CAS CLINIQUE / CASE REPORT

ABERRANT ORIGIN OF THE RIGHT VERTEBRAL ARTERY IN A PATIENT WITH TRUNCUS BICAROTICUS AND ABERRANT RIGHT SUBCLAVIAN ARTERY

A Possible Baroreceptor Nidus

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Rami EL HAGE[1*], Fouad BANDALY[1], Georges TABET[1]

El Hage R, Bandaly F, Tabet G. Aberrant origin of the right vertebral artery in a patient with truncus bicaroticus and aberrant right subclavian artery: a possible baroreceptor nidus. J Med Liban 2017 ; 65 (4) : 220-224.

ABSTRACT • Multiple aortic arch variations have been reported. Most of them are asymptomatic incidental findings, while others, like an aberrant right subclavian artery, can be symptomatic and require a surgical intervention. In this case report, we present the hemodynamic effects they might produce along with a short review of the literature. We, hereby, present the case of a patient with truncus bicaroticus, aberrant right subclavian artery and a right vertebral artery arising from the right common carotid artery. The patient was undergoing thymoma resection when she had two episodes of hypotension and bradycardia upon manipulation of the right vertebral artery origin. The presence of an ectopic baroreceptor is discussed.

Keywords : hypotension; bradycardia; pressoreceptors; vertebral artery/abnormalities; subclavian artery/abnormalities; carotid arteries/abnormalities

El Hage R, Bandaly F, Tabet G. Possible nidus de barorécepteurs sur une origine aberrante de l'artère vertébrale droite chez un patient ayant un truncus bicaroticus et une artère sous-clavière droite aberrante. J Med Liban 2017 ; 65 (4) : 220-224.

RÉSUMÉ • Plusieurs variantes de la crosse de l'aorte ont été rapportées dans la littérature, mais elles sont le plus souvent asymptomatiques. L'artère sous-clavière droite aberrante (Arteria lusoria) par contre est symptomatique parfois et nécessite une intervention chirurgicale. Nous présentons dans ce cas clinique les effets hémodynamiques causés par l'une de ces variantes : il s'agit d'une association entre un truncus bicaroticus – une artère sous-clavière aberrante – et une origine aberrante de l'artère vertébrale droite. Cette variante a été trouvée chez une patiente qui a présenté deux épisodes d'hypotension et bradycardie au cours d'une chirurgie de résection d'un thymome lors de la manipulation de l'origine de l'artère vertébrale droite. Nous discuterons la présence d'un foyer de barorécepteurs à ce niveau

Mots-clés : hypotension; bradycardie; barorécepteurs; anomalies de l'artère vertébrale; anomalies de l'artère sous-clavière; anomalies de l'artère carotide

INTRODUCTION

Multiple aortic arch variations have been reported on aortograms, CT/MRI imaging or cadaveric dissections. Most of them are asymptomatic and are incidental findings [1-3]. Others like aberrant right subclavian artery can be symptomatic in 10% of cases [4] and require a surgical intervention [5]. It is important to identify these variations before any surgical intervention in order to adapt the surgical approach [5] and to anticipate any hemodynamic effects they might produce as described in this case report. The case of a double axillary artery is a perfect example of how an asymptomatic anatomic variation affects surgical approach [6].

CASE PRESENTATION

We report the case of a 76-year-old woman, with a medical history of hypertension, dyslipidemia, coronaropathy and scleroderma with secondary pulmonary hypertension, who had two unexplained episodes of hypotension and bradycardia during resection of a thymoma.

The patient was screened three months earlier for evaluation of her cardiac function. An anterior mediastinal mass was then discovered on cardiac ultrasound. A CT scan of the chest revealed a 4.7 x 4 cm anterior mediastinal mass compatible with a thymoma. The CT scan also revealed a truncus bicaroticus (a common origin of common carotid arteries), an aberrant right subclavian artery and an aberrant origin of the right vertebral artery from the right common carotid artery (Figure 1). The supra-aortic branches emerged from the aortic arch in the following order, from proximal to distal: the truncus bicaroticus, the left subclavian artery and the right subclavian artery (Figure 1).

She had no history of hypotension, bradycardia, vertigo, malaise or dysphagia and was not a smoker. The thymoma was asymptomatic: the patient had no dyspnea and didn't have a chronic cough. The patient was clinically asymptomatic and had no neurological deficits upon repetitive stimulation; all the pulses were palpable and symmetric, and the patient had no audible cervical bruit. Brachial blood pressures were 123/70 mmHg bilaterally. Lab results were within normal range. Given the size of the thymoma, a decision to surgically resect it was taken. A barium swallow to further evaluate the aberrant right subclavian artery was declined by the patient.

