Kommerell Diverticulum, Right Aortic Arch and Anomalous Left Subclavian Artery: A Case Report

Viviane Tiemi Hotta[1,2], Andrei Skromov Albuquerque[1,3], Valdir Ambrósio Moisés[1,4]

Fleury Medicina e Saúde[1]; Instituto do Coração HC - FMUSP[2]; Hospital Alemão Oswaldo Cruz[3]; Universidade Federal de São Paulo[4], São Paulo, SP - Brazil

Introduction

The congenital anomalies of the aortic arch present an assorted clinical spectrum and may be associated with cardiac abnormalities and the anomalous origin of great vessels. The right-sided aortic arch is a rare anomaly, with an incidence from 0.05% to 0.10% reported in series of cases. It may appear as an isolated anomaly or associated with congenital heart defects and symptoms of tracheoesophageal compression[1-3]. In these cases, the noninvasive imaging methods are applied for identification and diagnosis of these lesions, anatomic definition and investigation of possible associated heart defects[4-6]. In patients without symptoms or signs of congenital heart defect, the diagnosis may be casually suspected during a chest X-ray or echocardiography performed due to other reasons.

Case Report

The case reported in this manuscript illustrates a congenital anomaly characterized by a right-sided aortic arch, associated with the anomalous origin of the left subclavian artery in the descending thoracic aorta. The twelve-year old patient attended the echocardiography service owing to complaint of palpitations on exertion. She denied chest pain, dyspnea, dizziness or syncope. The physical examination did not reveal murmurs or other abnormalities.

The transthoracic echocardiography revealed a medial displacement of the aortic arch and to the right, and a vessel with ascending artery flow on color flow mapping without a significant gradient. The vessel presented a path parallel to the ascending aorta, but a definition was not possible regarding the possible connection with the initial descending thoracic aorta, or if there was only an ascending upper path (Figure 1).

Owing to the suspected vessel anomaly associated with the right-sided aortic arch, the patient underwent Magnetic Resonance Angiography (MRA) of the thoracic aorta as diagnostic complementation. MRA revealed a right-sided aortic arch, with discrete impression regarding the right lateral position of the trachea, determining a discrete displacement to the left, in addition to an Aberrant Left Subclavian Artery (ALSA) and retroesophageal path, presenting normal caliber and opacification. In the ALSA origin, a focal dilatation (Kommerell diverticulum) was detected, and it may possibly be associated with the anomalous origin of this vessel (Figure 2).

Discussion

The Kommerell diverticulum is a rare congenital anomaly which is typically associated with the right-sided aortic arch, aberrant left subclavian artery, and is usually diagnosed in an incidental manner in asymptomatic patients. In this case, as the patient complained of palpitations, a symptom not connected to these findings, some aspects triggered the diagnostic suspicion of anomalous origin of the ALSA, insomuch as the absence of bifurcation of the brachiocephalic trunk and observation of an ascending artery emerging from the descending thoracic aorta[1-6]. In the case reported, since signs of tracheal or esophageal compression on MRA or congenital heart defects on transthoracic echocardiography were not found, the symptoms stated by the patient are most probably not correlated with the radiological findings.

Although these changes often did not bring on symptoms, it is important to be heedful of the development of atherosclerotic disease in the anomalous vessels, as well as dissection or aneurysmal dilatation with compression of adjacent mediastinal (esophagus and trachea) causing dysphagia, dyspnea or stridor, in addition to cough and chest discomfort[7]. Other important aspect is the association with the presence of right-sided aortic arch with the DiGeorge Syndrome (or velo-cardio-facial syndrome)[8], which may be present mainly when there are associated defects, such as cono-trunk malformations, atrial or ventricular septal defects and tetralogy of Fallot. The diagnosis of this syndrome is important for counseling purposes in possible pregnancies and rate of recurrence in families.

