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Case Report

An Adult-Onset Patient With Scimitar Syndrome: An Anatomical and Functional Evaluation by Cardiac Magnetic Resonance Imaging

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ABSTRACT

Scimitar syndrome is a rare congenital heart disease that presents with a partial anomalous venous connection and a hypoplastic right lung. It has been reported in 0.5% to 1% of all congenital heart diseases. A partial anomalous pulmonary venous structure (scimitar syndrome) draining into the inferior vena cava was detected in a 37-year-old female patient, who presented with mild dyspnea and chest pain. The anomalous pulmonary venous drainage into the inferior vena cava (scimitar syndrome) was diagnosed by cardiac magnetic resonance imaging (CMR) and 3D contrast-enhanced magnetic resonance angiography, which demonstrated the anomalous pulmonary venous connection with a pulmonary-to-systemic flow ratio noninvasively. The patient was referred for surgery without additional interventional imaging. She had a good postoperative course. We herein present 3D magnetic resonance angiography and phase-contrast CMR findings in an adult-onset patient with scimitar syndrome. ***(Iranian Heart Journal 2021; 22(2): 124-129)***

KEYWORDS: Partial anomalous pulmonary venous connection, Scimitar syndrome, Cardiac magnetic resonance imaging

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Scimitar syndrome is a rare congenital heart disease that presents with a partial anomalous venous connection and a hypoplastic right lung. The syndrome has been reported in 0.5% to 1% of all congenital heart diseases. [1] The aberrant pulmonary vein drains into a supradiaphragmatic or infradiaphragmatic portion of the inferior vena cava and usually drains the lower lobe of the right lung, causing a right-to-left shunt. The right pulmonary vein has an appearance of a curved shadow adjacent to the right atrium that resembles a curved Turkish sword. Here, we report a case of anomalous pulmonary venous drainage into the inferior vena cava (scimitar syndrome) diagnosed by cardiac magnetic resonance imaging (CMR) and 3D contrast-enhanced magnetic resonance angiography, which demonstrated the anomalous pulmonary venous connection with a pulmonary-to-systemic flow ratio noninvasively.

CASE REPORT

A 37-year-old female patient who resided in the rural parts of Turkey presented with mild dyspnea and chest pain. Her physical examination was unremarkable, and all laboratory findings were normal. Although the patient was asymptomatic, transthoracic echocardiography showed substantial dilation of the right atrium and the right ventricle, a pulmonary artery systolic pressure of 57 mm Hg, and findings of an increased right-heart pressure as a flattened interventricular septum and septal bouncing. The patient was referred to our hospital for detailed cardiac evaluations. CMR was performed to evaluate the anomalous pulmonary venous connection with right-heart function/volume and the pulmonary-systemic flow ratio. Four-chamber and short-axis cine-CMR revealed a dilated right ventricle (right ventricular end-diastolic volume =145 mL/m^2) with a preserved right ventricular ejection fraction (56%) without additional intracardiac anomaly or anomalous pulmonary arterial supply. Three-dimensional contrast-enhanced CMR demonstrated right lower pulmonary venous drainage by an anomalous large vertical vein (the scimitar vein) into the inferior vena cava just below the diaphragm (Fig. 1). Afterward, phase-contrast imaging was performed to calculate the pulmonary-systemic flow ratio (Fig. 2). For the calculation of the pulmonary-systemic flow ratio, the net pulmonary artery flow was divided by the net aortic flow. The calculated pulmonary-systemic flow ratio was 1.83 (Fig. 3). The patient was referred for cardiovascular surgery, and she had a good postoperative course.

DISCUSSION

Scimitar syndrome is a relatively uncommon congenital pathology that accounts for 0.5% to 1% of all congenital heart diseases. [1] The main component of this pathology is an anomalous pulmonary venous connection to the supradiaphragmatic or infradiaphragmatic segment of the inferior vena cava, as well as the hypoplasia of the right lung, with an aortic blood supply. [2, 3] An aberrant drainage into the inferior vena cava can be seen alone or with cardiac pathologies such as atrial septal defect, ventricular septal defect, coarctation of the aorta, total anomalous pulmonary venous connection, and patent ductus arteriosus (19%–31% of patients with scimitar syndrome). [2, 4, 5] Scimitar syndrome is classified into 3 groups: infantile, adult, and complex. Patients may present with severe pulmonary hypertension, cardiac failure, and cyanosis in infantile ages. Patients in adulthood are generally asymptomatic, but they may present with pulmonary complications such as recurrent pneumonia, mild exertional dyspnea, and hemoptysis. Patients with additional cardiac malformations in the third group tend to have more severe clinical presentations. [6] Scimitar syndrome could present with a wide spectrum of symptoms depending on the presence of other associated congenital heart diseases, the severity of right-to-left shunts, the presence of scimitar vein obstruction, the presence of arterial supply to the right lung, or bronchial abnormalities responsible for pulmonary complications. [3] Thus, the wide spectrum of symptoms and coexisting congenital anomalies at clinical onset renders the diagnosis difficult. Our patient had mild dyspnea and chest pain on admission. Although her physical examination was unremarkable, transthoracic echocardiography showed right-heart dilatation with an increased pulmonary artery systolic pressure of 57 mm Hg.

