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Scimitar Syndrome: A Fortuitous Diagnosis in an Asymptomatic Adult

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Abstract | **Case Report**

Scimitar syndrome is a group of cardiopulmonary malformations where the most prominent anomaly is an abnormal venous return affecting mostly the right lung. There are two clinical presentations: infants and adult forms, where the first one is the most severe. The diagnosis is suspected initially by a chest x-ray and confirmed by chest CT-angiography or MR-angiography. Cardiac examinations can help detect associated malformations. The treatment depends on the clinical form and the associated abnormalities, and can range from a simple monitoring to surgical intervention. We report the case of a 45-year-old woman, consulting for an isolated dry cough, whose chest x-ray showed a hypoplastic right lung, with displacement of the heart to the right side, and abnormal retro-cardiac opacity. complementary chest CT-angiography revealed an abnormal dilated vein joining the inferior veina cava as well as other features that allowed us to diagnose the patient with a scimitar syndrome.

Keywords: Scimitar Syndrome, Cardiopulmonary Anomalies, Incidental Findings, Chest Ct-Angiography, Chest Radiography.

INTRODUCTION

Scimitar syndrome, also known as Felson's venolobar syndrome, is defined by the association of multiple cardiopulmonary anomalies, where the main feature is the presence of an abnormal venous return from a part of the right lung to the inferior vena cava or, rarely, to the hepatic veins [1]. The case reported in this article concerns an asymptomatic 45-year-old woman diagnosed incidentally with scimitar syndrome.

CASE PRESENTATION

We report the case of a female patient, aged 45, who presented with a dry cough persistent for five days, with no spontaneous improvement. The clinical examination was without notable features. A chest X-ray showed a hypoplastic right lung, with displacement of the heart to the right, and a right retro-cardiac opacity (figure 1).

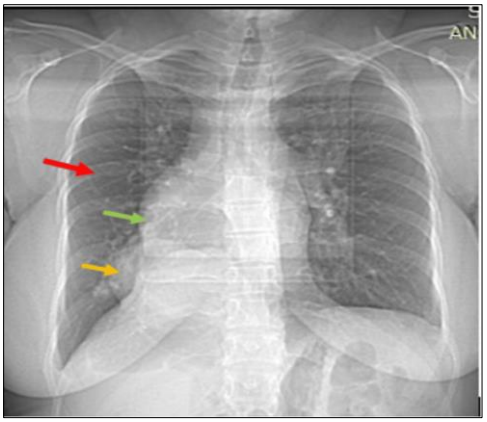

Figure 1: Chest X-Ray of the patient showing a hypoplastic right lung (red arrow), displacement of the heart to the right side (green arrow), and a right retro cardiac opacity (orange arrow)

In view of these findings, a thoracic CT angiography was ordered, confirming hypoplasia of the right lung, dextroposition of the heart to the right which is characterized by a normal appearance of the cardiac chambers contrary to the dextrocardia, and an abnormally dilated pulmonary vein, joining the inferior vena cava at the supra-diaphragmatic level just before its junction with the right atrium (figure *2* and figure *3*). We also note the presence of systemic arterial branches coming directly from the inferior thoracic aorta and the abdominal aorta, vascularizing the lower lobe of the right lung (figure *4*). Finally, we note the presence of a right diaphragmatic hernia with hepatic content which is the origin of the retro-cardiac opacity previously seen on the chest X-ray. Echocardiography didn't find additional malformations or pulmonary hypertension. Based on these findings, the patient was diagnosed with scimitar syndrome and referred to the cardiology department, where she received symptomatic treatment for her dry cough with a good clinical evolution, as well as periodic monitoring to assess her condition and detect the onset of complications.

