Respir Case Rep 2024;13(3):141-144 DOI: 10.5505/respircase.2024.36926

Scimitar Syndrome Diagnosed in Adulthood

Erişkin Dönemde Tanı Konulan Scimitar Sendromu

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Abstract

Scimitar syndrome is a congenital heart anomaly characterized by the drainage of one or more accessory veins, usually from the right pulmonary veins, into the right atrium. We present here the case of a 44-year-old patient who was followed up with a diagnosis of asthma but whose dyspnea continued despite treatment and who was subsequently diagnosed by thoracic tomography. It is important to perform additional investigations during the differential diagnosis of patients with respiratory symptoms.

Keywords: *Scimitar syndrome, partial pulmonary venous return anomaly, differential diagnosis of asthma.*

Öz

Scimitar sendromu, doğumsal bir kalp anomalisi olup, genellikle sağ pulmoner venlerden bir veya daha fazla aksesuar venin sağ atriyuma drenajı ile karakterizedir. Bu yazıda, 44 yaşında astım tanısı ile takip edilen ancak tedaviye rağmen nefes darlığı devam eden ve toraks tomografisi ile tanı konulan hasta sunulmuştur. Solunum semptomları nedeniyle takip edilen hastalarda ayırıcı tanıda ek incelemelerin yapılması önem arz etmektedir.

Anahtar Kelimeler: *Scimitar sendromu, parsiyel pulmoner venöz dönüş anomalisi, astım ayırıcı tanısı.*

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Submitted (Başvuru tarihi): 27.02.2024 ***Accepted (Kabul tarihi):*** 26.07.2024

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Scimitar syndrome is a congenital cardiac anomaly characterized by drainage of one or more accessory veins from the right pulmonary veins into the right atrium (1). In such cases, venous drainage of the right lung is abnormally sent to the right system, leading potentially to such symptoms as pulmonary hypertension, heart failure and severe respiratory distress (2).

Scimitar syndrome is usually diagnosed in childhood, with such symptoms as respiratory and cardiac problems such as respiratory distress, recurrent infections and impaired growth (3). Although the number of cases diagnosed in adults is low, it should be noted that the condition can also occur in adulthood (4).

CASE

A 44-year-old female patient who had been followed up with asthma for 1.5 years presented to our pulmonology outpatient clinic with exertional dyspnea, despite treatment. Head and neck, cardiac and respiratory examinations were normal. One-way posteroanterior chest radiography and pulmonary function test with reversibility were ordered for the patient, who had been followed up in an external center with a diagnosis of asthma, with the following results: FEV1: 65%, FVC: 82%, FEV1/FVC 68% and negative reversibility. The patient had no history of smoking, although chest radiography showed volume loss in the right lung (Figure 1). Thorax CT was ordered for further examination, the results of which pointed to Scimitar syndrome, with decreased right hemithorax volume (Figure 2), and the drainage of some pulmonary veins in the right hemithorax into the hepatic vein (Figure 3). The patient was referred to the cardiology outpatient clinic for further investigation and treatment, where a patent foramen ovale and thrombus in the interatrial septum were identified on echocardiography. A decision was made to continue with an anticoagulant, and for intermittent follow-up with echocardiography.

DISCUSSION

Scimitar syndrome, referred to also as pulmonary venous drainage anomaly, is a rare congenital cardiovascular malformation characterized by the partial or complete drainage of the right-sided pulmonary veins into an anomalous venous channel, typically the inferior vena cava (5). In our case, in the right lung, vena cava drainage was observed. Scimitar syndrome occurs in 3/100,000 cases, however, the number of cases detected does not reflect the real numbers due to its asymptomatic course (6,7). Scimitar syndrome is characterized by the following unique features: partial or total abnormal curved venous drainage of the right lung into the inferior vena cava; correlation with hypoplasia of the pulmonary arteries and varied right lung; dexterity of the heart; and unusual blood supply to the ipsilateral lung throughout the body (8). In our case, hepatic vein drainage was noted in the right lung and pulmonary artery hypoplasia, although the heart was normally located on the left.

The clinical presentations of Scimitar syndrome can be widely varied, ranging from asymptomatic incidental findings to severe respiratory distress and pulmonary hypertension. Heart abnormalities can lead to various symptoms, and diagnosis is typically made in childhood (infantile form). In those who exhibit the infantile form, the condition is typically associated with pulmonary hypertension and has a worse prognosis due to the greater severity of the illness. Adult diagnoses (adult version) are less common, and patients typically present with less severe symptoms or not at all (9). Our case was admitted with the complaint of shortness of breath.

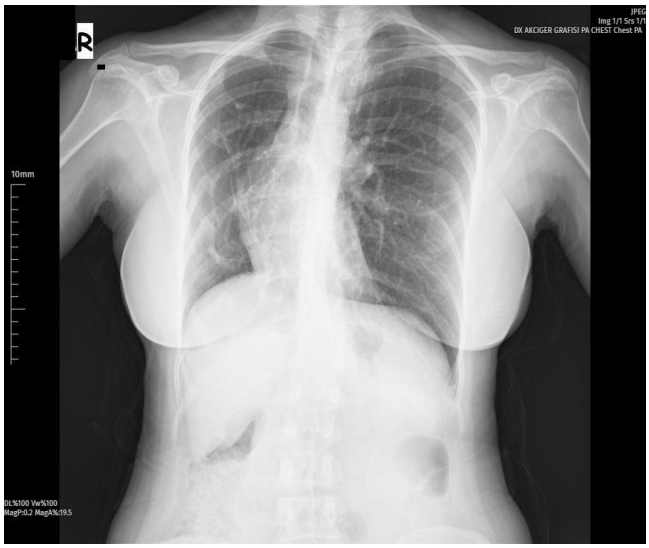

Figure 1: Chest X-ray image taken at the time of application revealing decreased right hemithorax volume and the deviation of the mediastinum to the right

