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SCIMITAR SYNDROME ASSOCIATED WITH PATENT DUCTUS ARTERIOSUS AND PULMONARY SEQUESTRATION IN ADULT: A CASE REPORT

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Abstract: Introduction: Scimitar Syndrome is a rare congenital anomaly, where the return of venous drainage from the right lung is presented in an anomalous way to the inferior vena cava. **Goals**: In this study, a case of Scimitar Syndrome was reported in a cardiology unit in the city of Manaus/AM (Brazil).**Methods**: The sample consisted of a female patient who underwent medical treatment at a reference institution in cardiac surgery in Manaus, aged between 50 and 70 years, in the preoperative period of patent ductus arteriosus. **Conclusion**: Scimitar syndrome is a rare disease, with few reports in the medical literature about this pathology, especially with the associations reported, and further studies are needed to assist in the management of this medical entity.
Keywords: Scimitar Syndrome, Pulmonary sequestration, Anomalous pulmonary drainage, Patent ductus arteriosus.

INTRODUCTION

Scimitar syndrome is characterized by anomalous drainage from the right lung into the inferior vena cava. Approximately half of elderly patients diagnosed after the first year of life are asymptomatic and identified by an incidental finding on chest radiography. In others, there may be fatigue, dyspnea, and recurrent pneumonia.

Other common associated abnormalities are atrial septal defect, pulmonary sequestration, hypoplastic lung, dextrocardia, pulmonary vein stenosis, other cardiac anomalies (eg, coarctation of the aorta, ventricular septal defect, patent ductus arteriosus), and in the bronchial tree.

Treatment is based on symptomatology and associated anomalies. Asymptomatic patients will not require surgical intervention, while those with marked shunting of blood to the inferior vena cava and/or pulmonary hypertension will require correction to

redirect venous return to the left atrium of the heart.

CASE REPORT

A 53-year-old patient, in the preoperative period for correction of patent ductus arteriosus, in irregular use of Amiodarone. On physical examination, pulmonary auscultation was physiological and symmetrical breath sounds. On cardiac auscultation, the rhythm was regular, with a murmur in the aortic area and good perfusion of the extremities. On abdominal examination, the abdomen was flat, pitted and with preserved bowel sounds. A chest X-ray was performed, among the tests necessary for cardiac risk, which showed an oval opacity with soft tissue density in the right hemithorax, between the right cardiac margin and the hepatic pocket. It was also noted bulging of the middle mediastinal arch (pulmonary artery trunk) and increased cardiac volume (Figure 1).

Chest tomography revealed some of the following findings: "1. Patent ductus arteriosus, measuring 0.9 x 0.4 cm in diameter (T x W) (Figures 2A and 2B) 2. In addition, dilatation of the pulmonary arteries is observed, its trunk measuring 4.8 cm in diameter (hypertension (Figure 3) 3. Oval image located on the medial postero-inferior aspect of the right hemithorax, surrounded by the pleural surface (medially) and the corresponding lower lobe of the lung (laterally), with soft tissue density and regular contours, measuring 8 9 cm (Figures 4A, 4B and 4C), compatible with intralobar pulmonary sequestration. An arterial vessel communicating this structure with the epigastric aorta (Figure 4D), at the time of emergence of the celiac trunk, measuring 1.1 cm in diameter 4. Anomalous drainage of the right inferior pulmonary vein, which communicates directly with the vena cava inferior and measures 1 cm in diameter, known as the Scimitar vein (Figure 5)."

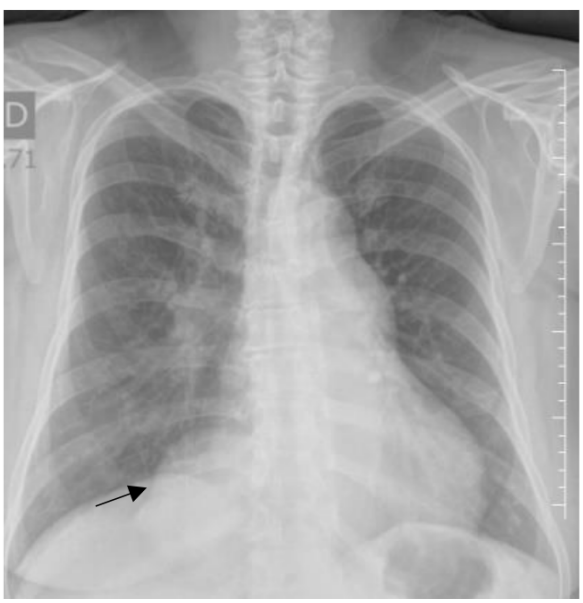

Figure 1: Chest X-ray: Nodular opacity with soft tissue density in the right hemithorax (black arrow), bulging of the middle mediastinal arch (pulmonary artery trunk) and increased cardiac volume.

