A rare association of avian aortic arch with a mid-arch narrowing of the aorta

Hend Keskes[a], Emna Bennour[a], Ahmed Sghaier[a], Ikram Kammoun[a], Meriem Affes[b], Sonia Marrakchi[a], Salem Kachboura[a]

[a] *Cardiology Department, Abderrahman Mami's hospital – Ariana, Tunisia*
[b] *Radiology Department, Abderrahman Mami's hospital – Ariana, Tunisia*

ARTICLE INFO

Article history:
Submitted: 24. 4. 2021
Revised: 17.5. 2021
Accepted: 19. 5. 2021
Available online: 15. 6. 2022

Klíčová slova:
Anomálie aortálního oblouku
Aortální oblouk u ptáků
Koarktace
Kazuistika

SOUHRN

Kontext: Anatomická rozmanitost aortálního oblouku a jeho větví umožňuje velkou řadu kombinací. Se zúžením středního segmentu aortálního oblouku se lze setkat vzácně, pouze v 2,5–3,5 % všech případů koarktace aorty. Aortální oblouk u ptáků má dvě větve se společným odstupem bilaterálních společných karotid a společným odstupem bilaterálních podklíčkových tepen. Zdá se, že u lidí se vždy jedná o výjimečnou anomálii, a i když je každá z nich samostatně popsána, obě současně se vyskytují velmi vzácně.
Popis případu: Popisujeme případ 50letého pacienta, který se dostavil na naši kliniku se závratěmi a námahovou únavou horních končetin. V jeho anamnéze nebyla přítomna hypertenze ani diabetes mellitus a klinické vyšetření prokázalo nízký krevní tlak, žádný srdeční šelest, výraznou pulsaci karotid s šelestem nad oběma karotidami a hmotu v laterocervikální oblasti vlevo. Puls na radiálních tepnách a na femorálních tepnách nebylo možno vůbec nahmatat. Elektrokardiografické vyšetření prokázalo hypertrofii levé síně a komory. Výsledek rentgenového vyšetření srdce a plic byl normální. Dvojrozměrná echokardiografie prokázala mírnou dilataci levé síně, komorovou hypertrofii s hypokinezí septa. Dopplerovský ultrazvuk supraaortálních kmenů prokázal bilaterální subklaviální steal syndrom a CT angiografie aorty, supraaortálních kmenů a mozkových cév zobrazila aortální oblouk se dvěma větvemi s těžkým těsným zúžením mezi nimi; vnější karotické tepny byly zvětšené a v pravé karotické tepně bylo nalezeno retrofaryngeální aneurysma. Chirurgické řešení koarktace proběhlo úspěšně. Pooperační zotavování bylo bez problémů. Při klinickém vyšetření nebyla přítomna hypertenze, puls na femorálních tepnách a tepnách horních končetin byl přítomen a symetrický. Kontrolní rentgenové vyšetření po pěti letech prokázalo propustnou dakronovou protézu a regresi aneurysmatu.
Závěr: Anomálie aortálního oblouku již přitáhly pozornost několika autorů, zvláště díky pokroku v oblasti endovaskulárních intervencí a složitých chirurgických výkonů. Povědomí o těchto anomáliích je nesmírně důležité před jakýmikoli intervenčními a chirurgickými výkony v oblasti hrudníku a krku.