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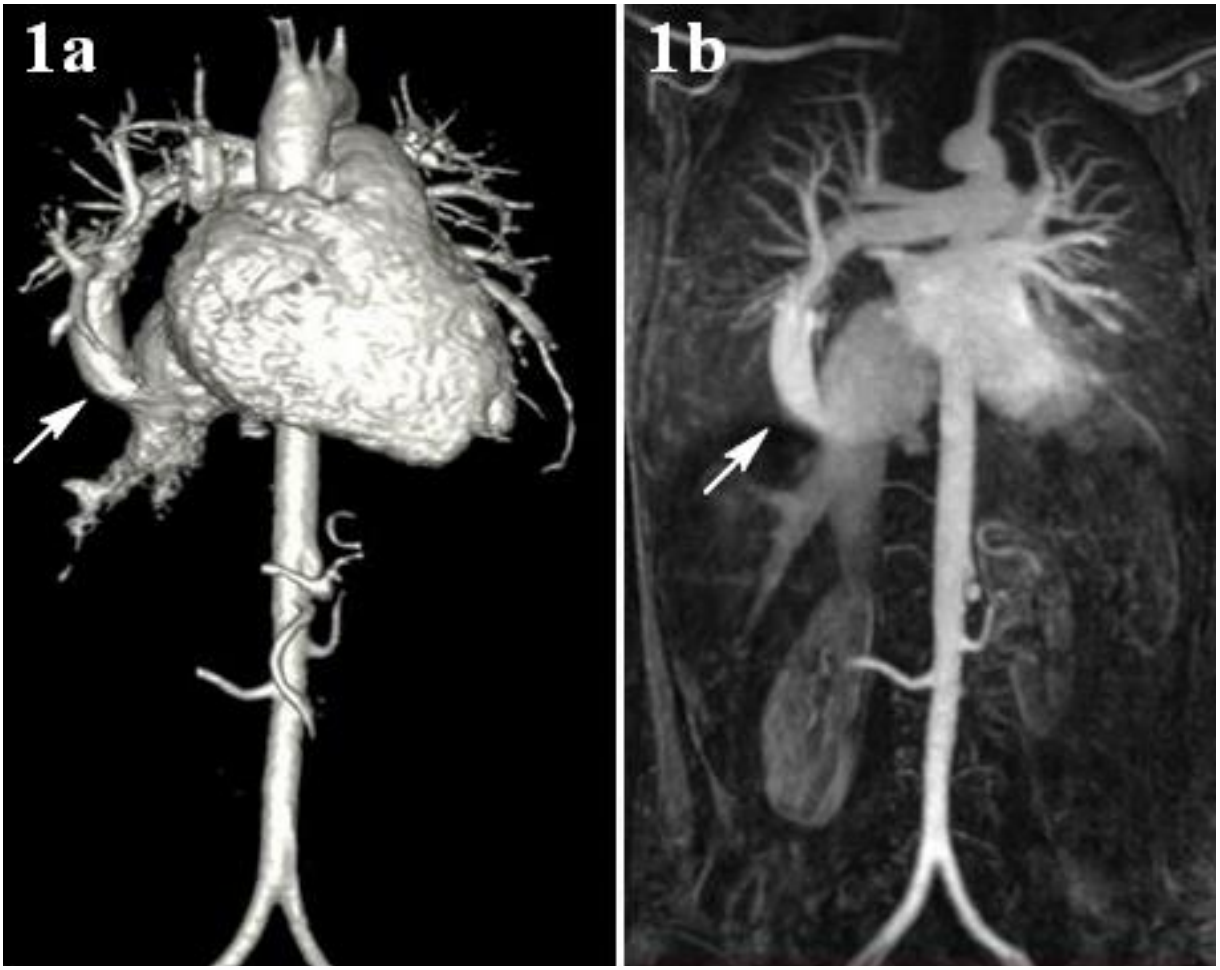

Figure 1: Gadolinium-enhanced magnetic resonance angiography is illustrated herein. Three-dimensional magnetic resonance angiography (a) and maximal-intensity projection image (b) in the coronal plane shows the drainage of the right pulmonary vein into the right atrium and the inferior vena cava connection.

