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CASE REPORT
ΕΝΔΙΑΦΕΡΟΥΣΑ ΠΕΡΙΠΤΩΣΗ

Scimitar syndrome in an adult presenting with effort-induced dyspnea
A missed diagnosis

Scimitar syndrome is a rare congenital constellation of cardio-pulmonary anomalies, classically described as an association of partial anomalous pulmonary venous return, hypoplasia of the right lung and cardiac dextroposition. The case is described of a patient with the adult form of the syndrome, with partial anomalous pulmonary venous return to the inferior vena cava and a hypoplastic right lung that had evaded diagnosis for years and was eventually successfully treated surgically.

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ΑΡΧΕΙΑ ΕΛΛΗΝΙΚΗΣ ΙΑΤΡΙΚΗΣ 2021, 38(1):95–98

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Σύνδρομο «γιαταγανίου» σε ενήλικα με δύσπνοια προσπαθείας. Μια διαλάθουσα διάγνωση

Περίληψη στο τέλος του άρθρου

Key words

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Scimitar syndrome is a rare congenital constellation of cardio-pulmonary anomalies, classically described as an association of partial anomalous pulmonary venous return (PAPVR), hypoplasia of the right lung and cardiac dextroposition.[1,2] Its name stems from the characteristic chest X-ray finding of a crescent-like shadow in the right lower lung field, produced by the anomalous venous channel that resembles a curved Turkish sword or scimitar.[2,3] Scimitar syndrome is classically divided into infant and adult forms.[4,5] The case is presented here of a patient with the adult form of the syndrome, with PAPVR to the inferior vena cava (IVC) and a hypoplastic right lung, which was corrected surgically.

CASE REPORT

The 56-year-old male presented here was suffering from

long-standing effort-induced dyspnea that had deteriorated during the previous 6 months, designated New York Heart Association (NYHA) grade III. His medical history included diabetes mellitus, dyslipidemia, and arterial hypertension. Previous cardiac and pulmonary evaluation, including transthoracic echocardiography (TTE), functional tests, and coronary angiography (CANG) had failed to establish a cause for his symptoms. Because of his persistent symptomatology, chest X-ray and computed tomography (CT) of the thorax were performed. The chest X-ray was pathognomonic for scimitar syndrome (fig. 1). The CT showed mild right lung hypoplasia, mild cardiac dextroposition, a dilated right pulmonary vein (PV) draining into the IVC, dilated right cardiac cavities and dilated main pulmonary artery (MPA) and branches, suggesting pulmonary hypertension (PHT) (figures 2a, 2b).

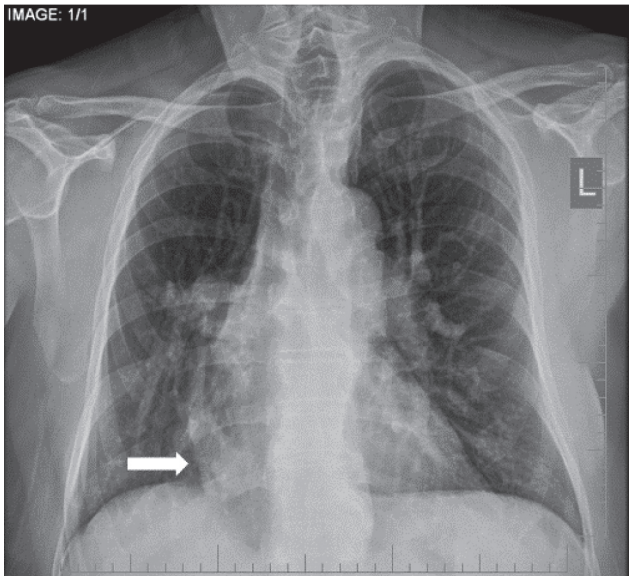

Figure 1. Scimitar syndrome in a 56-year-old male: Chest X-ray showing the anomalous draining vein as a tubular structure paralleling the right heart border in the shape of a scimitar (Turkish sword) (white arrow).

At that stage the patient came to our center, self-referred, for evaluation. The 6-minute walking test was stopped prematurely after approximately 20 meters because of severe dyspnea. The electrocardiogram (ECG) showed sinus rhythm with incomplete right bundle branch block. The TTE and transesophageal echocardiography showed normal dimensions and function of the left ventricle, with mild hypertrophy, mild dilation of the left atrium (LA), with prolapse of the posterior leaflet of the mitral valve and mild eccentric regurgitation (MR). The other valves were structurally normal. There was moderate dilation of the right chambers, and the MPA and its branches were also dilated. No atrial septal defect (ASD) of any type or patent foramen ovale was present. The left PVs were draining normally into the LA but the right PVs could not be demonstrated. The LA appendage was free of thrombi. The ascending and descending aorta were normal, without atherosclerosis. Mild to moderate PHT was suggested by Doppler ultrasound. Echocardiographic findings excluded the presence of additional endocardial defects and confirmed the scimitar vein as the cause of right heart volume overload.