[1]Cardiothoracic and Vascular Surgery Department, Hôtel-Dieu de France University Hospital, Beirut, Lebanon.
*Corresponding author : *Rami El Hage, MD*.
e-mail: hage.rami@gmail.com

The patient was put under general anesthesia, and a median sternotomy was performed. The anterior, inferior and lateral sides of the mass were dissected uneventfully. Upon dissection of the superior mediastinal space, the aberrant origin of the vertebral artery was reached. There was a slight bulge at its origin (as shown on the CT reconstruction in Figure 2) that was reminiscent of the carotid bulb. The origin of the vertebral artery was then very lightly manipulated for further dissection, bradycardia (sudden reduction of pulse from 55 to 34 beats per minute) and hypotension (85/50 mmHg) occurred and lasted for 30 seconds. A second episode of bradycardia and hypotension was reproduced when the origin was manipulated 10 minutes later for completion of the dissection.

Further manipulation of this area was avoided afterwards and no further episodes of bradycardia or hypotension occurred. The patient recovered promptly from surgery, and the postoperative course was uneventful. She was discharged on the fourth day following surgery.

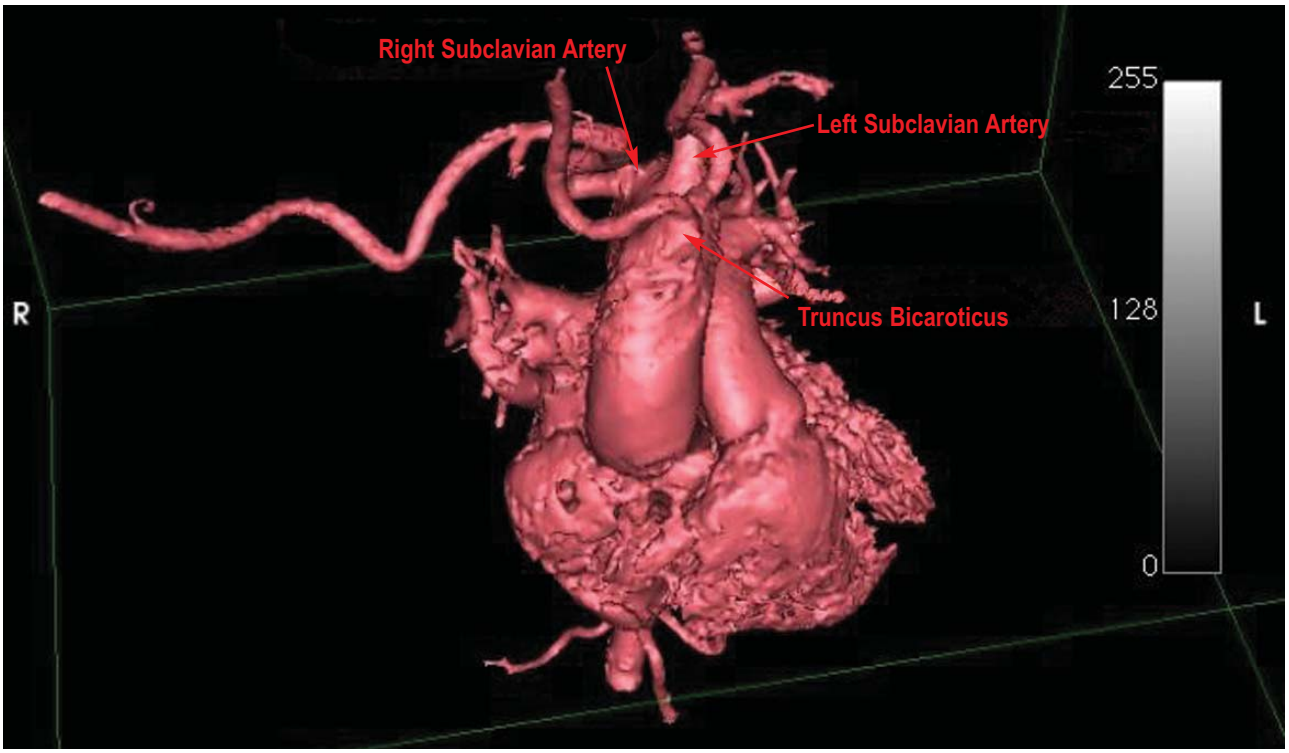

Figure 1. 3D Chest CT reconstruction showing the supra-aortic branches from proximal to distal: truncus bicaroticus, left subclavian artery and the aberrant right subclavian artery.