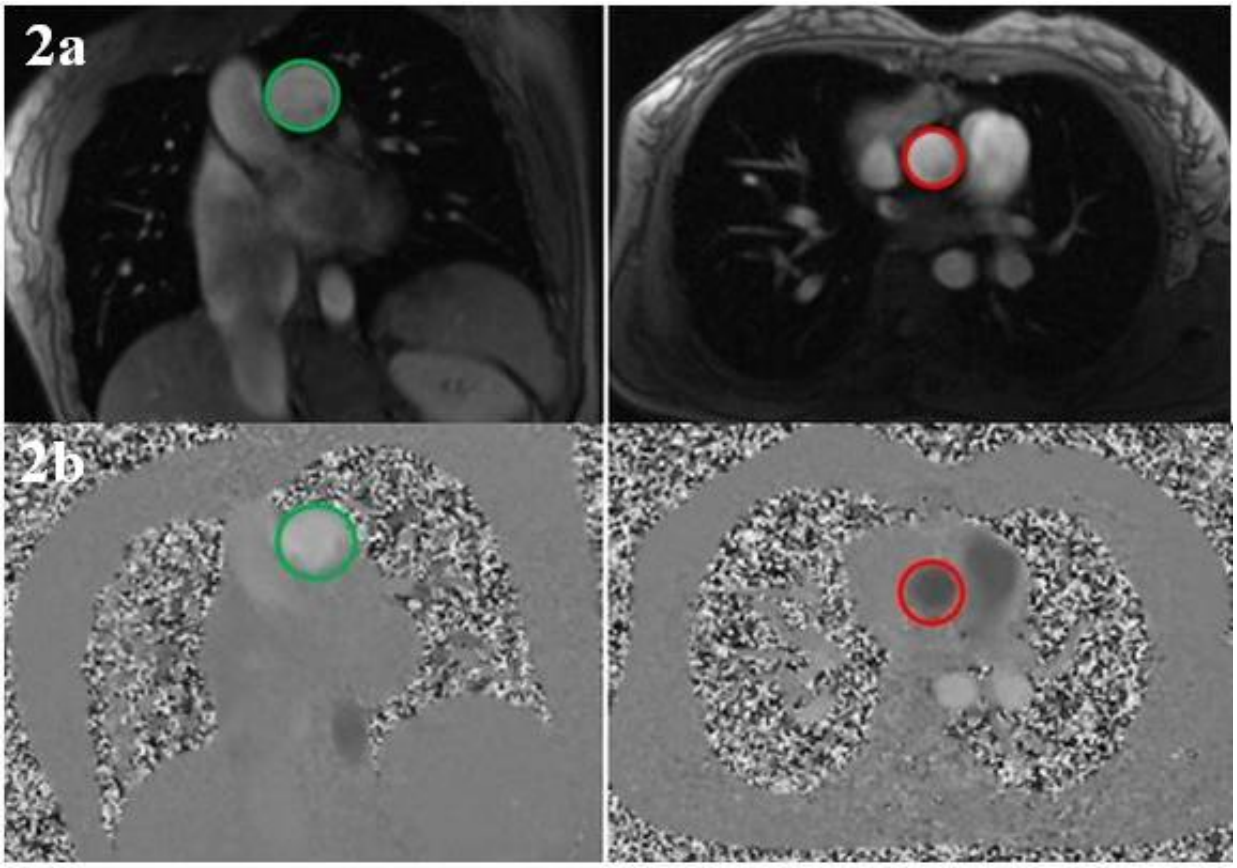

Figure 2: The magnitude image shows the anatomy of the main pulmonary artery (MPA) (green curve) and the ascending aorta (AAO) (red curve). (a) A phase image obtained in systole shows the flow through the MPA in black and the flow through the AAO in white (b).