The patient's clinical status (NYHA III-IV) and his poor quality of life led the heart team to recommend surgical intervention. The pre-operative CANG and right heart catheterization showed normal epicardial coronaries and mild PHT. The patient underwent a successful open-heart surgery with creation of an ASD and pericardial baffle directing flow from the anomalous ectopic PV to the newly created ASD (figures 3a, 3b). After an uncomplicated hospitalization,

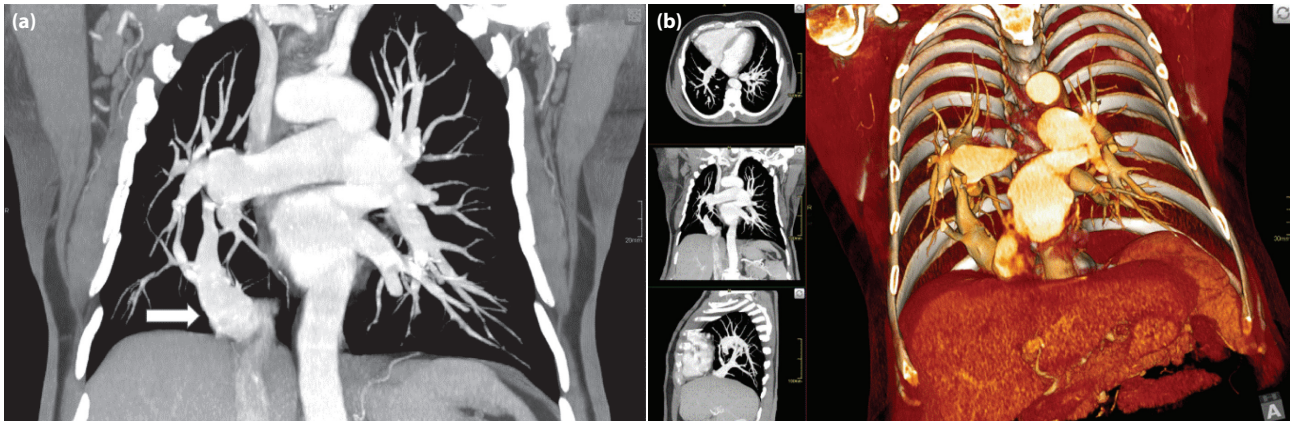

Figure 2. Scimitar syndrome in a 56-year-old male: **(a)** Coronal reformatted maximum intensity projection (MIP) image demonstrating the anomalous scimitar vein draining the hypoplastic right lung into the inferior vena cava (white arrow). **(b)** Volume rendering (VR) reconstructed computed tomography (CT) image showing the previously mentioned anomalous venous drainage (scimitar vein).