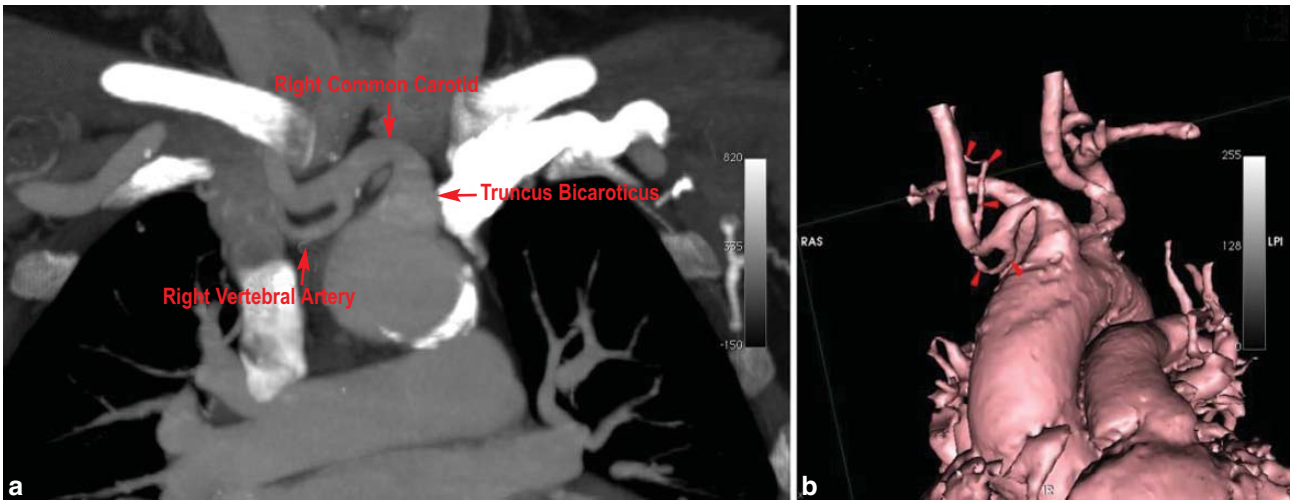

Figure 2. Chest CT reconstruction showing the aberrant origin of the right vertebral artery from the right common carotid. (**a**: Thick slab planar reconstruction **b**: 3D reconstruction).

DISCUSSION

1. Prevalence

Aortic arch anomalies are quite frequent, with the "bovine arch" (common origin of the brachiocephalic trunk and the left carotid artery) being the most common [2] with an incidence of 19.6% [7]. An aberrant right subclavian artery represents 1% of all arch vessel anomalies according to Velioglu *et al*. [2] and Müller *et al*.[8]; whereas it has an incidence of 0.4% to 2% according to Nachtigall *et al*. [7]. There are several types of anomalies that affect the origin of the vertebral arteries. The most common being a left vertebral artery originating directly from the aortic arch [1,2] with an incidence of 2.4% to 5.8% [1,5,8]. The other vertebral artery variants published and cited in case reports or case series, include:

- Right vertebral artery originating from the right common carotid artery: 20 cases (of which 10 were also associated with an aberrant right subclavian artery) [1]. According to Palmer it has an incidence of 0.18% [9].
- Right vertebral artery originating from the aorta: 9 cases [1].
- Duplicated origin of the vertebral artery: 13 cases [1].

A truncus bicaroticus has been found in 29% of cases of an aberrant right subclavian artery [4,7]. And there are several cases in the literature of an aberrant right subclavian artery with an aberrant right vertebral artery origin [1,4,10]. In fact 9.8% of patients with an aberrant right subclavian artery had a right vertebral artery originating from the right common carotid [11].

To the best of our knowledge there's only one case in the literature of a truncus bicaroticus associated with an aberrant origin of the right vertebral artery [12] and only a single case combining the three abnormalities that are present in our patient [5]. (Truncus bicaroticus, an aberrant right subclavian artery and a right vertebral artery originating from the right common carotid artery).

However, it is the first report where these three abnormalities are associated with hypotension and bradycardia upon manipulation of the origin of the right vertebral artery from the right common carotid. The hypotension and bradycardia couldn't be explained by the anesthetics given at the time, and were reproduced by a second manipulation of the trigger area.