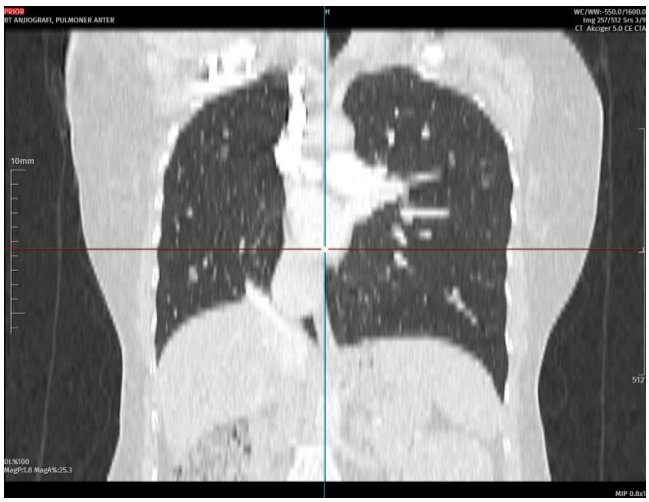

Figure 2: Patient's thorax CT parenchyma images showing the mediastinal structures displaced to the right. Evaluation of the sections in the lung parenchyma window revealing reduced right hemithorax volume

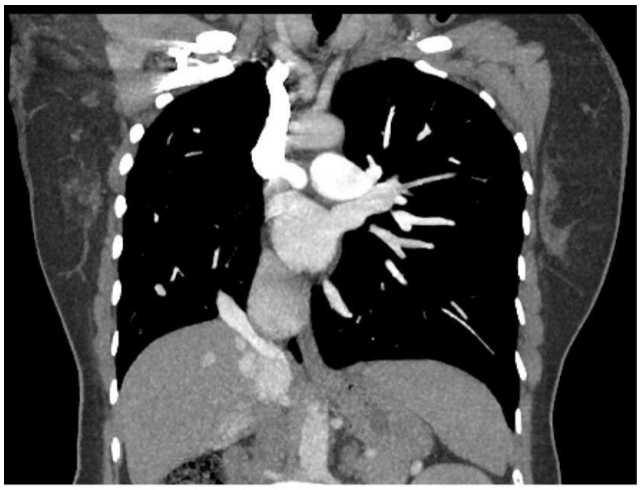

Figure 3: Mediastinal structures displaced to the right. The sectional evaluations in the lung parenchyma window reveal the right hemithorax volume to be reduced. In the right hemithorax, the pulmonary veins drain into the hepatic vein. This appearance is typical of Scimitar syndrome

Echocardiography is the primary diagnostic modality and is often supported by advanced imaging techniques such as computed tomography or magnetic resonance imaging for the delineation of anatomical details. In some cases, anomalous pulmonary venous drainage may not be visually apparent, necessitating the use of contrast echocardiography to confirm the diagnosis (10). The diagnosis of our case was made without the need for contrast-enhanced echocardiography.

The accurate diagnosis and prompt management of Scimitar syndrome are crucial, as significant morbidity and mortality can develop if left untreated (9). The management of Scimitar syndrome is complex and may require a multidisciplinary approach. Surgical correction may be necessary in patients with significant hemodynamic compromise or pulmonary hypertension, typically involving the redirection of the anomalous pulmonary venous drainage to the left atrium. Surgical interventions can be a safe and effective approach to some conditions, and are most commonly performed in children. Adult-age therapies may also be needed, especially in symptomatic patients (5). Our case was also decided to be followed up due to a mild clinical course.

CONCLUSION

Scimitar syndrome is a rare cardiac anomaly that may also be detected in patients presenting with respiratory symptoms. For this reason, angiographic and cardiologic investigation of patients with suspected Scimitar syndrome is vital. Respiratory diseases such as asthma and chronic obstructive pulmonary disease in patients with respiratory symptoms tend to be prioritized due to their frequency, although additional investigations such as pulmonary function tests and chest radiographs are important in a differential diagnosis. As Scimitar syndrome can lead to serious complications if left untreated, accurate and early diagnosis are important for treatment and follow-up. Patients diagnosed with Scimitar syndrome should be treated with a multidisciplinary approach and subjected to long-term follow-up.

In our study, the appropriate evaluations and examinations were performed in all branches, without limitations, although echocardiography images of the patient could not be obtained due to technical problems.

CONFLICTS OF INTEREST

None declared.

AUTHOR CONTRIBUTIONS

Concept - G.K., K.C., O.A., H.D., H.Ö.; Planning and Design - G.K., K.C., O.A., H.D., H.Ö.; Supervision - G.K., K.C., O.A., H.D., H.Ö.; Funding - G.K., K.C., O.A., H.D., H.Ö.; Materials - G.K., K.C., O.A., H.D., H.Ö.; Data Collection and/or Processing - G.K., K.C., O.A., H.D., H.Ö.; Analysis and/or Interpretation - G.K., K.C., O.A., H.D., H.Ö.; Literature Review - G.K., K.C., O.A., H.D., H.Ö.; Writing - G.K., G.K., K.C., O.A., H.D., H.Ö.; Critical Review - G.K., K.C., O.A., H.D., H.Ö.

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