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An Incidental Diagnosis of a Rare Case of Scimitar Syndrome in a Female

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Abstract

Scimitar syndrome is a rare birth defect characterized by abnormal right pulmonary venous return to the inferior vena cava. It can be partial (PAPVR) or total (TAPVR). Anomalous right-sided pulmonary veins might drain into the superior vena cava, azygos vein, inferior vena cava, or less commonly into the right atrium. Two clinical forms can be distinguished: an infantile form with severe pulmonary arterial hypertension (PAH) and heart failure, with a poor prognosis; and a pediatric or adult form without PAH, well tolerated and with a good prognosis. We report a case of accidentally discovered scimitar syndrome in a 66-year-old patient with dyspnea on exertion as a symptom. The diagnosis was suspected on the chest x-ray and confirmed on a CT scan which showed a single large right pulmonary vein emptying into the right atrium associated with dextro cardia and pulmonary sequestration. Our patient was asymptomatic, apart from stage 1 dyspnea, that's why therapeutic abstention and monitoring were indicated for her.

Keywords: ***Scimitar syndrome; Anomalous pulmonary venous return; Respiratory system abnormalities; CT scann***

1. Introduction

Scimitar Syndrome is a rare congenital disorder which consists of an anomalous pulmonary venous return. It can be partial (PAPVR) or total (TAPVR). Anomalous right-sided pulmonary veins might drain into the superior vena cava, azygos vein, inferior vena cava, or less commonly into the right atrium.

2. Case Summary

This is the case of a 66-year-old patient with no medical history presenting with a few months of exertional dyspnea. Chest radiographic findings include cardiomegaly and opacity at the right posterior lower zone. CT angiography showed a mediastinal shift to the right, main pulmonary artery dilation with a diameter of 42 mm, a partial abnormal pulmonary venous return

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(APVR) with connection of the right pulmonary artery to the inferior veina cava, agenesis of the right upper lobe. It isassociated with a decrease in the volume of the right lung with a pulmonary hyperlucency and trans-mediastinal hernia (FIG. 1).

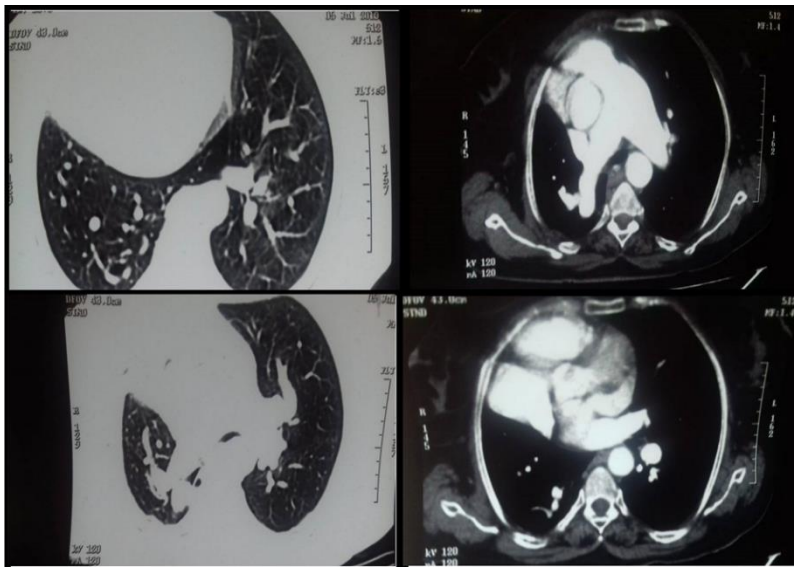

FIG. 1**. CT angiography: a mediastinal shift to the right, main pulmonary artery dilation with a diameter of 42 mm, a partial abnormal pulmonary venous return (APVR) with connection of the right pulmonary artery to the inferior veina cava, agenesis of the right upper lobe.**

Trans-thoracic-echocardiography showed signs of chronic pulmonary heart disease with a mild left ventricular systolic dysfunction. No mitral or aortic valve dysfunction was detected, and the systolic pulmonary artery pressure was 30 mmHg. Our patient was asymptomatic, apart from stage 1 dyspnea, that's why therapeutic abstention and monitoring were indicated for her. No worsening of her condition was noted with two years of follow up.

3. Discussion

Scimitar syndrome; also known as congenital venolobar syndrome, Halasz syndrome [1]. It is a rare clinical syndrome which consists of anomalous right pulmonary venous return to the inferior vena cava. Scimitar syndrome classically involves heart dextroposition, broncho-pulmonary sequestration, bilobar right lung, and/or congenital heart defects such as interatrial communication [2]. The chest-radiography show the characteristic appearance of scimitar sign which is a curvilinear density in the right middle and lower pulmonary fields resembling a curved Turkish sword. The overall prevalence among new born infants vary from 1/100000 to 1/33000. Patients are predominately female. The causes are not fully understood. Abnormal pulmonary venous return associated to lung sequestration was found in 50% of patients with scimitar syndrome [3]. The systemic artery most commonly arises from the lower part of the thoracic aorta and less commonly from the abdominal aorta [4].

Systematic screening for lung sequestration is imperative before indicating a surgical treatment [5]. The clinical expression of the disease is highly variable from a severe presentation from the first days of neonatal life to completely clinically

unrecognized asymptomatic form in adults. It depends up on the degree and extent of the hyperplasia and the arteriovenous fistulation through the sequestration [6]. In most cases, it presents in the neonatal period with a congestive heart failure due to pulmonary hypertension and respiratory failure[4]. The diagnosis is usually suspected from the chest radiographic image of a right basilar curve-shaped opacity extending from the perihilar region towards the diaphragm. Clinical investigations include morphological and functional assessment to detect other abnormalities and determine the impact of APVR on the right ventricular function. The gold standard for the lung tissue and trachea-bronchial tree morphological study and the diagnosis of pulmonary arterial and systemic abnormalities, is today the CT angiography with volume rendering. The functional assessment is evaluated by measuring the pulmonary to systemic flow ratio usually determined by echocardiography. In recent years, cardiovascular flow measurement with phase-contrast MRI may have major role in the evaluation of these two flows [7], by comparing pulmonary arterial flow and aortic flow. The curative treatment of scimitar syndrome in some cases involves combined surgery: redirection of right pulmonary veins to the left atrium and repairing other cardiovascular malformations like resection of pulmonary sequestration [3] and closure of interatrial communication. The surgical repair of total anomalous pulmonary venous return depends on finding a right heart failure and a pulmonary to systemic flow ratio greater than two [8]. In our case the patient was asymptomatic, apart from stage 1 dyspnea, that's why therapeutic abstention and monitoring were indicated for her.

4. Conclusion

Scimitar syndrome is an exceptional, unrecognized disease whose clinical expression is insidious and nonspecific. The chest x-ray often helps to suggest the diagnosis, which will be confirmed by CT angiography, pulmonary angiography or MRI. No treatment is necessary for asymptomatic patients; however, surgical treatment may be offered in case of a severe left right shunt, sequestration or recurrent pulmonary infections.

5. Funding

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6. Conflicts of Interest

Not declared.

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