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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 (Qp/Qs = 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.

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 PaO_2 88 mm Hg, PCO_2 46 mm Hg, HCO_3 27 mm Hg, and pH 7.39 at room temperature, and venous blood gas analysis showed PO_2 39 mm Hg, PCO_2 53 mm Hg, HCO_3 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:

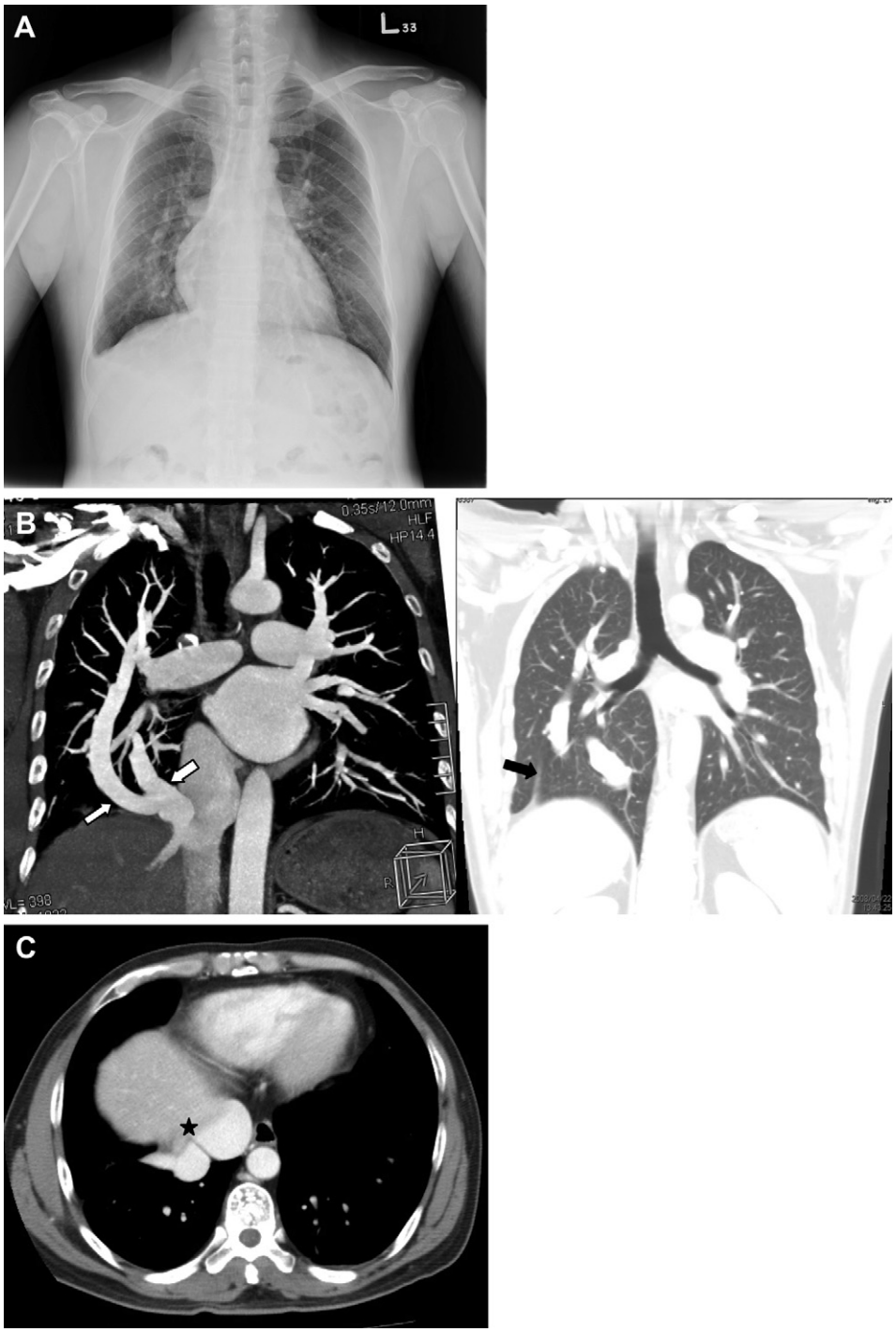

Fig. 1. (A) Chest x-ray; (B) the double scimitar vein drained into the supraphrenic inferior vena cava (IVC) with two convergent pulmonary veins (white arrow). Chest computed tomography (CT) coronal view with lung window showed an absence of right minor fissure and lack of right middle lobe. Only the 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).