Right aortic arch with aberrant left subclavian artery revealed by a dyspnea: A case report

Lahfidi Amal, Boukhalit Hind, Echikhi Meriam, Jroundi Laila, Laamrani Fatimazahrae

ABSTRACT

Introduction: Congenital cardiovascular diseases affect about 1% of live births. In 15–20% of cases, this is an anomaly of the aortic arch that includes various malformations with a common embryological origin and which may be asymptomatic or manifest by signs of compression of the tracheoesophageal axis. The diagnosis is sometimes accidental during a radiographic examination, especially in adults. Case Report: We report a clinical case of asymptomatic right aortic arch, discovered during dyspnea. Conclusion: Right-sided aortic arch is a rare anatomical variant present in 0.1% of adult population. It is associated with an aberrant left subclavian artery and half of the cases causing dyspnea or dysphagia. Specific imaging feature can lead to diagnosis and help choose an appropriate treatment.

Keywords: Chest angio-CT, Dyspnea, Right aortic arch

INTRODUCTION

The anomalies of the aortic arches have been known since 18th century. Their anatomical description was initially performed in 1737 by Hommel, then their radiological aspects in 1926 by Arkin, and those of the right subclavian retro-oesophageal in 1936 by Kommerell, the clinical description of double Bow by Wolman in 1939 [1].

The right aortic arch (RAA) is one of many types of congenital aortic arch abnormalities. It is present in 0.05–0.1% of adult cases [1, 2]. Several classifications of these anomalies have been proposed according to the arrangement of the vessels by supply to the esophagus or the presence of congenital cardiac anomalies [1, 2].

The RAA is often asymptomatic in adults, discovered incidentally during the exploration of another associated cardiac malformation, or during a morphological evaluation of another pathology. When it is symptomatic, dysphagia and dyspnea are the most frequently reported [1, 2].

The Edwards classification describes three types of RAA [3, 4]:

Type I: RAA with mirror image branching
Type II: RAA with aberrant left subclavian artery
Type III: RAA with isolation of the left subclavian artery

CASE REPORT

We report a case of a 34-year-old female patient with no pathological history who had difficulties in swallowing for several years. However, the fibroscopy results were normal. At the time of this study, she was consulting for chest pain and dyspnea. Currently she is consulting for chest pain and dyspnea.
The chest radiograph (Figure 1) showed an enlargement of the superior mediastinum with absence of the aortic knob normally visible on the left and the para-aortic line.

Chest angio-computed tomography (CT) (Figure 2) showed an RAA, the butt of the aorta carries a retrotracheo-oesophageal path compressing the esophagus, the ascending and descending thoracic aorta located on the right. It is an RAA with successive births of: the left common carotid artery LCC (4), the ascending aorta which travels in pre-tracheal in its path to the left side, then the right common carotid RCC (5), and the right subclavian artery SCD (6). Thus on the left edge of the posterior part of the aortic arch appears the Kommerell’s diverticulum giving rise to the aberrant retrooesophageal left subclavian artery (LSC).

**DISCUSSION**

The aortic artery abnormalities are rare, diagnosed most often during childhood, and the most common is the RAA. They result from an abnormal embryological development of primitive aorta (abnormal of the fourth primitive branchial arch) characterized by the persistence of the right aortic root and the regression of its left counterpart. In this case the aortic arch and the descending thoracic aorta found on the right side of the spine and trachea unlike the normal subject. These abnormalities may be associated with congenital heart disease mainly tetralogy of Fallot, atrial septal defect, ventricular septal defect, or coarctation of aorta [2].

According to Edwards’ double-aortic arch hypothesis, there are bilateral aortic arches and arterial canals, common carotid arteries, and subclavian arteries emerging from their respective arches. The fourth aortic arch cutting sites define the anatomy of the aortic arch. The numbers (Figure 3) represent the sites of interruptions of the arc at different levels. The normal site is located in 1; the other four sites correspond to the anomalies of the fourth aortic arch [2–5]:

Figure 1: Chest X-ray shows: enlargement of the superior mediastinum (arrow), absence of left aortic knob and the para-aortic line.