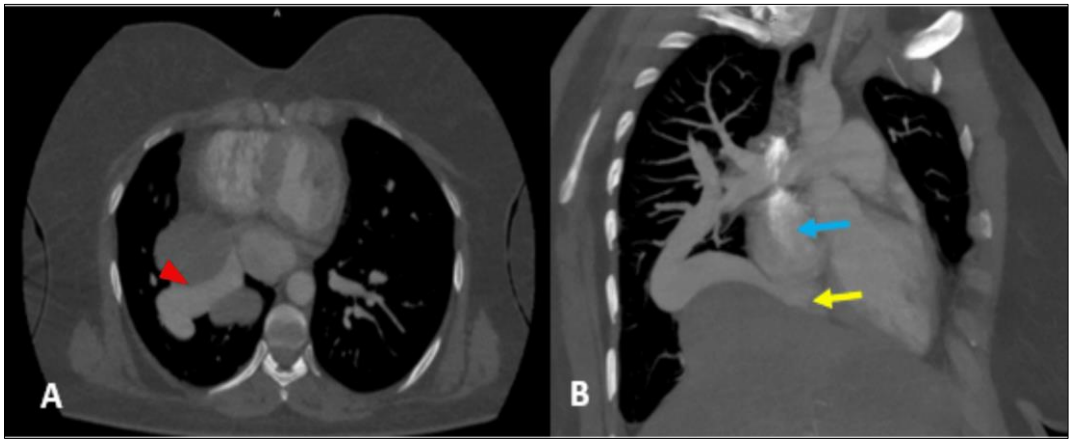

Figure 2: (A): axial slice of a thoracic CT-Angiography showing a dilated right pulmonary vein crossing above the diaphragmatic hernia (red arrow head). (B): coronal image with maximum intensity projection (MIP) of a thoracic CT-Angiography revealing the dilated pulmonary vein joining the IVC (yellow arrow) before its junction with the right atrium (blue arrow)

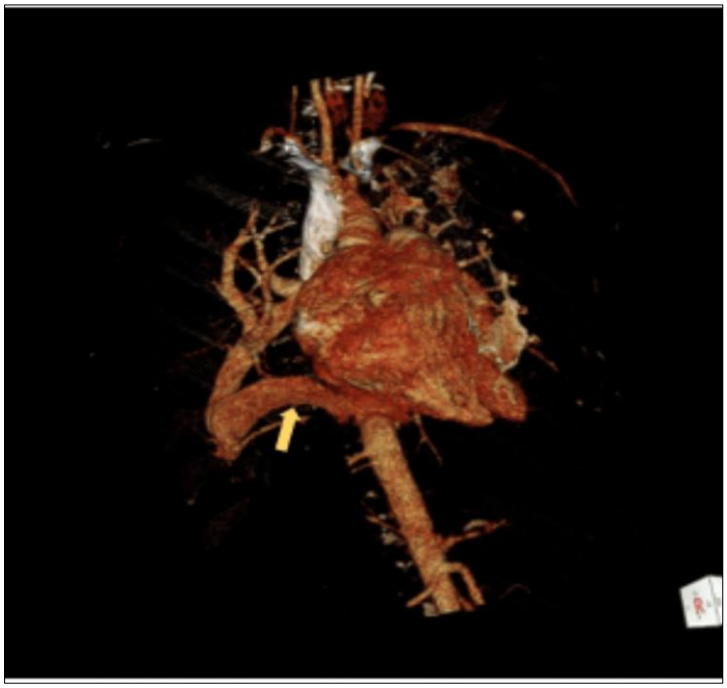

Figure 3: 3D reconstruction of the anomalous right pulmonary vein (yellow arrow)