ABSTRACT

Background: Variations of the aortic arch and its branches appear in a large number of possible combinations. Mid-aortic arch narrowing is rare, and represents 2.5–3.5% of all aortic coarctations. Avian arch is a two-branch arch with common origin of bilateral common carotids and common origin of bilateral subclavian arteries. It seems to be exceptional in humans. Although these two anomalies are well identified in isolation, their association seems very rare.
Case presentation: We report a case of a 50-year-old patient presenting to our department complaining of dizziness and on-exertion fatigue of the upper limbs. Medical history revealed no hypertension and no diabetes mellitus and clinical examination showed a low blood pressure, no heart murmur, marked pulsation of the carotids with a bilateral carotid murmur and a left latero-cervical mass. Radial pulses were weak and femoral pulses were absent. Electrocardiogram showed left atrial and ventricular hypertrophy. Chest X-ray was normal. 2-dimensional echocardiography showed moderately left atrial dilatation, ventricular hypertrophy with septal hypokinesia. Doppler ultrasound of supra-aortic trunks showed bilateral subclavian steal phenomenon and computed tomography angiography of the aorta, supra-aortic trunks, and brain vessels showed a two-branch aortic arch with a severe in-between narrowing, external carotid arteries were enlarged and a retropharyngeal aneurysm was present in the right carotid artery. Surgical treatment of the coarctation was successfully done. Postoperative recovery was good. At clinical examination, there was no hypertension, femoral and upper limbs pulses were present and symmetric. A radiological control 5 years later showed a permeable Dacron tube and a regression of the aneurysm.
Conclusion: Aortic arch anomalies have called the attention of several authors especially with advancements of endovascular interventions and complex surgical procedures. Their knowledge is of crucial importance before any interventional and surgical procedures in thorax and neck

Keywords:
Avian arch
Aortic arch anomaly
Case report
Coarctation

Address: Ahmed Sghaier, MD, Cardiology Department, Abderrahman Mami's hospital – Ariana, Tunisia, e-mail: sghahmed991@gmail.com
DOI: 10.33678/cor.2021.061

Please cite this article as: Keskes H, Bennour E, Sghaier A, et al. A rare association of avian aortic arch with a mid-arch narrowing of the aorta. Cor Vasa 2022;64:320–323.

Background

Commonly, the aortic arch is left-sided and has three branches. Variations of the aortic arch and its branches appear in a large number of possible combinations with various frequencies.

Mid-aortic arch coarctation is rare,[1] and represents 2.5–3.5% of all aortic coarctations.[2] Anomalies of the arch vessels associated with coarctation are unusual.[3]

Avian arch is a two-branch arch with common origin of bilateral common carotids and common origin of bilateral subclavian arteries.[4] It seems to be exceptional in humans with a prevalence rate of 0.16 to 1%.[5]

Although these two anomalies are well identified in isolation, their association seems very rare.

We present a case describing an unusual aortic branching pattern with an associating avian arch (common bi-carotid trunk and common bi-subclavian trunk) to mid-arch narrowing of the aorta between the two common trunks.

Case presentation

A 50-year-old patient presented to our Cardiology Department with complaints of dizziness and on-exertion fatigue of the upper limbs evolving for 6 months with a recent progressive aggravation without chest pain, nor syncope.

Medical history revealed no hypertension and no diabetes mellitus.

Clinical examination showed a low blood pressure (90/40 mmHg) symmetrical in both arms, no heart murmur, but marked pulsation of the carotids with a bilateral carotid murmur and a left latero-cervical mass. Radial pulses were weak and femoral pulses were absent on both sides.

Electrocardiogram showed a sinus rhythm and a left atrial and ventricular hypertrophy. At chest X-ray there was no cardiomegaly and no notching of the ribs.

2-dimensional echocardiography showed moderately left atrial dilatation, ventricular hypertrophy with septal hypokinesia, and an ejection fraction of 50%. The pulsed-wave Doppler in the suprasternal notch found a gradient of 30 mmHg.

Doppler ultrasound of supra-aortic trunks showed bilateral subclavian steal (BSS) phenomenon. Computed tomography angiography of the aorta, supra-aortic trunks and brain vessels showed a two-branch aortic arch with a severe in-between narrowing (Fig. 1). The pre-stenotic branch was a bi-carotid trunk and the post-stenotic was a bi-subclavian trunk._

External carotid arteries were enlarged and a retropharyngeal aneurysm was present in the right carotid artery (Fig. 2 panel A: RPA). Vertebral arteries were dilated and there was a well-developed cervical collateral circulation (Fig. 2 panel B: Arrow).

Coronary computed tomography was normal.

