Marfan Syndrome with Bilateral Subclavian Artery Aneurysm Associated with Coarctation of the Aorta

Síndrome de Marfan con aneurisma subclavio bilateral asociado a coartación aórtica

NIDHESSY PAGÉS MORALES[1], ELIZABETH RAMÍREZ REYES[1], BRISNEY MENGANA CHIVAS[1]

Marfan syndrome is an inherited disease described in children and adults that affects the connective tissue of multiple organs and systems, such as the cardiovascular and musculoskeletal systems, eyes and skin. (1) It involves autosomal dominant inheritance due to a genetic alteration in chromosome 15; prevalence is estimated at 1 in 5000-10 000 newborns, and it affects both sexes equally. (2) Coarctation of the aorta is a highly complex and life-threatening cardiovascular malformation characterized by narrowing of the descending aorta, which in some cases may be a long hypoplastic aortic segment. It is usually located at the ductus arteriosus insertion point, distal to the left subclavian artery. It tends to be associated with anomalies in the aortic arch and results in left ventricular pressure overload. (3)

We present the case of a 51-year-old female patient with a history of arterial hypertension, ischemic heart disease and Marfan syndrome, who presents with increased soft tissue volume in the left anterior thorax. During anamnesis, she denies any pain or any other symptoms.

A computed tomography (CT) of the chest is performed with contrast agent using CT angiography technique, with axial slices, and both sagittal and coronal reconstructions. Aneurysmal dilation of both subclavian arteries is observed in the supraclavicular region (Figures 1 and 2). The left subclavian artery is more prominent, with an 81-mm transverse diameter and a 95-mm longitudinal diameter, and a thrombus on the wall leaving a 41-mm lumen. The contralateral artery has a 57-mm longitudinal diameter and a 36-mm transverse diameter, with absence of thrombi. The ascending aorta is dilated (53-mm), showing coarctation after the left subclavian artery emergency (Figure 2).

In Marfan syndrome, dilated aortic root is the most common cardiovascular occurrence, estimated to affect 60-80% of patients and to cause 90% of deaths. (4)

Aneurysms of the subclavian artery are extremely rare, with an incidence ranging from 0.01% to 3.5% as reported by different authors. It results from infections or degeneration of the *tunica media* of the artery, but it may be part of a clinical spectrum of diseases, including Marfan syndrome. (5) Other causes of subclavian artery aneurysm are thoracic outlet syndrome and trauma secondary to gunshot wound, clavicle fracture or iatrogenic causes. (5,6)

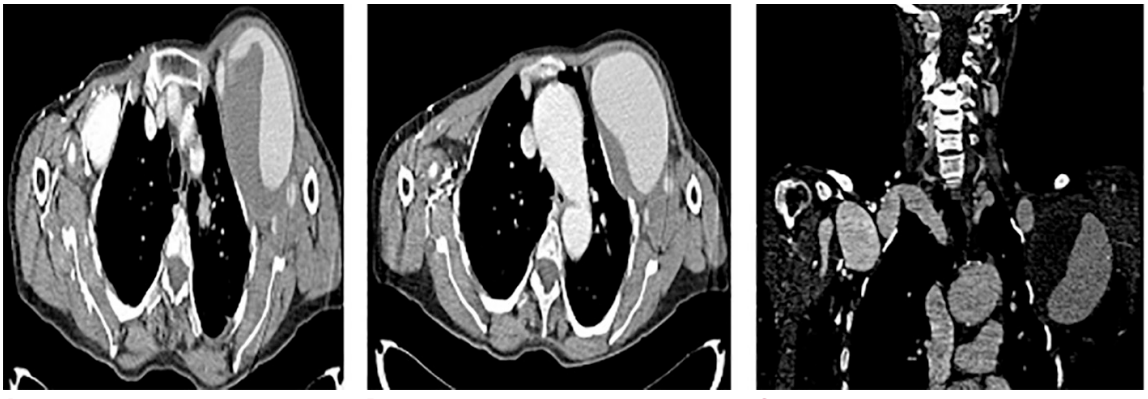
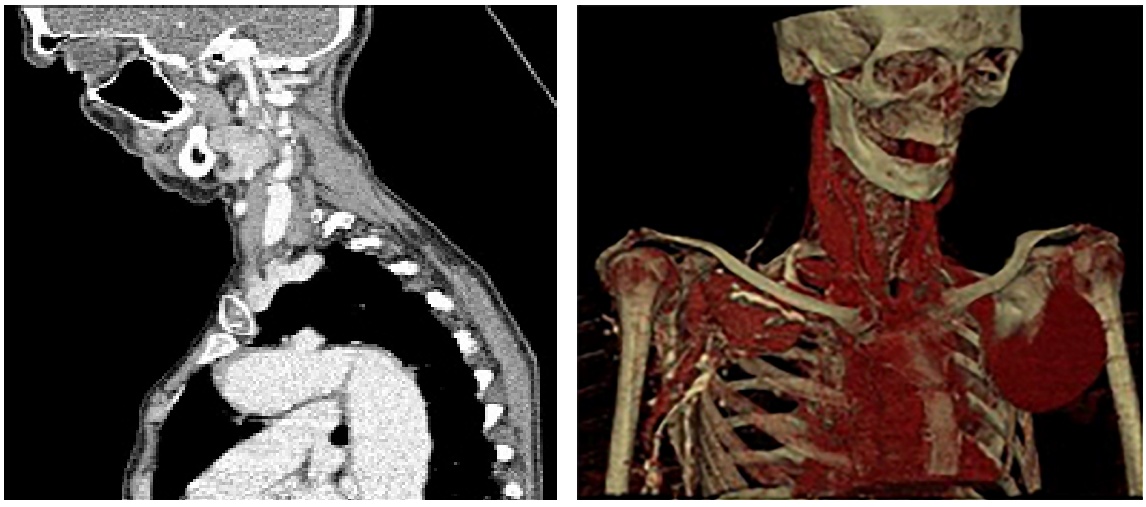