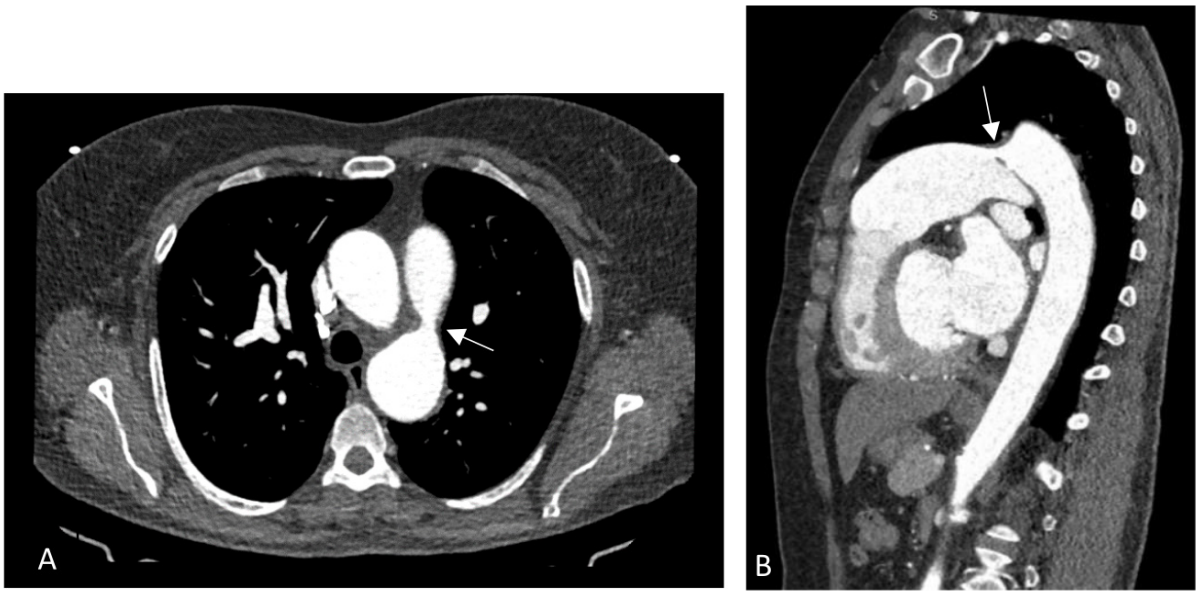

Figure 2: Chest tomography with intravenous contrast. (A) axial plane and (B) sagittal plane, showing patent ductus arteriosus (white arrow).

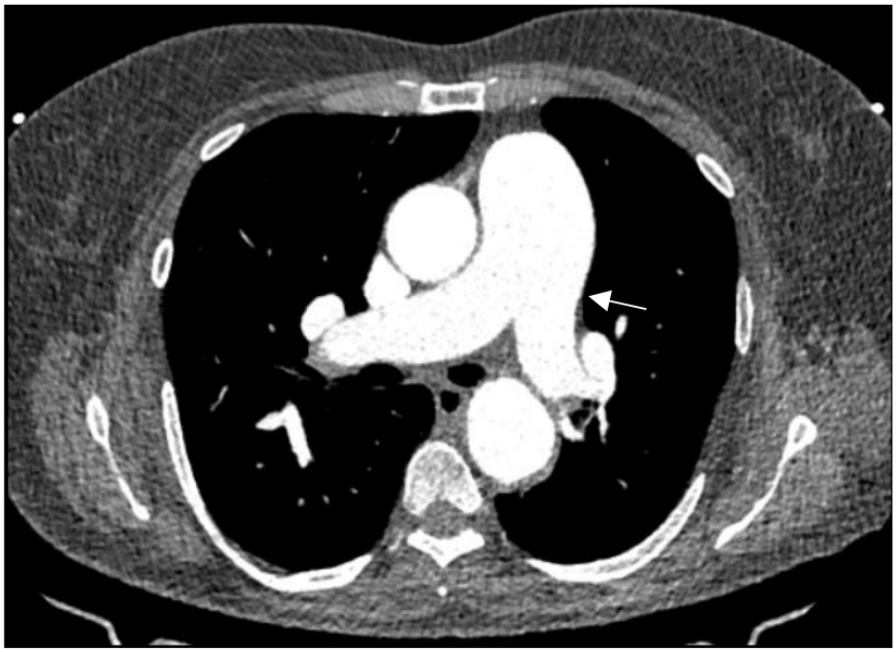

Figure 3: Chest tomography with intravenous contrast. In the axial plane, there is dilatation of the pulmonary arteries (presumed hypertension) – white arrow.