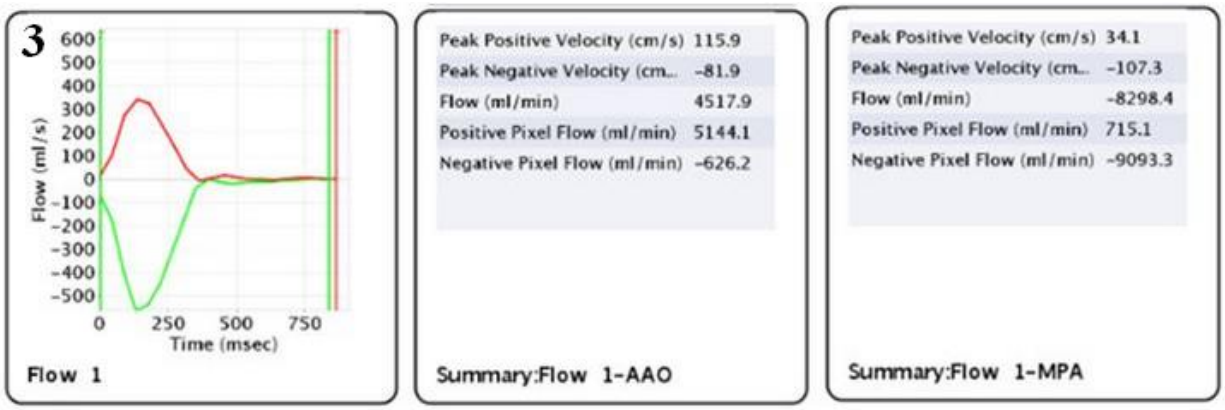

Figure 3: The images present the time-velocity curves of the main pulmonary artery (MPA) (green curve) and the ascending aorta (AAO) (red curve). For the calculation of the pulmonary-systemic flow ratio, the net pulmonary artery flow was divided by the net aortic flow. The calculated pulmonary-systemic flow ratio was 1.83.

Although chest plain radiography can suggest the diagnosis, further imaging is frequently needed. Conventional angiography, echocardiography, computed tomography, and CMR are other imaging modalities to confirm the diagnosis, to

demonstrate the anomalous pulmonary vein drainage, and to exclude the associated cardiopulmonary abnormalities. A complete evaluation requires the demonstration of all the pulmonary veins with anomalous pulmonary vein connections to the systemic circulation with the measurement of the ventricular size and function, the quantification of blood flow, and the pulmonary-systemic flow ratio, which is frequently required for surgical decision-making. [7, 8] CMR is a noninvasive imaging modality. This modality provides detailed information concerning major cardiovascular anatomic anomalies such as aberrant systemic arterial blood supplies and abnormal pulmonary venous drainages. Additionally, without radiation exposure, CMR confers quantitative functional and hemodynamic information regarding the volume and function of the ventricles, the volume and velocity of blood flow, the pulmonary-to-systemic blood flow ratio, and the blood flow distribution between the 2 lungs. [9] In phase-contrast CMR, phase-contrast velocity mapping determines the velocity, volume, and pattern of blood flow by measuring the phase shifts of moving particles compared with stationary particles, [10] and flow velocity is determined by the extent of shifting. [11] Contrary to Doppler ultrasound, the volume and pattern of the flow in imaging planes can be calculated by phase-contrast CMR with 5% inaccuracy. [10] By phase-contrast CMR, information regarding flow parameters such as flow curves, peak flows, and net flows can be demonstrated in imaging planes, and the pulmonary-to-systemic blood flow ratio and the blood flow distribution between the 2 lungs can be calculated. [9, 12] We assessed biventricular volume and function in 4-chamber and short-axis cine-CMR (Fig. 1). The aberrant course of the vascular anomaly was demonstrated on 3D magnetic resonance angiography (Fig. 2). We also evaluated the flow parameter of the pulmonary artery and the aorta and calculated the pulmonary-systemic flow ratio on phase-contrast CMR (Fig. 3). Our calculation showed a pulmonary-systemic flow ratio of 1.83, which was sufficient for surgical correction without additional interventional studies.

Surgical correction may be preferred in symptomatic patients or asymptomatic patients with an increased pulmonary-systemic flow ratio of greater than 1.5:1 to prevent the development of pulmonary hypertension and right ventricular dysfunction. In our patient, the calculated pulmonary-systemic flow ratio was 1.83; therefore, the patient was referred for cardiovascular surgery. There are different surgical approaches to scimitar syndrome such as the direct reimplantation of the aberrant pulmonary vein into the left atrium [13, 14] or baffling between the anomalous drainage and the left atrium via a tunnel. [15] In some selected cases with systemic-to-pulmonary drainage and intracardiac defects, the catheter-based closure of the aortopulmonary vascular structures and intracardiac defects followed by the correction of the anomalous veins can be another therapeutic choice. [16] Still, there is no consensus as to which is the best option.

In conclusion, scimitar syndrome can be seen in the infantile period of life or adulthood with a wide spectrum of clinical onsets. Although a chest plain graph is usually suggestive of the diagnosis, identifying a detailed anatomic structure, evaluating the functional and physiological information, and calculating the shunt degree are required for surgical decision-making. Assessment can be made noninvasively by CMR with 3D contrast-enhanced magnetic resonance angiography

and phase-contrast CMR without radiation exposure.

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