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RESEARCH ARTICLE

PARTICULARITIES OF SCIMITAR SYNDROME IN ADULT PATIENTS

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Abstract

Scimitar syndrome or Felson's veno-lobar syndrome is a very rare congenital disease characterized by a combination of cardiopulmonary abnormalities, including partial right-sided pulmonary venous drainage to the inferior vena cava, the inferior cavo-atrial junction, or low on the right atrium. We report the case of a 53-year-old female patient who presented with recent gradually worsening dyspnea. The diagnosis was suspected on the chest x-ray and confirmed on Cardiac echography andComputed Tomography scan that showed a wide collector gathering the three right superior pulmonary veins that joins the lower part of the superior vena cava, thus joining the right atrium; while the right inferior pulmonary vein is drained into the inferior vena cava. The patient was treated surgically by performing a derivation of the right superior pulmonary venous collector to the left atrium with a tricuspid annuloplasty with a good outcome.

Introduction:-

Scimitar syndrome (SS) is a rare congenital anomaly characterized by a partial (usually the right lower lobe) or complete unilateral anomalous pulmonary venous drainage in association with ipsilateral lung hypoplasia [1].The anomaly, also known as pulmonary venolobar syndrome or hypogenetic lung syndrome, was first described in 1836 [2]

The term scimitar was first used by Halasz et al (1956) [3] to describe the radiographic appearance of the anomalous pulmonary vein trajectory that drains into the inferior vena cava just below or above the right hemidiaphragm as a broad, gently curved shadow resembling the silhouette of a short, curved Persian sword known as a shimshir. In 1960 [4].The familial occurrence and the clinical spectrum were defined first using the term scimitar syndrome.

Case report:

A 53-year-old female patient with no history of personal or familial diseases; she carried to term 5 pregnancies, with vaginal delivery with no complications.She presented with recent gradually worsening dyspnea, dry coughs, palpitations and asthenia.The physical examination was without abnormality.

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EKG showed a regular sinus rhythm, an average ventricular rate at 84bpm, and slightly deviated axis to the right with signs of right ventricle hypertrophy; Incomplete right bundle branch block. We noticed during monitoring the patient some transitional passages to atrial fibrillation.

The chest x-ray showed cardiomegaly with a diaphragmatic heart tip, a discreet right paracardiac arciform opacity with a right hilar overload overflow and straight left middle arch,

The echocardiography showed severely dilated right cavities (figure 1), a dilated tricuspid ringwith significant tricuspid insufficiency.