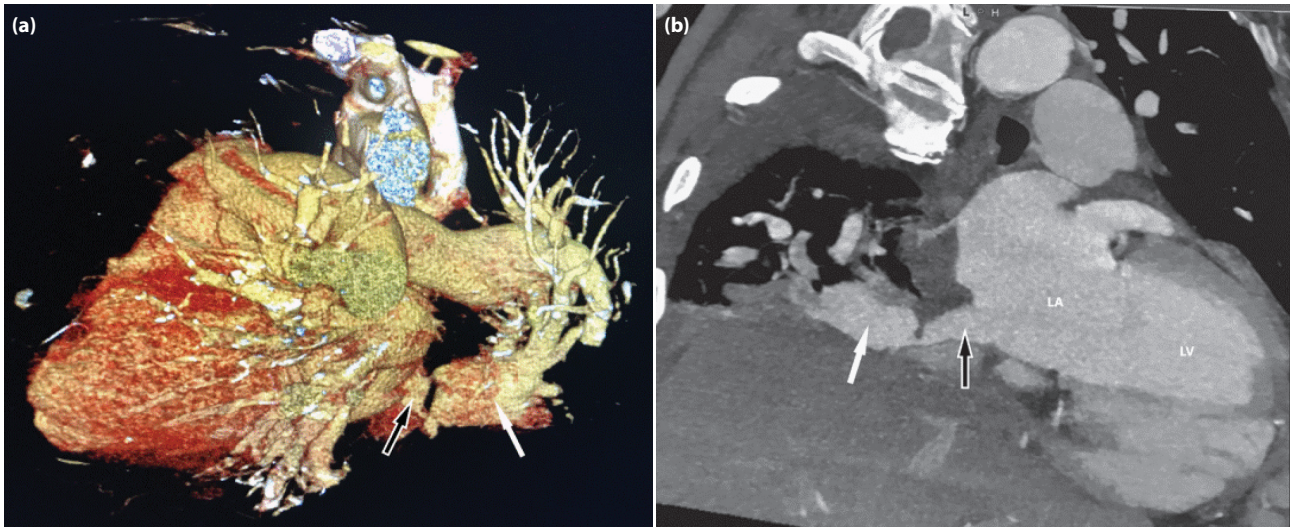

Figure 3. Scimitar syndrome in a 56-year-old male: **(a)** Volume rendering (VR) reformatted computed tomographic (CT) image showing the anomalous pulmonary vein (white arrow) and the baffle (black arrow). **(b)** Multiplanar reconstruction (MPR) CT image showing the anomalous pulmonary vein (white arrow) and the baffle (black arrow). LA: Left atrium, LV: Left ventricle.

the patient recovered fully, and at 6-week follow-up he was classified NYHA I, and he was on a low dose of diuretic. The TTE showed less dilation of the right cavities, and only mild mitral and tricuspid regurgitation.

DISCUSSION

APVR disorders are a group of rare congenital heart diseases caused by the abnormal drainage of a part of, or the entire right lung to the IVC, the RA or a variety of venous connections from the anomalous PV to a systemic vein; the estimated incidence is 2/100,000 births.[6,7] Scimitar syndrome is characterized by anomalous venous drainage of part or the entire right lung to the IVC, variable right lung hypoplasia, and variable systemic blood supply to part of the right lung.[8] Scimitar syndrome shows a bimodal presentation, with either an infantile or a pediatric/adult form.[6] In its infant form, it is diagnosed in the first two months after birth, causing symptoms of failure to thrive, tachypnea, heart failure, and cyanosis, and carries a mortality of approximately 45%. Patients with the pediatric/adult form usually have milder symptoms, depending on the degree of lung hypoplasia, or are asymptomatic.[6,7]

The diagnosis can be made by the pathognomonic scimitar sign on chest X-ray, which is present in about 70% of the patients.[6] CT and magnetic resonance imaging are of great value in delineating the anatomy of the anomalous drainage and assessment of concomitant congenital defects of the bronchovascular tree, lung and thoracic spine.[6,7]

Surgical repair is almost always required for the infantile form, while in the adult form it needs to be considered only in the case of recurrent right lung infections, significant left-to-right shunting, PHT, and cardiac symptoms.[6,8,9] The main aim of the surgical procedures, which consist of redirection of the anomalous pulmonary venous drainage into the LA, is to avoid sequelae of chronic heart overload. Because of the significant risk of post-operative scimitar drainage stenosis/occlusion, surgery should be tailored according to a comprehensive hemodynamic evaluation and to the patient's age.[8]

To our knowledge, this is the first case of scimitar syndrome ever reported in Cyprus to be was successfully treated surgically. Adult cardiologists encounter scimitar syndrome only rarely in their careers and this case report demonstrates that the diagnosis can easily be missed.

ΠΕΡΙΛΗΨΗ

Σύνδρομο «γιαταγανίου» σε ενήλικα με δύσπνοια προσπαθείας. Μια διαλάθουσα διάγνωση

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Το σύνδρομο «γιαταγανίου» (scimitar) είναι ένα σπάνιο συγγενές σύνδρομο καρδιο-πνευμονικών ανωμαλιών και κλασικά χαρακτηρίζεται από μερική ανώμαλη σύνδεση πνευμονικών φλεβών, υποπλασία του δεξιού πνεύμονα και δεξιοκαρδία. Περιγράφεται ενήλικας ασθενής με μερική ανώμαλη σύνδεση πνευμονικών φλεβών στην κάτω κοίλη φλέβα και υποπλαστικό δεξιό πνεύμονα που διέλαθε διάγνωσης για έτη και, τελικά, αντιμετωπίστηκε επιτυχώς χειρουργικά.

Λέξεις ευρετηρίου: Ακτινογραφία, Διαθωρακικό υπερηχοκαρδιογράφημα, Διοισοφάγειο υπερηχοκαρδιογράφημα, Σύνδρομο scimitar, Υπολογιστική τομογραφία

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