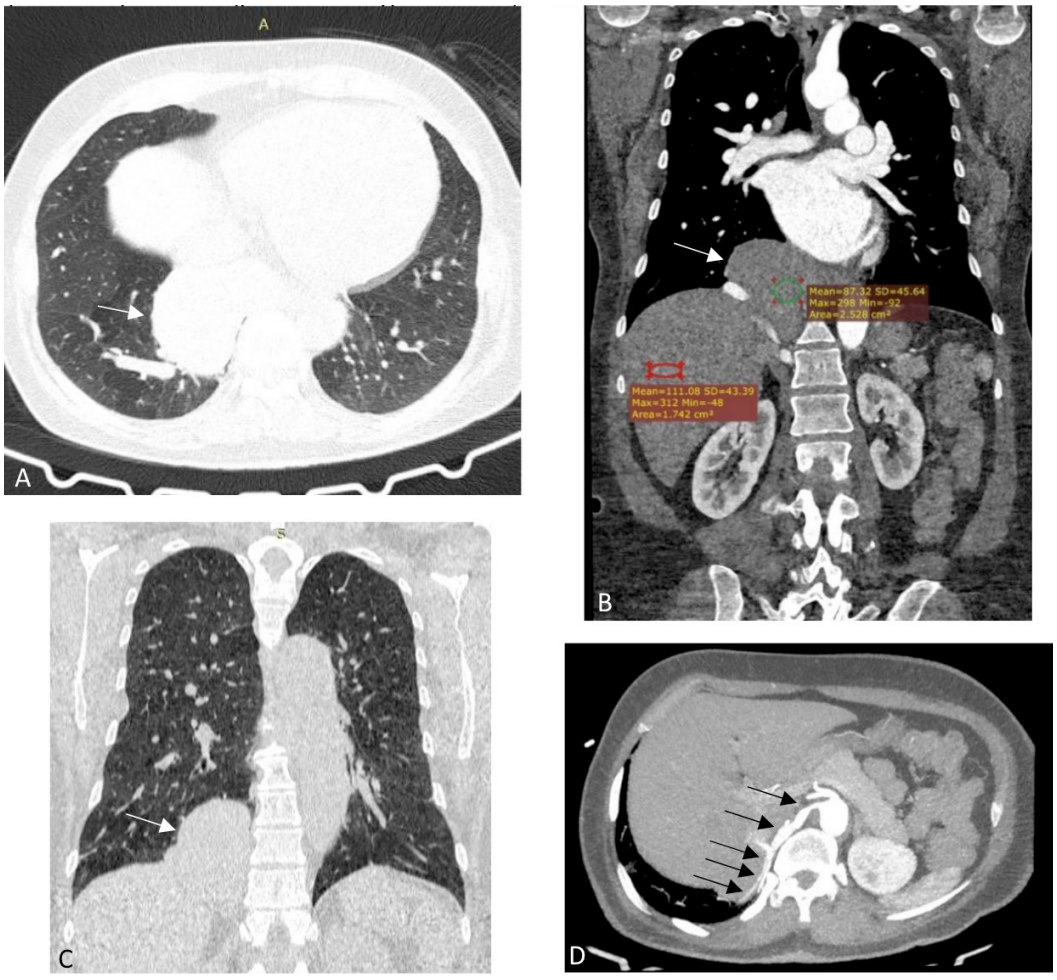

Figure 4: Chest tomography with intravenous contrast. (A) and (D) in the axial planes, and (B) and (C) in the coronal planes, demonstrating the presence of an oval image in the right hemithorax with a density different from that of the liver (white arrow) and being vascularized by a branch coming from the epigastric aorta (black arrows) and having a pleural lining shared with the lung.adjacente.

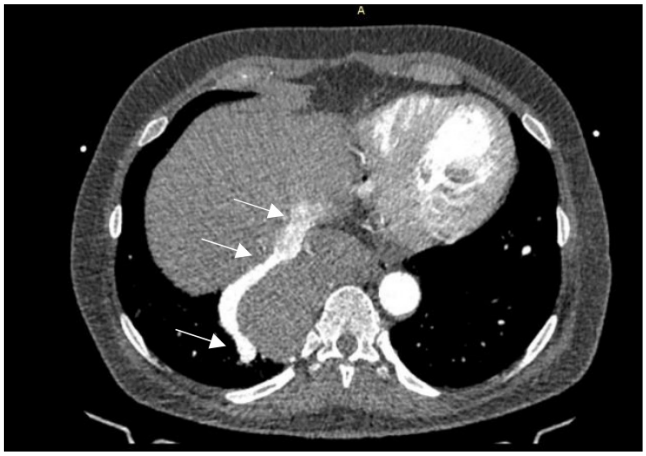

Figure 5: Chest tomography with intravenous contrast. (A) axial plane, showing the presence of anomalous drainage of the right inferior pulmonary vein that communicates directly with the inferior vena cava (Scimitar vein) – white arrows.