2. Baroreceptor development

It is well established that baroreceptors are highly present in the aortic arch and carotid sinuses [13]. They seem to develop at their definitive locations after the aortic arches take on their final disposition [14]. Some sparse baroreceptors are found in the heart and pulmonary vessels, and they are called the "cardiopulmonary receptors". Others are found in the main branches of the aortic arch [13] but they're low in numbers thus they are not known to induce hypotension and/or bradycardia during surgical manipulation of the aortic branches. All of the extra-carotid baroreceptors transmit their afferent information via the vagus nerve [13].

We, therefore, suspect the presence of a nidus containing a high concentration of baroreceptors at the origin of the vertebral artery.

The structure that we manipulated during surgery was the origin of the vertebral artery and not a low carotid bifurcation as demonstrated by the CT scan. Indeed, a thoracic carotid bifurcation is found in 2% of angiograms [15].

3. Totally aberrant aortic arch

Furthermore, in our literature review we found two cases of truncus bicaroticus associated with an aberrant right subclavian artery, an aberrant origin of the right vertebral artery from the right common artery and a left vertebral artery emanating from the aortic arch [2,3].

4. Aortic arch development

Knowledge of normal aortic arch embryologic development is helpful to understand the development of this triad of anomalies.

The aortic arch develops during the sixth to eighth week of gestation [2]. As shown in Figure 3a, there are six pairs of primitive aortic arches, one for each pharyngeal arch. Among those, the third and fourth pairs are associated with the development of the aortic arch system. The third pair of cervical aortic arches gives rise to the left and right common carotid arteries. At seven weeks gestation, both common carotid arteries arise from a common vascular trunk. The persistence at this stage of development in the derivatives of the embryonic ventral aorta gives the vascular pattern called common origin of carotid arteries (truncus bicaroticus). The prevalence of this anomaly is less than 0.2% [2].

The right subclavian artery develops from the 7th cervical intersegmental artery. The caudal part of the dorsal aorta obliterates during development just distal to the confluence of the right dorsal aorta and the C7 intersegmental artery. The vertebral artery arises from the longitudinal anastomosis between the C1 and C7 intersegmental arteries [16].

If the obliteration zone is proximal to the origin of the C7 intersegmental artery, the right subclavian artery becomes the most distal branch of the aortic arch (as depicted in Figure 3b) distal to the left subclavian artery [1]. The right vertebral artery originates from the normal position from the right subclavian artery.

An added abnormal origin of the right vertebral artery origin from the right common carotid artery (Figure 3c) can be explained by:

- A migration of the vertebral artery onto the dorsal aorta [1];

or

- A longitudinal cervical intersegmental artery anastomosis that stops at the level of C6 instead of C7 [1].

The variants of the origin of the vertebral artery from the carotid arteries frequently display persisting embryological vessel and hypoplasia or aplasia of the proximal vertebral artery [1].