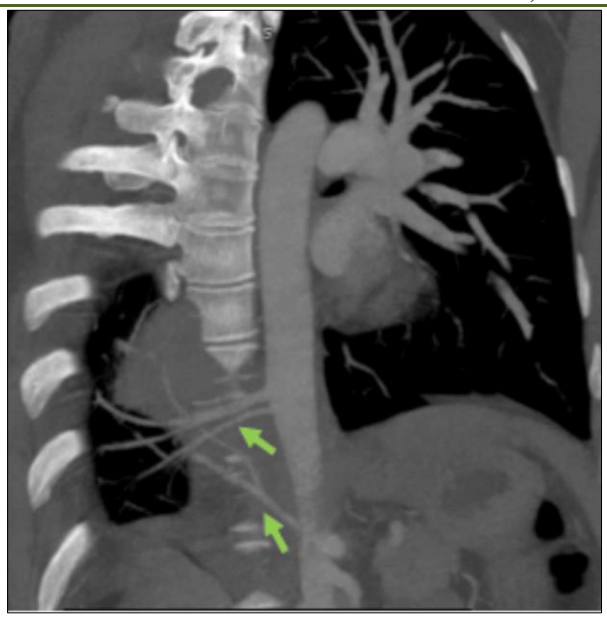

Figure 4: Adjusted coronal image with MIP of a chest CT-Angiography showing the systemic branches arising directly from the thoracic and abdominal aorta, and vascularizing the inferior right lobe (green arrows)

DISCUSSION

Scimitar syndrome, also known as pulmonary venolobar syndrome, was described for the first time in 1836 by Cooper and Chassinat, but the term "Scimitar" wasn't introduced until 1956 by Halasz and al, based on the resemblance of the anomalous pulmonary venous return “APVR” to a Turkish sword [1, 2]. It is a rare condition, primarily affecting female newborns, and its incidence ranges from three to ten in 1,000,000 live births [3, 4].

The characteristic of Scimitar Syndrome is anomalous pulmonary venous return, which most commonly affects the right lung. Rarely, the left lung can also be involved. The abnormal pulmonary veins drain into the superior vena cava or inferior vena cava or rarely the hepatic veins [5, 6]. When these veins are connected with the IVC, the connection site is more frequently subdiaphragmatic, but can also be supradiaphragmatic [7].

There is a wide variability in clinical manifestations, which vary according to the period of onset of symptoms. Infantile forms have a higher risk of morbidity and mortality, and are associated with congenital malformations. They can also present a congestive heart failure, pulmonary hypertension, and respiratory distress [3-5]. In contrast, adults can be asymptomatic or mildly symptomatic with low mortality and this syndrome is often discovered incidentally during imaging studies, or after recurrent respiratory infections, or interstitial pulmonary disorders [3].

Imaging Studies are primordial to the diagnosis, and usually, the first step is a chest X-ray which may reveal a curvilinear opacity creating the aspect of the scimitar sign, or evidence of right lung hypoplasia. But CT Angiography or Cardiac MR Angiography are more precise and allow us to confirm the diagnosis by providing detailed visualization of the anomalous venous return. However, it should be differentiated from the meandering right pulmonary vein where the anomalous pulmonary vein connects to the left atrium [1, 2].

In addition, a cardiac Evaluation for patients with this syndrome is mandatory. Transthoracic or transesophageal echocardiography helps to assess the left-to-right shunt and detect additional cardiac anomalies frequently associated with the syndrome like atrial or ventricular septal defects, patent ductus arteriosus, pulmonary vein stenosis, tetralogy of Fallot, and aortic coarctation [5].

Multiple visceral malformations can also be encountered in this syndrome, and may include dextrocardia, defined as a rightward displacement of the heart with normal arrangement of heart chambers, or septal defects, right lung hypoplasia, systemic arterial supply to the right lung by branches of the aorta that penetrate the diaphragm and divide in the lower part of the right lung, and finally Bochdalek hernia [4-6].

Indications for treatment vary according to the patient's clinical situation. Curative treatment based on surgery, is indicated in cases of right heart failure, a pulmonary-to-systemic blood flow ratio > 1,5. Surgery can be extensive, involving implantation of the pulmonary vein in the left atrium and surgery for other cardiovascular malformations [7].

In the presence of anomalous pulmonary venous return associated with hemoptysis, pulmonary

sequestration or recurrent pulmonary infections, lobectomy may be discussed. However, in asymptomatic patients, simple surveillance appears to be the best therapeutic option [6, 7].

CONCLUSION

Scimitar Syndrome remains a rare congenital anomaly, with its severity and presentation varying greatly according to age. Nowadays, an early diagnosis is possible using advanced imaging techniques and management range from simple monitoring in asymptomatic patients to surgical intervention which are the key to manage severe cases effectively.

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