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IMAGE IN CARDIOLOGY

More than a rare cause of pulmonary hypertension in the elderly

Mais do que uma causa rara de hipertensão arterial pulmonar no idoso

Rui Plácido[a,*], **Isabel Sousa**[b], **Dulce Antunes**[b], **Tatiana Guimarães**[a], **Fausto J. Pinto**[a]

[a] *Cardiology Department, Santa Maria University Hospital (CHLN), Lisbon Academic Medical Centre, and Centro Cardiovascular da Universidade de Lisboa, Faculdade de Medicina, Lisboa, Portugal*
[b] *Radiology Department, Santa Maria University Hospital (CHLN), Lisbon Academic Medical Centre, Lisboa, Portugal*

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An 80-year-old woman with a history of systemic hypertension and dual-chamber pacemaker implantation for sick sinus syndrome presented to the hospital with worsening dyspnea and fatigue for several months. The chest X-ray (Figure 1A) showed a markedly enlarged cardiac silhouette along with a tubular opacity (dotted lines) paralleling the right heart border. Echocardiographic examination depicted dilated right heart chambers, right ventricular dysfunction and a high probability of pulmonary hypertension.

Computed tomography angiography (Figure 1B-E) revealed an abnormal connection between a single right pulmonary vein and the inferior vena cava, a finding consistent with scimitar syndrome (Supplementary Video 1). There were no right pulmonary artery or lung hypoplasias. It also identified an anomalous origin of the left circumflex coronary artery from the right pulmonary artery (LCx/ALCAPA), close to the pulmonary bifurcation. The left anterior descending coronary artery arose from the left aortic sinus (Supplementary Videos 1 and 2).

Hemodynamic assessment confirmed the presence of pulmonary hypertension with a precapillary phenotype (mean pulmonary artery pressure: 46 mmHg; pulmonary capillary wedge pressure: 8 mmHg), and significantly increased pulmonary vascular resistances (8 Wood units), which precluded surgical correction. The patient's symptoms improved after decongestive therapy and dual pulmonary vasodilator therapy, which included a phosphodiesterase-5 inhibitor (sildenafil) and an endothelin receptor antagonist (bosentan).

Scimitar syndrome is a rare congenital anomaly of pulmonary venous return, in which the right pulmonary vein connects anomalously to the inferior vena cava. Coronary artery anomalies are rarely associated, with anomalous left

* Corresponding author.
E-mail address: placidorui@gmail.com (R. Plácido).

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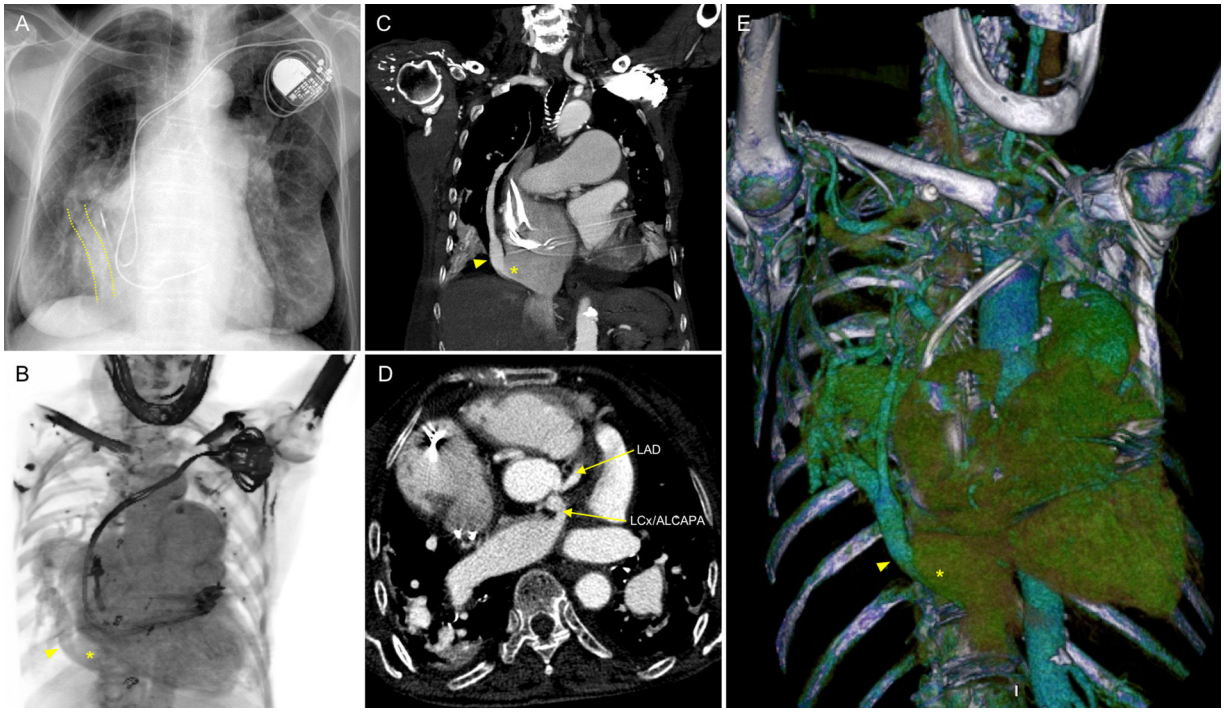

Figure 1 (A) Chest X-ray showing a markedly enlarged cardiac silhouette along with a right-sided tubular opacity (dotted lines) paralleling the heart border; (B and C) computed tomography angiography (CTA), coronal plane, depicting a single right pulmonary vein (arrowhead) connected to the inferior vena cava (*); (D) CTA, axial plane, revealing the presence of an anomalous origin of the left circumflex coronary artery from the right pulmonary artery (arrow), close to the pulmonary bifurcation; (E) CTA with three-dimensional volume rendering. LAD: left anterior descending coronary artery; LCx/ALCAPA: anomalous origin of the left circumflex coronary artery from the right pulmonary artery.

circumflex artery from the pulmonary artery only anecdotally reported in pediatric patients.

Conflicts of interest

The authors have no conflicts of interest to declare.

Appendix A. Supplementary material

Supplementary material associated with this article can be found in the online version at doi:10.1016/j.repc.2019.06.007.