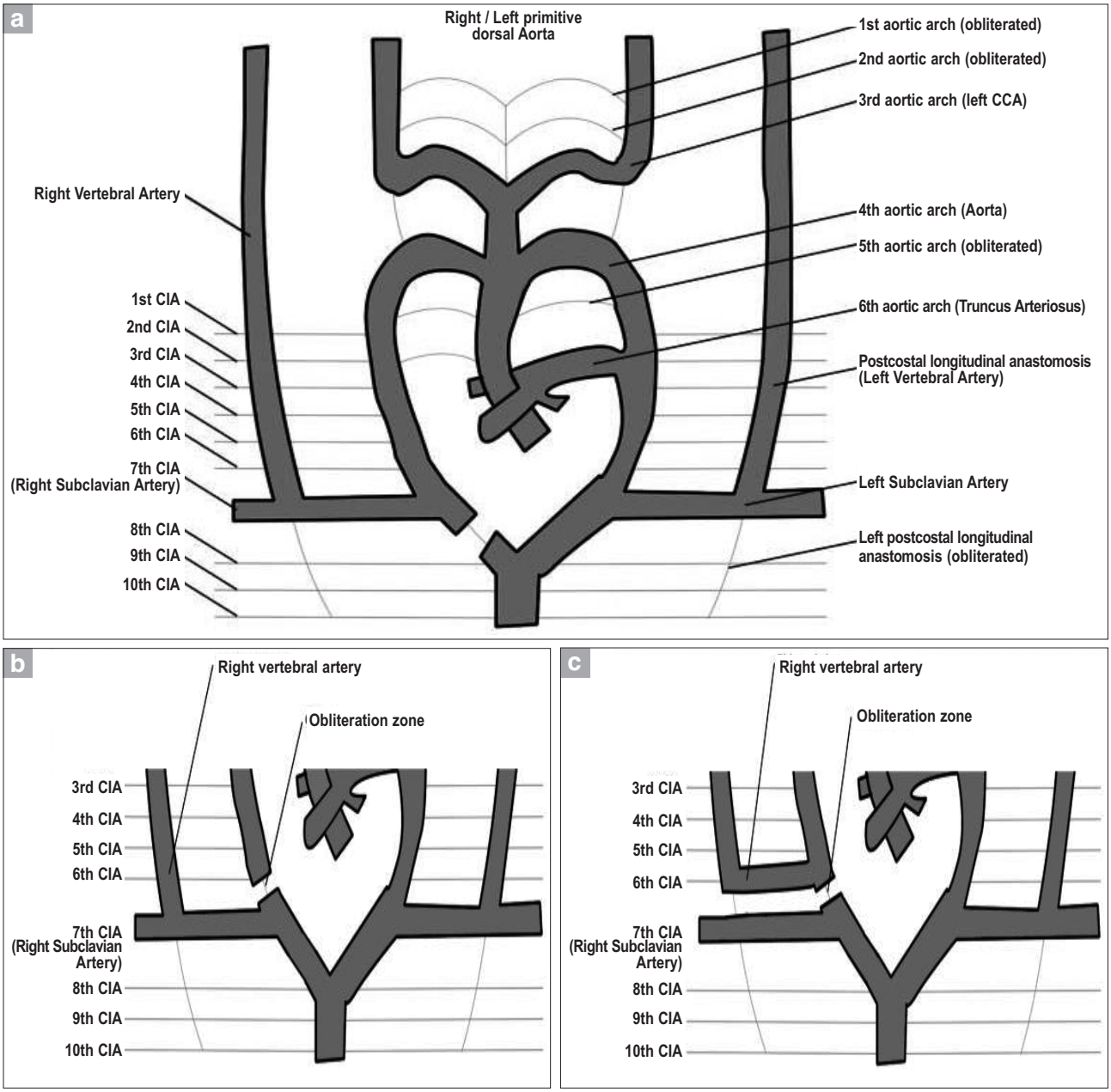

Figure 3. Diagram of aortic arch embryological development: **a.** Normal aortic arch **b.** Aberrant right subclavian artery **c.** Aberrant right subclavian artery with aberrant origin of right vertebral artery.

5. Associated anomalies

In addition, it is hypothesized that anomalous origins of the vertebral arteries predispose the patient to the formation of intracranial aneurysms due to altered hemodynamics [8]. Reports claim that patients with such anomalies should therefore be screened for coexisting aneurysms. However, within the current literature there is no conclusive evidence to suggest that an anomalous origin of the vertebral arteries predisposes an individual to cerebrovascular disorders [8].

Furthermore, some variations like a "bovine arch" have been associated with thoracic aortic diseases [17]. and with other syndromes and anatomic variations. These include esophageal atresia, tracheo-esophageal fistula, DiGeorge anomaly, and anomalous origin of the left coronary artery from the pulmonary artery, congenital polyvalvular disease, truncus arteriosus, aortico-pulmonary window, trisomy 13, 18 and 21 syndromes, acrocephalosyndactyly (especially Apert syndrome), tetralogy of Fallot not associated with DiGeorge anomaly, and clinical Noonan phenotype [5].

6. Treatment

Treatment is indicated when the vascular variants are symptomatic. This is sometimes the case with the aberrant

right subclavian artery, where the patient develops dysphagia secondary to compression of the esophagus by the aberrant right subclavian artery. Its origin can also dilate and form an aneurysm (Kommerell's divertivulum), which needs to be treated either to relieve symptoms due to compression or to avoid rupture of the aneurysm. Open surgical correction has been the mainstay for years, but in recent years multiple cases were treated by hybrid approaches or even total percutaneous approaches [18,19].

CONCLUSION

In summary, an aberrant origin of the right vertebral artery from the right common carotid artery should be handled with care, especially if there's a dilation reminiscent of the carotid bulb. We hypothesize that a nidus containing a high number of baroreceptors is present at this level; however, further evaluation is needed for confirmation.

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