We indicated surgical treatment of the coarctation. The usual technique was used. A median sternotomy and pericardial incision were performed under general anesthesia with cannulation of the right femoral artery, left primitive carotid artery, right primitive carotid artery, and right atrium. The constricted aortic segment was resected and bypassed by inserting a Dacron tube (n°20). The procedure was difficult because the aortic wall was very thin and fragile adjacent to the coarctation. End-to-end anastomosis was done by two layers and reinforcement with a Teflon strip was performed.

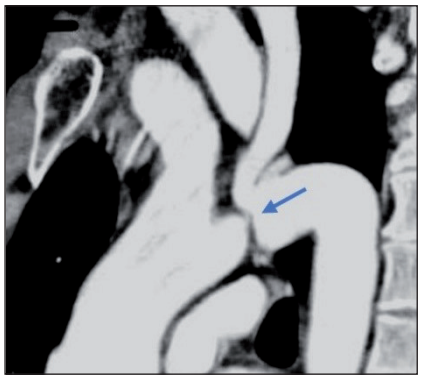

Fig. 1 – Aortic CT-scan – volume rendering and multiplanar reconstructions: Severe mid-arch narrowing (arrow) with a prestenotic bi-carotid trunk and a post-stenotic bi-subclavian trunk.

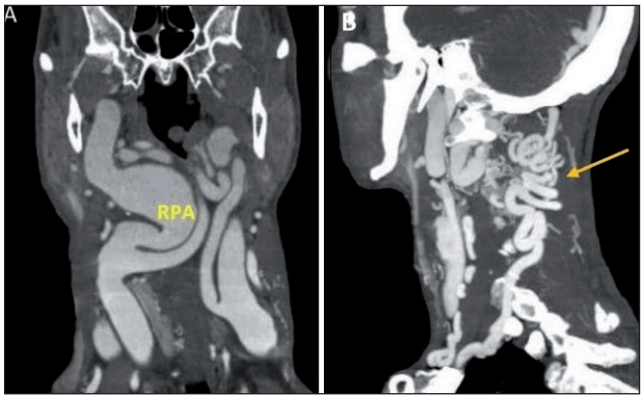

Fig. 2 – (A) Coronal reconstruction showing the enlargement of external carotid arteries and the retropharyngeal aneurysm (RPA). Panel (B) shows the well-developed cervical collateral circulation (arrow).

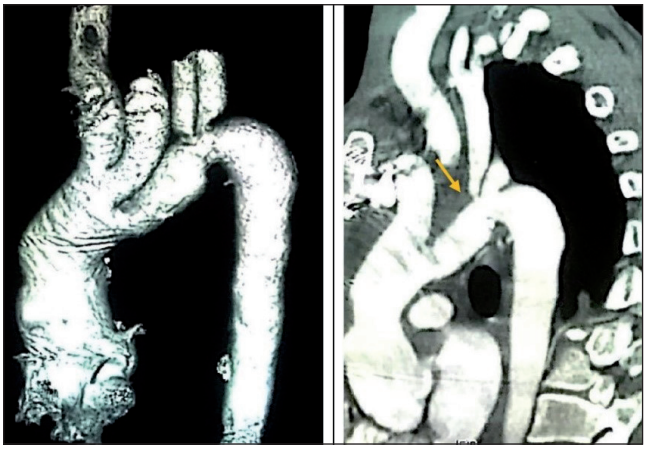

Fig. 3 – CT-angiography: Volume rendering reconstruction showing the permeable Dacron tube and the stenosis at the origin of the right subclavian artery (arrow).

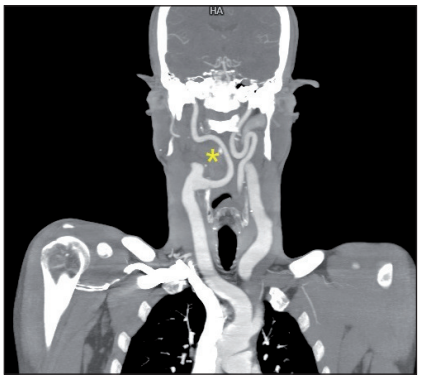

Fig. 4 – Angio-MRI: Coronal reconstruction showing no retropharyngeal aneurysm (*).

