pulmonary vein shape resembling curved Turkish swords.

vena cava-Right atrium junction and a large atrial septal defect. The right lung drained through three veins into the inferior vena cava; two of these veins were large and typically had the shape of a Turkish sword. The third vein drained a single segment of the lower lobe and did not form a typical shadow. Thus, the X ray, CT, and angiogram showed a unique "Double Scimitar" sign (Figure 1 and Video) . The small vein which was draining a single segment of lower lobe was severely stenosed at its insertion to the inferior vena cava, with a gradient of 12 mm Hg. The Qp/Qs and PVRI were 2.2 and 1.69 Wood units, respectively. A small systemic collateral from abdominal aorta to the right lung was embolised with a coil during the cardiac catheterisation. Surgery was performed through right antero-lateral thoracotomy. The pulmonary veins were translocated to the posterior wall of the right atrium and baffled to the left atrium through the atrial septal defect using bovine pericardium. Child is clinically well with unobstructed pulmonary venous drainage to the left atrium on one-year follow-up.

When there is hemi-anomalous drainage of right pulmonary veins in Scimitar syndrome, the pulmonary veins join together and often drain as a single vein into the inferior vena cava. Hence, the classical Scimitar sign is noted on X ray and angiogram. In our case, there was a unique Double Scimitar sign, due to two large veins. There are very few reports of two scimitar veins;[1,2] however, even in those reports, the classical double shadow was not demonstrated. This case is reported for the unique images on X ray, angiogram, and CT.

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Competing interests. None.

Ethical standards. The authors assert that all procedures contributing to this work comply with the ethical standards of the relevant national guidelines on human experimentation and with the Helsinki declaration of 1975, as revised in 2008, and has been approved by the institutional committees.

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