Besides infrequent, the aortic arch anomalies associated with vessel abnormalities may be safely diagnosed with noninvasive imaging methods, showing a significant correlation among the findings of different methods. The right-sided aortic arch may be suspected in the longitudinal suprasternal section due to the need for anti-clockwise rotation of the transducer to fully view the arch and the descending aorta. In cross sections, if the aortic arch is right sided, the brachiocephalic trunk is on the left and bifurcates[9,10]. In the case described, the Transthoracic Echocardiography (TTE) was important for screening and clinical suspicion of vessel anomaly, leading to the performance of thoracic MRA, which allowed the anatomical diagnosis at a higher accuracy. Additionally, the imaging methods are important in the clinical follow-up and detection of complications associated with these anomalies,

Keywords

Heart defects congenital; Aorta thoracic/abnormalities; Diagnostic imaging; Echocardiography.

Mailing address: Viviane Tiemi Hotta •
Avenida Dr. Enéas Carvalho de Aguiar, 44, 05403-000, São Paulo, SP - Brazil
Email: viviane.hotta@grupofleury.com.br
Manuscript received on June 11, 2014; revised on June 14, 2014; accepted on July 21, 2014.

DOI: 10.5935/2318-8219.20140037

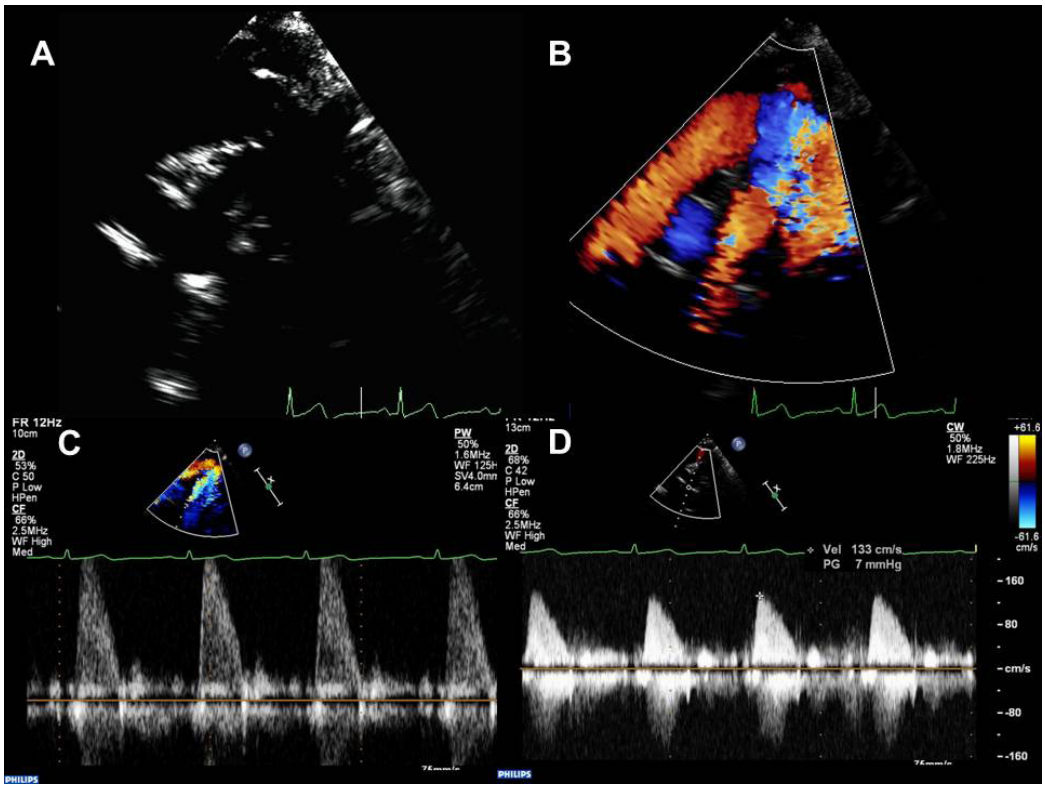

Figure 1 – *Two-dimensional echocardiographic and transthoracic Doppler echocardiography images obtained from a longitudinal suprasternal section. A: 2D echocardiography showing the medial displacement of the aortic arch and to the right; B: Vessel with artery flow observed on color flow mapping parallel to the ascending aorta; C: Pulsatile Doppler study of the anomalous left subclavian artery; D: The continuous Doppler study of the same vessel did not show a pressure gradient.*

such as compression symptoms, resulting in dyspnea, dysphagia and vascular complications (such as dissection and atherosclerotic disease) connected to the anomalous vessel.