Postoperative recovery was good. At clinical examination, there was no hypertension, femoral and upper limbs pulses were present and symmetric.

CT-angiography and magnetic resonance imaging of the aorta and supra-aortic trunks, 5 years later, showed a permeable Dacron tube, a stenosis at the origin of the right subclavian artery (Fig. 3), an occlusion of the left vertebral artery, and a complete disappearance of the aneurysm with a decrease of the collateral circulation (Fig. 4). These anomalies were respected as they didn't cause any symptoms. Close clinical and radiological surveillance was advised.

Discussion

Classically, aortic arch gives off three branches: brachiocephalic trunk, left common carotid artery and left subclavian artery. This classical pattern was reported to occur in 74.0% to 89.4% of cases in radiological series.[6–8]

Many variations in the branching pattern of the aortic arch may occur. Classifications take into consideration the aortic arch (interrupted, right sided, left sided, double aortic arch, or cervical aortic arch) and the order or pattern of branching of the great vessels.[9]

The incidence of aortic arch anomalies varies between authors (2.6–35.1%).[10] They are noted in 1–10/1,000 live births and account for 15–20 % of all congenital heart diseases.[11]

The present case report describes an unusual constellation associating an avian aortic arch with a mid-arch narrowing between the two common trunks.

Aortic coarctation accounts for 5–8% of all congenital heart defects.[12] It is a narrowing of the aorta most commonly found just distal to the origin of the left subclavian artery. Mid-aortic arch coarctation is rare,[1] and represents 2.5–3.5% of all aortic coarctations according to medical and surgical series.[2] Anomalies of the arch vessels associated with coarctation are unusual.[3] Reinshagen et al. observed a higher prevalence of bi-carotid trunk in patients with aortic coarctation (17.8% vs 14.8%). However, they could not prove a significant association between bi-carotid trunk and aortic coarctation.[13]

Avian arch is a two-branch arch.[4] It seems to be exceptional in humans with a prevalence rate of 0.16–1%.[5]

Although the two components of this specific constellation (Avian arch + mid-arch coarctation) are well identified in isolation, their association seems very rare.

Clinical presentation of aortic arch anomalies is highly variable. Many patients remain asymptomatic throughout life and these variations are discovered incidentally.[14] Symptomatic forms generally result from compression of surrounding mediastinal structures by vascular ring.[11] Furthermore, the anomalous origins of the large aortic arch vessels can lead to changes in cerebral hemodynamics and cause cerebral abnormalities.[15]

In our case, symptoms were essentially consecutive to the alteration of normal blood flow resulting in a bilateral subclavian steal (BSS). BSS is a rare condition usually due to reversal of vertebral blood flow in the setting of proximal subclavian or innominate artery severe stenosis or occlusion. Most patients with subclavian steal are asymptomatic. In symptomatic cases, patients present with arm ischemia symptoms including exertional pain and weakness.[16] Symptoms of vertebrobasilar ischemia of brainstem such as dizziness, vertigo, and syncope can also occur.[17] Our patient presented with both arm ischemia signs (exertional pain and weakness) and vertebrobasilar ischemia signs (dizziness).

Avian arch variation is often asymptomatic,[17] but in this specific case, the particular configuration has probably influenced the clinical expression of the coarctation. Our patient had neither collateral circulation nor hypertension. The origin of the subclavian arteries distally to the coarctation may probably explain this, and explain the absence of discernible differences in pulse pressure between right and left arm.

No specific treatment is needed for avian arch,[4] but in this case, surgical correction of the coarctation was clearly indicated. Mid-arch coarctation presents difficulties in the conduct of surgical repair.[1] In addition, in the present case, a special attention had to be granted to the presence of a bi-carotid trunk as it can lead to some serious neurologic complications during the surgical procedure.

