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Case Report

Asymptomatic Scimitar Syndrome in Adults

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Abstract

Scimitar syndrome, also called hypogenetic veno-lobar or right lung syndrome, is characterized by abnormal right pulmonary venous return in the inferior vena cava [2]. This syndrome is rare, not well known. Anomalous pulmonary venous drainage was first described in 1836 by Cooper and Chassinat. In 1949, Dotter, first described the radiological signs of Scimitar syndrome, Halasz, first used the word "Scimitar" in 1956 The term "Scimitar syndrome" was first given and described by CA Neill in 1960 [1]. It is essentially a combination of pulmonary hypoplasia and partial abnormal pulmonary venous return (RVAP). This happens almost exclusively on the right side. Hemodynamically, there is a cyanotic shunt from left to right. The abnormal vein usually discharges, inferior vena cava more common, right atrium or portal vein. It mainly affects female births, clinically there are signs of heart failure especially infants, which may be due to an associated congenital heart disease, adults are most often asymptomatic but may present with recurrent lung infections or dyspnea on exertion. The incidence ranges from 1/100,000 to 1/330,000 live births [3]. The exact causes of scimitar syndrome have not yet been known. More associated with familial risk factor with abnormality on chromosome 4q12 [4]. Diagnosis is made by transthoracic or transesophageal echocardiography, angiography, or CT or MRI angiography. Chest X-ray shows a small hypoplastic lung with ipsilateral mediastinal displacement and, in a third of cases, the abnormal drainage vein can be seen as a tubular structure parallel to the right edge of the heart in the shape of a Turkish sword (scimitar), the right edge of the heart may be blurred. The objective of this study is to show some parenchymal and vascular anatomical variants associated with Scimitar syndrome with slight symptomatology in adulthood.

Keywords: Scimitar syndrome; Unilateral pulmonary hypoplasia; tronc céliaque; Adult; asymptomatic; CT

Clinical Image

It is a 30-year-old lady, with a history of lymph node tuberculosis diagnosed 9 months ago and undergoing treatment , was presented to our department for dry and chronic cough, physical examinations were in favor of increased liver volume (hepatomegaly), REVOIR LES TERMES EN ANGLAIS →An echogram was done that aimed at a portal hypertension, a thoracic scanner was performed showing the following findings: a hypoplic right lung, deviation of mediastinum to the right, abnormal pulmonary venous gear (pulmonary artery that is injected into the inferior vena cava **(Figure 1)**, associated with parenchymal and vascular variants such as: a tongue of the right lung parenchyma passing between the aorta and the heart appearing in confluence with the left lung in relation to horseshoe lung et a branch of celiac trunk communicates with right pulmonary artery **(Figure 2)**.

An Rx of throrax was requested with individualization of a right paracardiac opacity oblique downwards and inwards with discreet mediastinal enlargement in relation to which it receives afferents giving an appearance of the Turkish sword **(Figure 3-5)**.

Discussion

There are 4 normal pulmonary veins, two on each side, which end in the left atrium. It is important to know that there are many variants in 30% of cases [6]. Scimitar syndrome is a rare pathology, characterized by hypoplasia of the right lung, which is a form of congenital malformation of pulmonary development that can be partial (most compatible with life) as is the case of our patient or complete (not compatible with life) of frequent form results clinically in respiratory disease recur and radiologically it presents as a pulmonary field affected by reduced volume, Pulmonary hypoplasia may be visible radiologically as a small unilateral lung [5].

Diagnosis is often made by chest CT angiography with 3D reconstruction or cardiac MRI angiography. For venous return abnormalities, frequently to the inferior vena cava with more than 50% followed by superior vena cava, the right atrium, or at the level of the abdominal aorta and its dividing branches and other cardiac malformations may be associated such as coartation of the aorta, tetralogy of Fallot, pulmonary sequestrations up to atrial or ventricular septal defect, These are frequent abnormalities in the neonatal period with variable clinical symptoms characterized by the volume of blood circulating

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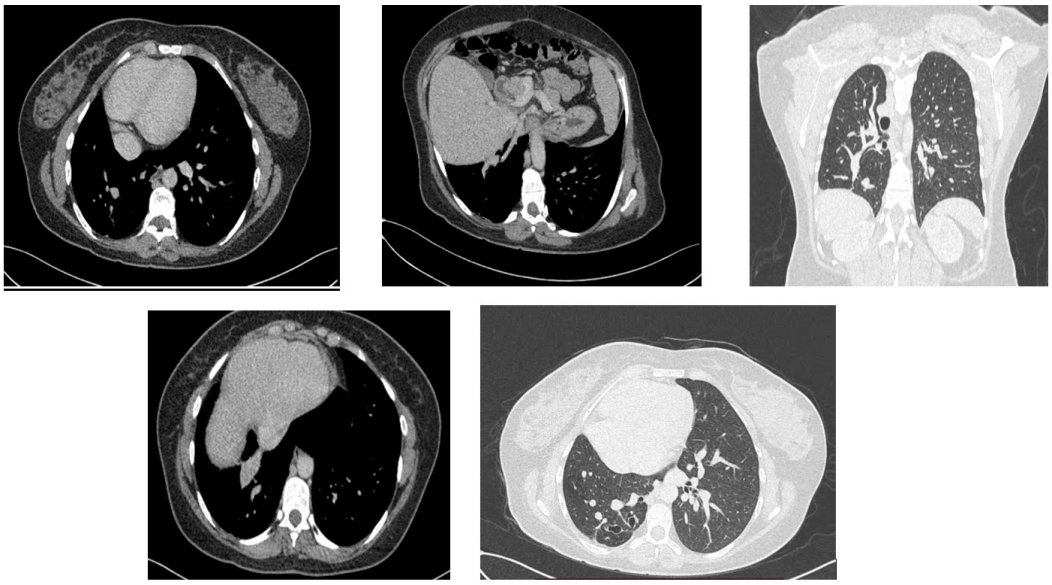

in the veins abnormal which, following severe in infants and asymptomatic in adults, varies other visceral malformations are possible affecting the diaphragm (Bochdalek hernia) [6]. Our patient has abnormal venous return that can be significant with the portal trunk branch communicating with the right pulmonary artery which are also of small caliber, which is why it remains asymptomatic with incidental findings.

Conclusion

Scimitar syndrome is an exceptional, little-known disease that is often asymptomatic. the diagnosis is evoked by a right para cardiac opacity and confirmed by thoracic CT angiography and sometimes MRI angio.

References

1. Scimitar syndrome radiopaedia.
2. Upuisj CD, Remy J, Remy-Jardine M, Reye C, Breviere GM, Kerkoub A. Syndrome du cimeterre avec absence anatomique ou fonctionnelle de l'artère pulmonaire droite. A propos de quatre observations.
3. Nesrine Fahem, Saoussen Cheikh Mhamed, Ahmed Ben Saad, Ghassen Trigui, Asma Migaou, Ahmed Zrig, et al. Syndrome de cimeterre : à propos d'une observation. Scimitar syndrome. A case report.
4. Cristiano-Antonino. Syndrome Du Cimeterre: Causes, Symptômes, Diagnostic, Traitement, Pronostic Et Mortalité, 2022.
5. The African Médical journal cause inhabituelle d'une infection respiratoire récidivant: hypoplasie du poumon gauche.
6. Doyen G Giraud. Les anomalies des retours veineux pulmonaires ; magali saguintaah, catherine baud, alain couture, corinne veyrac, sabrina flunker, jean-louis ferran : Radiologie pédiatrique - Hôpital Arnaud de Villeneuve - 371 av.