Authors' Contribution

Investigation conception and design: Hotta VT; Data collection: Hotta VT, Albuquerque AS, Moisés VA; Data analysis and interpretation: Hotta VT, Albuquerque AS, Moisés VA; Manuscript drafting: Hotta VT; Critical review of the manuscript regarding the important intellectual content: Hotta VT, Albuquerque AS, Moisés VA.

Potential Conflicts of Interest

No relevant potential conflicts of interest.

Sources of Funding

This study had no external funding sources.

Academic Association

This study is not associated with any graduate programs.

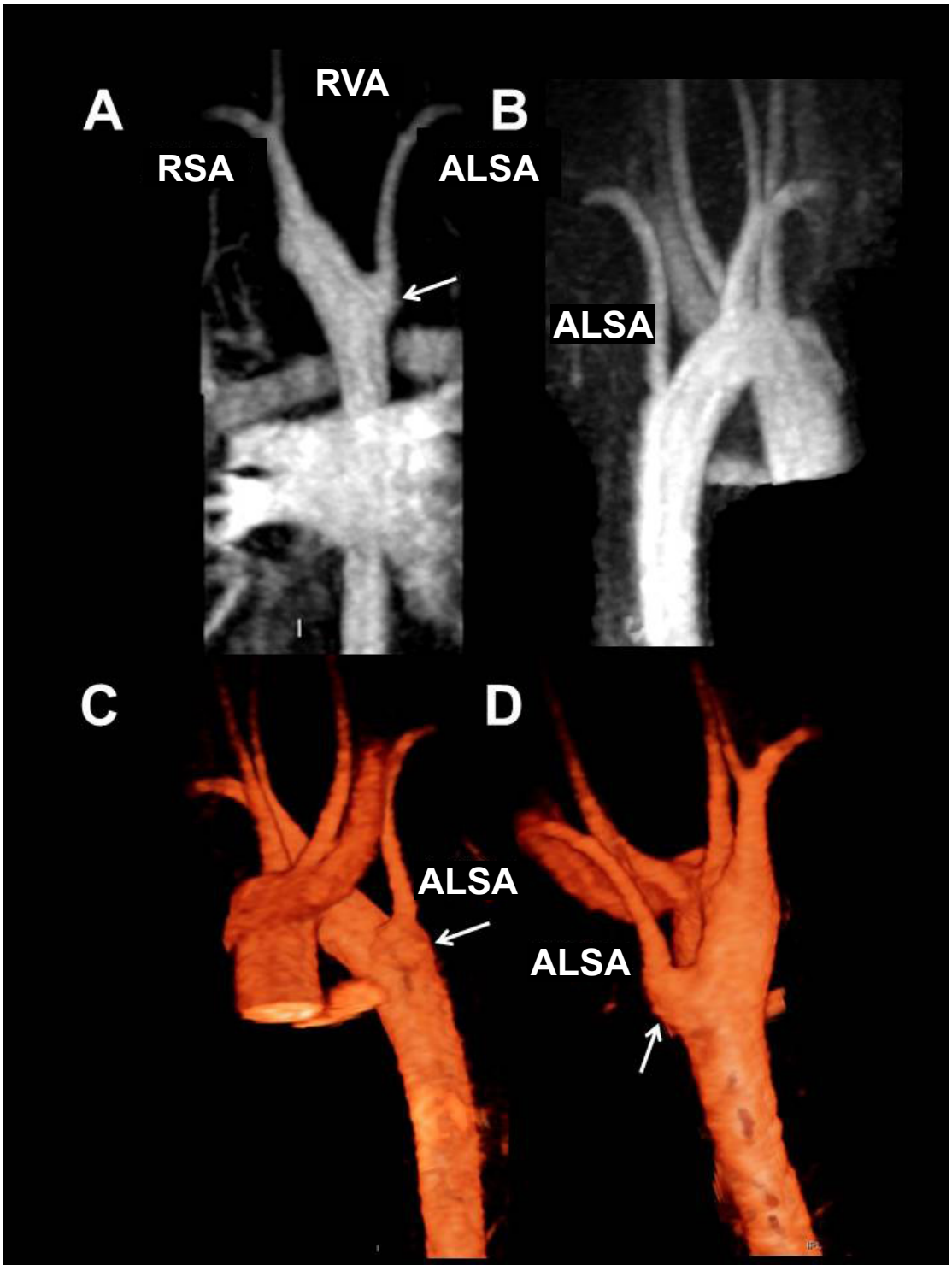

Figure 2 – *A: 3D reconstruction in maximum intensity projection (MIP) of the thoracic aorta in anterior view, excluding the ascending aorta and the proximal half of the arch. Right-sided aortic arch and medianized descending aorta. Right Subclavian Artery (RSA) with origin in common topography. Left Subclavian Artery (LSA) with anomalous origin (aberrant) in the focal dilatation located in the left lateral position, at the aortic isthmus, characterizing a Kommerell diverticulum (arrows); B: 3D reconstruction in maximum intensity projection (MIP) of the thoracic aorta in right posterolateral view. ALSA with anomalous origin (aberrant) at the aortic isthmus, distally for the origin of the other arch branches; C: 3D reconstruction in Volume Rendering (VR) of the aortic arch in a left anterolateral view. The following arteries are originated: left common carotid artery, right common carotid artery, right subclavian and left subclavian artery, from proximal to distal; D: 3D reconstruction in Volume Rendering (VR) of the aortic arch in a posterior view. Focal dilatation highlighted (Kommerell diverticulum, arrows) in the origin of aberrant ALSA in C and D images. RVA: right vertebral artery.*

References