The transthoracic echocardiogram performed showed the presence of a patent ductus arteriosus (PDA) measuring 0.8 cm with aortopulmonary shunt, mild pulmonary arterial hypertension (pulmonary artery systolic pressure: 40 mmHg), enlargement of the left chambers, and mitral and aortic regurgitation. and light tricuspid. Cardiac catheterization did not show significant changes.

DISCUSSION

Scimitar syndrome is defined as an anomalous drainage of the right pulmonary vein from the entire right lung or partially into the inferior vena cava (DUPUIS, 1992). Its prevalence is 1-3 of every 100,000 live births, with a predominance of females (CHOWDHURY, 2021).

The first description of this syndrome was made by George Cooper and Raoul Chassinat in 1836, but without using the term "scimitar". This nomenclature comes from the radiological image of a tubular image, downward curve, vertical, resembling the Turkish scimitar sword. In 1960, Catherine Neil was the first to use the vein as a syndrome (MULLIGAN, 1999).

Dupuis et al classified the syndrome into three forms: childhood, with pulmonary hypertension and worse prognosis; the adult, asymptomatically and with a good prognosis, and the form with associated congenital cardiac anomalies (2,3). It may also be associated with pulmonary sequestration, hypoplastic lung, diaphragmatic hernia, genitourinary alterations (VIDA, 2010; BERROCAL, 2004).

The case patient has a patent ductus arteriosus, which is an uncommon cardiac abnormality (9%) in a multicenter study including 68 scimitar patients (VIDA, 2010).

Symptoms depend on the severity of the shunt, ranging from its absence or mild dyspnea in adults to severe pulmonary hypertension in infants.

Surgical correction must be recommended for symptomatic patients in the presence of significant left-to-right shunt and pulmonary hypertension. Surgery in adults is still discussed, due to the low symptomatology of the cases. The type of intervention depends on the associated anomalies, the treatment being to redirect venous return to the left atrium, through an unrestricted path to the scimitar vein. Individually, some drugs such as diuretics, β-agonists, ipratropium bromide, antibiotics and corticosteroids can also be considered (CHOWDHURY, 2021).

CONCLUSION

Scimitar syndrome is a disease characterized by anomalous venous drainage from the right lung to the inferior vena cava. Diagnosis in adulthood is uncommon and most cases are asymptomatic. When symptomatic, which was the case in this report, surgical intervention is safe and effective and must be performed.

This association of Scimitar syndrome with PDA and pulmonary sequestration is uncommon and there are no reports of similar cases, so further studies are needed in this area that describe the pathology, for the recognition and adequate management of the syndrome, according to the clinical picture presented.

REFERENCES

BERROCAL, T. *et al.* Congenital anomalies of the tracheobronchial tree, lung, and mediastinum: embryology, radiology, and pathology. **Radiographics**, v. 24, n. 1, p. e17-e17, 2004.

CHOWDHURY, U. K. *et al*. Surgical management of the scimitar syndrome. **Journal of Cardiac Surgery**, v. 36, n. 10, p. 3770-3795, 2021.

DUPUIS, Claude et al. "Infantile" form of the scimitar syndrome with pulmonary hypertension. **The American journal of cardiology**, v. 71, n. 15, p. 1326-1330, 1993.

DUPUIS, C. *et al.* The "adult" form of the scimitar syndrome. **The American journal of cardiology**, v. 70, n. 4, p. 502-507, 1992.

GUDJONSSON, U.L.F; BROWN, J. W. *et al*. Scimitar syndrome. In: **Seminars in thoracic and cardiovascular surgery: pediatric cardiac surgery annual**. WB Saunders, 2006. p. 56-62.

MULLIGAN, M. E. History of scimitar syndrome. **Radiology**, v 210, n. 1, p. 288-290, 1999.

VIDA, V. L. *et al.* Scimitar syndrome: a European congenital heart surgeons association (ECHSA) multicentric study. **Circulation**, v. 122, n. 12, p. 1159-1166, 2010.

YAMANARI, T.R.; CANTONI, S.T.O. Malformações pulmonares congênitas. In: ZATTAR, L.; VIANA, P.C.C.; CERRI, G.G. **Radiologia diagnostica prática: manual da residência do Hospital Sírio-Libanês.** 1. Ed. Barueri: Manole, p. 204, 2017.