Figure 2: Thoracic CT scan in axial: a right aortic arch (1) with successive births of the vessels of the bartering of the supra-aortic, a posterior diverticulum (diverticulum of Kommerell) (2) giving birth to aberrant retrooesophageal left subclavian artery (3), aorta ascending (7) and descending (8) to the right, (9) trachea.

Figure 3: Cutting sites of fourth aortic arches explaining the different anomalies of the aortic arches [6]. ACG/ACD: left/right carotid, ASG/ASD: left/right subclavian, AA: ascending aorta, AD: descending aorta, AP: pulmonary artery.
• Cleavage at site 2, between the right primitive carotid forward and the right subclavian artery behind, is responsible for a right aortic arch with successive births of right carotid artery, left carotid artery, left subclavian artery, and retrooesophageal right subclavian artery [5].
• Cleavage at site 3, between the ascending aorta forward and the left carotid artery behind, is responsible for a right aortic arch with successive births of the right carotid artery, the right subclavian artery, and an aberrant left brachiocephalic artery trunk [5].
• Cleavage at site 4, between the left primary carotid anterior and the left subclavian artery behind, is responsible for a right aortic arch with successive births of the left carotid, right carotid, the right subclavian artery, and the aberrant left subclavian artery (retrooesophageal) [5].
• Cleavage at site 5, between the left subclavian artery forward and the descending aorta behind is responsible for a right aortic arch with successive births of a left brachiocephalic artery trunk, the right carotid artery, and the right subclavian artery [5].

The birth defects of the subclavian arteries give either a right retro-oesophageal subclavian artery on aorta on the left, or a left subclavian artery on butt on the right. In the case where the aortic arch is on the right, the aberrant retro-oesophageal left subclavian artery may present at its origin with a bulge which is Kommerell’s diverticulum (as the case of our patient) with sometimes a compressive ligament left arterial compressive [7, 8]. This diverticulum is a remnant of the fourth right aortic arch. It is manifested by the compression of neighboring organs (trachea and esophagus) [9].

The RAA with aberrant left subclavian artery may be asymptomatic or may cause chronic respiratory digestive symptoms [10].

The diagnosis is mainly radiological. It includes [10]:
• The chest X-ray shows indirect signs: the aortic knob and the descending thoracic aorta are seen on the right; tracheal axis is medial or deviated to the left.
• The barium enema: allows you to look for abnormal fingerprints on the esophagus. It can affirm the diagnosis of abnormality of the aortic arches and sometimes specifies the type of anomaly. It is currently supplanted by the magnetic resonance imaging (MRI).
• Thoracic CT scan: Chest angio-CT is the preferred examination to (100% sensitivity and 98% specificity) a complete morphological evaluation of aortic artery anomalies and associated congenital heart disease. It confirms the diagnosis and specifies the position of the aortic arch and supra-aortic trunks in relation to the esophagus and trachea.
• Magnetic resonance imaging allows both anatomical and functional exploration of all portions of the aorta. From the technical point of view, the basic sequences produced are the sequences in T1 spin echo, three-dimensional gradient echo sequences with gadolinium injection and cine-MRI sequences.
• Bronchoscopy or upper gastrointestinal tract (UGI) endoscopy: asserts or denies the vascular character of tracheal and esophageal compression by the pulsatile character (Kommerell’s diverticulum) and specifies the seat of compression. It can objectify an associated tracheomalacia.

The treatment of the RAA depends on the clinical situation. In asymptomatic cases radiologic monitoring is desirable, however, in patients suffering from compressive symptomatology surgical intervention is indicated. In case of association of asymptomatic right aortic arch with another pathology, the therapeutic modalities as well as the prognostic and evolutionary factors belong to the associated injury. Surgical treatment was required in our case [11].

CONCLUSION

Right-sided aortic arch is a rare anatomical variant present in 0.1% of adult population. It is associated with an aberrant left subclavian artery, and half of the cases causing dyspnea or dysphagia. Specific imaging feature can lead to diagnosis and help choose an appropriate treatment.