Fig. 1. Computed tomography. A. Axial slice showing both vascular dilations at the subclavian arteries level. B. Axial slice showing aneurysmal dilation of the left subclavian artery, with both dilation and coarctation of the aorta. C. Coronal slice showing aneurysmal dilation of both subclavian arteries, the left one with a thrombus on the wall.

Rev Argent Cardiol 2024;92:291-292. http://dx.doi.org/10.7775/rac.v92.i4.20806

Correspondence: Nidhessy Pagés Morales. E-mail: nidhepages@gmail.com

[1] Imaging Service of the University Hospital "Manuel Ascunce Domenech" Camagüey, Cuba



A B

Fig. 2. Computed tomography. A. Sagittal section showing dilation and coarctation of the aorta. B. Volumetric reconstruction showing both aneurysmal dilations of the subclavian arteries

Complications depend on the location, and the most commonly reported are brachial plexus injury or compressed upper extremity vessels possibly leading to edema. Surgical procedures have been reported for treatment, with associated complications in almost one third of cases. (6)

Ethical considerations

Not applicable.

Conflicts of interest

None declared (See authors' conflicts of interest forms on the website).

Financing

None.

REFERENCES

1. Serrano RG, Marcano Sanz LE, Bacallao Carril D. Cardiovascular considerations of Marfan syndrome in pediatric ages. Rev Cubana Pediatr [Internet]. 2012;84):176-87.
2. Romero-Asín A, Vidal-Casal JJ, Asín-Rodríguez D. Marfan syndrome, a genetic disease with cardiovascular complications. Rev Med Electron. [Internet]. 2022;44:425-33.
3. Lizano MJ, Álvarez Urbina J. Congenital aortic coarctation: an update in the diagnostic and therapeutic approach. Revista Ciencia Y Salud Integrando Conocimientos. 7(2). https://doi.org/10.34192/cienciaysalud.v7i2.617
4. Michel-Martinez AJ, Chavez-Camacho RJ, Laguna-Chavez RA. Cardiovascular involvement in marfan syndrome: a case report: Rev Cient Cienc Med 2020;23:252-7. https://doi.org/10.51581/rccm.v23i2.18.
5. Trujillo-Santos AJ, Jurado Gámez B. Left subclavian artery aneurysm: an infrequent cause of superior lung mass. Ann Med Inter (Madrid) [Internet]. 2001;18:40-1. https://doi.org/10.4321/S0212-71992001000200007
6. Peña-Juárez RA, Medina-Andrade MA, Ramírez-Cedillo D, Landeros A, Abarca-De la Paz JP. Congenital subclavian aneurysm in a pediatric patient. Arch Cardiol Mex 2021;91:514-5. https://doi.org/10.24875/ACM.20000543.