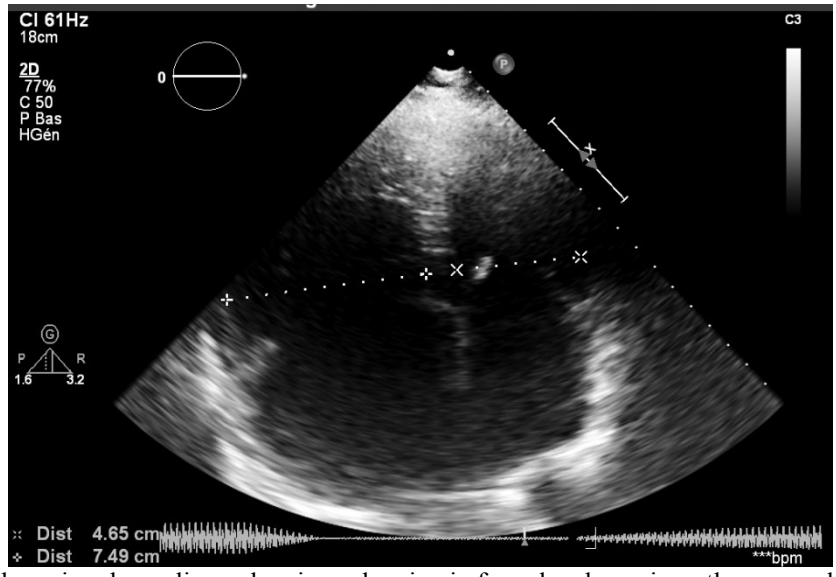
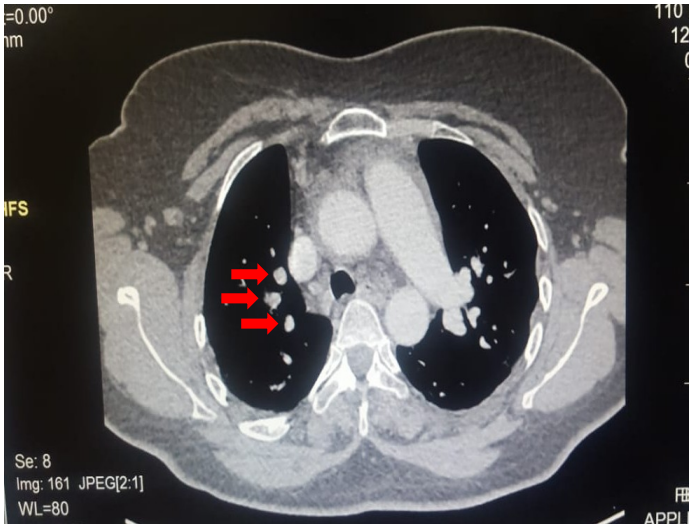
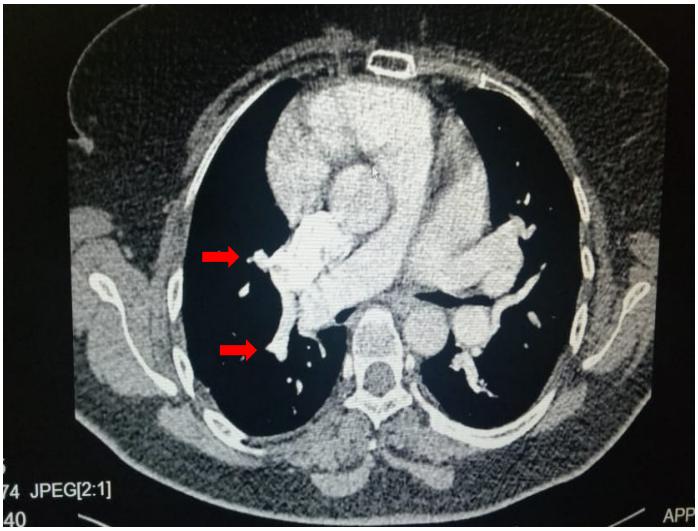


Figure 1:- Transthoracic echocardiography views showing in four chambers views the severe dilation of the right chambers.

The left ventricle was of normal size and function.The Computed Tomography scan (figure 2) showed a wide collector gathering the three right superior pulmonary veins that joins the lower part of the superior vena cava, thus joining the right atrium; while the right inferior pulmonary vein is drained into the the retro-hepatic portion of the inferior vena cava.





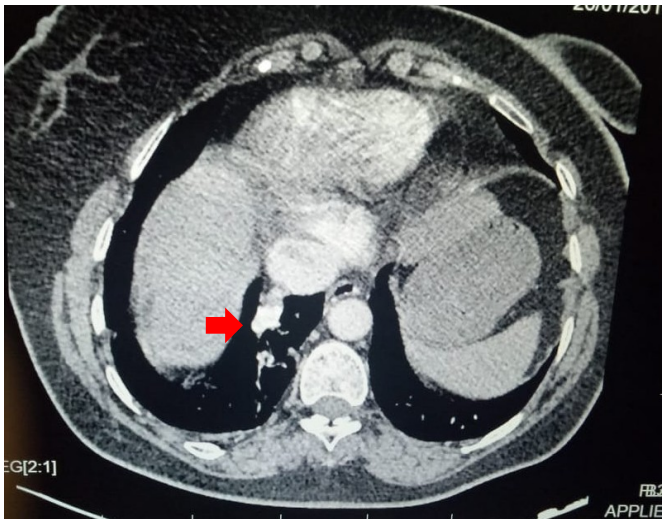


Figure 2:- Axial section of thoracic CT angiography showing in mediastinal view, the wide collector that drains the three right superior pulmonary veins joining the lower part of the superior vena cava; while the right inferior pulmonary vein is drained into the inferior vena cava.