REFERENCES

1. Brevière GM. Anomalies des arcs aortiques. Encycl Méd Chir (Editions Scientifiques et Médecales Elsevier SAS, Paris), Cardiologie 2003;11-940-D-80:9p.
2. Hsu KC, Tsung-Che Hsieh C, Chen M, Tsai HD. Right aortic arch with aberrant left subclavian artery—prenatal diagnosis and evaluation of postnatal outcomes: Report of three cases. Taiwan J Obstet Gynecol 2011;50(3):353–8.
3. Cinà CS, Althani H, Pasenau J, Abouzahr L, Kommerell’s diverticulum and right-sided aortic arch: A cohort study and review of the literature. J Vasc Surg 2004;39(1):131–9.
4. Lee CH, Son JW, Park JS. Comprehensive three-dimensional analysis of right-sided aortic arch with multiple vascular anomalies. BMC Cardiovasc Disord 2004;14:104.
5. Arazinska A, Polgaj M, Szymczyk K, Kaczmarska M, Trębiński L, Stęfańczyk L. Right aortic arch analysis – Anatomical variant or serious vascular defect? BMC Cardiovasc Disord 2017;17(1):102.
6. Hanneman K, Newman B, Chan F. Congenital variants and anomalies of the aortic arch. Radiographics 2017;37(1):32–51.

International Journal of Case Reports and Images, Vol. 11, 2020. ISSN: 0976-3198
7. Celik M, Celik T, Iyisoy A, Guler A. An unusual combination of congenital anomalies in an adult patient: Patent ductus arteriosus, Kommerell’s diverticulum with aberrant right subclavian artery, and heterotaxy syndrome. Hellenic J Cardiol 2011;52(5):469–72.

8. Ruzmetov M, Vijay P, Rodefeld MD, Turrentine MW, Brown JW. Follow-up of surgical correction of aortic arch anomalies causing tracheoesophageal compression: A 38-year single institution experience. J Pediatr Surg 2009;44(7):1328–32.

9. Le Bret E, Papadatos S, Folliguet T, Laborde F. Anomalies des arcs aortique. EMC (Elsevier, Paris), Techniques chirurgicales-Thorax 2002;42-767:12p.

10. Corone P, Vernant P. Anomalies des arcs aortiques. Encycl Med Chir, Cardiologie (Elsevier SAS Paris) 1970:11-040-M-M-50:1–8.

11. Atkin GK, Grieve PP, Vattipally VR, Ravikumar KH, Das SK. The surgical management of aortic root vessel anomalies presenting in adults. Ann Vasc Surg 2007;21(4):525–34.

*********

Author Contributions
Lahfidi Amal – Conception of the work, Design of the work, Acquisition of data, Analysis of data, Interpretation of data, Drafting the work, Revising the work critically for important intellectual content, Final approval of the version to be published, Agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved

Boukhalit Hind – Conception of the work, Design of the work, Acquisition of data, Analysis of data, Interpretation of data, Drafting the work, Revising the work critically for important intellectual content, Final approval of the version to be published, Agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved

Echikhi Meriam – Conception of the work, Design of the work, Acquisition of data, Analysis of data, Interpretation of data, Drafting the work, Revising the work critically for important intellectual content, Final approval of the version to be published, Agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved

Jroundi Laila – Conception of the work, Design of the work, Acquisition of data, Analysis of data, Interpretation of data, Drafting the work, Revising the work critically for important intellectual content, Final approval of the version to be published, Agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved

Laamrani Fatimazahrae – Conception of the work, Design of the work, Acquisition of data, Analysis of data, Interpretation of data, Drafting the work, Revising the work critically for important intellectual content, Final approval of the version to be published, Agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved

Guarantor of Submission
The corresponding author is the guarantor of submission.

Source of Support
None.

Consent Statement
Written informed consent was obtained from the patient for publication of this article.

Conflict of Interest
Authors declare no conflict of interest.

Data Availability
All relevant data are within the paper and its Supporting Information files.

Copyright
© 2020 Lahfidi Amal et al. This article is distributed under the terms of Creative Commons Attribution License which permits unrestricted use, distribution and reproduction in any medium provided the original author(s) and original publisher are properly credited. Please see the copyright policy on the journal website for more information.