1. Davies M, Guest PJ. Developmental abnormalities of the great vessels of the thorax and their embryological basis. Br J Radiol. 2003; 76(907): 491-502.
2. Shuford WH, Sybers RG, Edwards FK. The three types of right aortic arch. Am J Roentgenol Radium Ther Nucl Med. 1970;109(1):67-74.
3. Demirkol S, Balta S, Bozlar U, Sari S, Unlu M, Arslan Z. Kommerell diverticulum associated with aberrant left subclavian artery and right-sided aortic arch. Eur Heart J Cardiovasc Imaging. 2013;14(8):764.
4. Donnelly LF1, Fleck RJ, Pacharn P, Ziegler MA, Fricke BL, Cotton RT. Aberrant subclavian arteries: cross-sectional imaging findings in infants and children referred for evaluation of extrinsic airway compression. AJR. Am J Roentgenol. 2002;178(5):1269-74.
5. Barranhas AD, Indiani JM, Marchiori E, Santos AA, Rochitte CE, Nacif MS. Atypical presentation of Kommerell's diverticulum. Arq Bras Cardiol. 2009;93(6):e88-90, e101-3.
6. Lee SJ, Lee SH, Kim JH, Lee H, Lee DJ, Kim JH, et al. Son JW, Son JW, Hong GR. A Case of Kommerell's Diverticulum Initially Detected by Transesophageal Echocardiography. J Cardiovasc Ultrasound. 2013;21(1):30-2.
7. Adams MT, Wilhelm M, Singh N, Andersen C. Right aeortic arch with Kommerell Diverticulum: a rare cause of dysphagia. JAMA Otolaryngol Head Neck Surg. 2014. Apr 17 [Epub ahead of print]
8. Snider AR, Serwer GA, Ritter SB, Gersony RA. Echocardiography in pediatric heart disease. 2nd ed. Saint Louis:Mosby; 1997.p.452-96.
9. Celano V, Pieroni DR, Gingell RL, Roland J-MA. Two-dimensional recognition of the right aortic arch. Am J Cardiol. 1983; 51(9):1507-12.
10. Monaco M, Lillo S, La Marca Giordano A, Contaldo A, Schiavone V. Endovascular repair of a right-sided thoracic aortic aneurysm with Kommerell Diverticulum and aberrant left subclavian artery. Ann Vasc Surg. 2014;28(5):1323.e1-5.