The patient was treated surgically by performing a derivation of the right superior pulmonary venous collector to the left atrium with tricuspid annuloplasty by an EDWARDS ring No.36; after good clinical evolution, she was discharged 10 days later without any complications with uneventful follow-up.

Discussion:-

The scimitar syndrome or veino-lobar syndrome is defined as a partial or total anomalous pulmonary venous return of the right lung veins to the inferior vena cava (IVC), the inferior cavo-atrial junction, or low on the right atrium. In 2/3 of cases the Scimitar vein provides drainage for the entire right lung, but in 1/3 the SV drains only the lower portion of the right lung with normally connected upper pulmonary vein [1].

The SS is a rare sporadic syndrome with a frequency of 1 in 2–3 million, with female preponderance.The etiology is not completely understood. The responsible gene locus was located on chromosome 4q12.

Most of the time, the SS is associated with other malformations; an abnormal right lung lobation and right lung hypoplasia (virtually 100%, with widely varying degrees of hypoplasia), dextroposition of the heart, hypoplasia of the right pulmonary artery (60%), systemic arterial blood supply to the right lower lung from the infra-diaphragmatic aorta (60%), atrial septal defect (ASD) of the secundum type (40% overall, 80% to 90 % in the infantile variant), right-sided diaphragmatic hernia (15%) and horseshoe lung [2].

Clinical symptoms vary from severe form in infants associated with significant mortality, to asymptomatic form in adults [5] with diagnosis being made incidentally because of radiographic abnormalities. Some patients can be pauci-symptomatic and present slight dyspnea at exertion, recurrent pulmonary infections, atrial fibrillation associated with chronic right-sided overload; while some other patients are asymptomatic with good tolerance of the SS and often lead normal lives.

Chest X-ray shows the aberrant vein that has convex course in the lower part of the right lung which Neil [4] compared to the shape of the Turkish curved sword, “the scimitar”.

However, this sign is often absent. Reasons cited for the absence include the hypoplasia of the right lung and resultant abnormal rightward positioning of the heart, obfuscating the view of the vein in frontal projection.

Electrocardiography in 50% of cases shows evidence of right ventricular hypertrophy withright bundle branch block.

The echocardiography is used to assess the importance of the left right shunt, do screening for associated malformations that will require simultaneous management; Pulmonary artery pressures are normal or only mildly elevated.

The certainty of diagnostis is often carried by the thoracic CT angiography with 3D reconstruction or by cardiac angio-MRI [6, 7].

Treating Scimitar syndrome in adults can be of debate; it is advised when patients present symptoms of recurrent lung infection;Qp/Qs greater than 1.5 in an asymptomatic patient.

Dupuis et al [8] casted doubt on the utility of corrective surgery in adults with SS while comparing the prognosis of two groups, one treated surgically and the second non-surgically; there were no significant differences related to the severity of the symptoms and the surgical decision depended on the attitude of the medico-surgical team. Good results were reported in the non-surgical (93 %) with less complication during a follow-up of more than 10 years.

The curative treatment of scimitar syndrome involves a corrective approach rerouting SV flow to the left atrium and surgery of other possible associated cardiovascular malformations including surgery of pulmonary sequestration, ligature of abnormal systemic arteries to the right lung and closure of ASD[3]

In cases of hemoptysis complicating sequestration, isolated treatment by radiological vaso-occlusion of the sequestered artery can be proposed in emergency in purely vascular sequestrations.In case of partial venous return of the right lower lobe with complicated sequestration of hemoptysis and infections, a lower lobectomy can be discussed in the absence of scissural abnormality that could make this lobectomy difficult [9]

Conclusion:-

The scimitar syndrome is an exceptional unrecognized disease. Clinical presentation may be insidious with no specific sign. The diagnosis is suggested in front of a right para-cardiac opacity in the chest X-raythat will be confirmed by chest CT scan, pulmonary angiography or MRI. No treatment is required for asymptomatic patients; however, surgical treatment may be offered in cases of severe left-sided shunts, sequestration or repeated pulmonary infections.

Consent:

The authors confirm that written consent for submission and publication of this case report including image(s) and associated text has been obtained from the patient.

Conflict of Interest

The authors declare that they have no conflict of interest to report.

Author Contributions

All authors have contributed to the elaboration of the manuscript.

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