Conclusion

Aortic arch anomalies have called the attention of several authors especially with advancements of endovascular interventions and complex surgical procedures. Their knowledge is of crucial importance.

We have reported here an interesting case of an extremely rare association of an avian aortic arch and a mid-arch coarctation.

Conflict of interest
The authors declare that they have no competing interests.

Funding
No funding was received.

Informed consent
Written informed consent was obtained from the patient for publication of this case report and accompanying images.

Authors' contributions

All authors have contributed equally to the work. [HK] and [MA] took care of the patient and contributed to the material preparation. [EB] and [AS] wrote the first draft of the manuscript and contributed the material preparation as well. [IK], [SM] and [SK] revised the manuscript and suggested some correction along the way.
All authors read and approved the final manuscript.

References

1. Katsumata T, Westaby S. Operation of mid-arch coarctation. Ann Thorac Surg 1999;67:1386–1390.
2. Winer HE, Kronzon I, Glassman E, Cunningham JN Jr, Madayag M. Pseudocoarctation and mid-arch aortic coarctation. Chest 1977;72:519–521.
3. Ellis FH Jr, Clagett OT. Coarctation of the Aorta Proximal to the Left Subclavian Artery: Experience with Six Surgical Cases. Ann Surg 1957;146:145–151.
4. Ergun O, Tatar IG, Durmaz HA, et al. A case of a rare aortic arch variation: the avian form. JBR-BTR 2014;97:349–350.
5. Rueda-Medina I, García-Ruíz R, Del Saz-Saucedo P, Turpín-Fenoll L. Common origin of carotid and subclavian arteries as an exceptional anatomical variant of the aortic arch. Neurología 2011;26:244–255.
6. Jakanani GC, Adair W. Frequency of variations in aortic arch anatomy depicted on multidetector CT. Clin Radiol 2010;65:481–487.
7. Natsis KI, Tsitouridis IA, Didagelos MV, et al. Anatomical variations in the branches of the human aortic arch in 633 angiographies: clinical significance and literature review. Surg Radiol Anat 2009;31:319–323.
8. Boyaci N, Dokumaci DS, Karakas E. Multidetecor computed tomography evaluation of aortic arch and branching variants. Turk Gogus Kalp Dama 2015;23:51–57.
9. Stojanovska J, Cascade PN, Chong S, et al. Embryology and imaging review of aortic arch anomalies. J Thorac Imaging 2012;27:73–84.
10. Karacan A, Türkvatan A, Karacan K. Anatomical variations of aortic arch branching: evaluation with computed tomographic angiography. Cardiol Young 2013;22:1–9.
11. DeRoo S, Dillman JR, Cascade P, et al. Aortic Arch Anomalies. In: Saremi F (ed.) Cardiac CT and MR for Adult Congenital Heart Disease. Springer: New York, NY, 2014.
12. Tsikrikas S, Charalampous C, Letsas KP. A case of severe asymptomatic aortic coarctation in an adult female. Case Rep Cardiol 2011;2011:535463.
13. Reinshagen L, Vodiskar J, Mühler E, et al. Bicarotid Trunk: How Much Is "Not Uncommon"? Ann Thorac Surg 2014;97:945–949.
14. Türkvatan A, Büyükbayraktar FG, Ölçer T, Cumhur T. Congenital Anomalies of the Aortic Arch: Evaluation with the Use of Multidetector Computed Tomography. Korean J Radiol 2009;10:176–184.
15. Lemke AJ, Benndorf G, Liebig T, Feli R. Anomalous origin of the right vertebral artery: review of the literature and case report of right vertebral artery origin distal to the left subclavian artery. AJNR Am J Neuroradiol 1999;20:1318–1321.
16. Amini R, Gornik HL, Gilbert L, et al. Bilateral Subclavian Steal Syndrome. Case Rep Cardiol 2011;2011:146267.
17. Lord RS, Adar R, Stein RL. Contribution of the circle of Willis to the subclavian steal syndrome. Circulation1969;40:871–878.