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ANATOMIC VARIATIONS

Right-sided aortic arch with isolation of the left innominate artery and hypoplasia of the left internal carotid artery

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Abstract

Purpose Here, we report a case of the right-sided aortic arch with isolation of the left innominate artery and hypoplasia of the left internal carotid artery.

Methods A 42-year-old male patient underwent a whole-body computed tomography angiography (CTA) examination upon the clinical suspicion of vasculitis.

Results CTA revealed a right-sided aortic arch with the isolation of the left innominate artery and hypoplasia of the left internal carotid artery.

Conclusion The right-sided aortic arch, with the isolation of the left innominate artery, is a scarce vascular variation that may occur with other cardiovascular anomalies such as ventricular septal defect. It can be asymptomatic or can present with symptoms of subclavian steal syndrome. Although its association with the agenesis of the left internal carotid artery has been reported, its association with the hypoplasia of the left internal carotid artery has not been reported previously to the best of our knowledge.

Keywords Right aortic arch · Isolated left innominate artery · Hypoplasia of internal carotid artery · Ventricular septal defect · Subclavian steal

Introduction

The frequency of aortic arch anomalies ranges from 0.5 to 3%. A right-sided aortic arch occurs in approximately 0.1% of adults [4, 5]. The right-sided aortic arch with isolation of the left innominate artery is extremely rare and is associated with structural congenital heart defects and subclavian steal syndrome [2, 9]. The addition of left internal carotid artery dysgenesis to this complex makes this variation even more complex, which is associated with intracranial arterial aneurysms [7, 10].

Case report

A 42-year-old male patient with a medical history of asthma, type 2 diabetes mellitus, and a past surgical history of membranous ventricular septal defect closure in childhood underwent a whole-body computed tomography angiography (CTA) examination because of the clinical suspicion of vasculitis. CTA (Table 1) revealed a right-sided aortic arch with the isolation of the left innominate artery. The right common carotid artery was the first branch arising from the right-sided aortic arch, followed by the right subclavian artery (Fig. 1). The left innominate artery was atretic and had no connection with the aortic arch. The left subclavian and hypoplastic common carotid artery were supplied retrogradely from the left vertebral artery and via a tortuous mediastinal collateral vessel originating from the descending thoracic aorta (Fig. 2). The left common carotid artery was hypoplastic and had a diameter of 3 mm. The initial segment

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Table 1 CT scan parameters (Dual-source CT scanner, Somatom Force, Siemens Healthineers, Germany)

Tube voltage (kVp)	90
Tube current (reference mAs)	334
Gantry rotation time (s)	0.25
Collimation	2×192×0.6
Pitch	0.2
Matrix	512×512
FOV (cm)	351
Scan slice thickness (mm)	0.6
Reconstruction slice thickness (mm)	1.0
Reconstruction interval (mm)	0.5
Contrast material injection parameters	
Contrast phase	Arterial phase (20.s)
Volume (ml)	90 (Opaxol 350/100 mg/ml, Opakim, Ankara, Turkey)
Flow rate (ml/s)	5

of the left ICA (length of 2 mm) was 3.2 mm in diameter, followed by the absence of other ICA segments to the visualized petrous bone level (Fig. 3). Although present, the left carotid canal was smaller in diameter when compared to the right. The caliber of the left vertebral artery arising from the left subclavian artery was thinner than the right vertebral artery.

Also, there were calcifications in the membranous interventricular septum that were likely linked to the closure surgery for the membranous ventricular septal defect. Our patient had no neurological or upper extremity complaints of subclavian steal syndrome. Also, the patient's neurological examination was unremarkable, and there was no history of stroke.

Discussion

The aorta and its branches develop between the 6th and 8th week of embryogenesis. The third set of paired embryonic aortic arches form the common carotid arteries and cervical segments of the internal carotid arteries. Typically, the left fourth embryonic aortic arch forms a part of the adult aortic arch, while the right fourth embryonic aortic arch forms the right subclavian artery. When this pattern is reversed, a right-sided aortic arch forms. The frequency of aortic arch anomalies ranges from 0.5 to 3% and a right-sided aortic arch occurs in approximately 0.1% of adults [4, 5]. The left subclavian artery derives from the 7th intersegmental artery. The left pulmonary artery and ductus arteriosus originate from the proximal and distal parts of the left 6th embryonic arch, respectively.

Right aortic arch anomalies include mirror image branching, which occurs in up to 98% of the right aortic arches. The

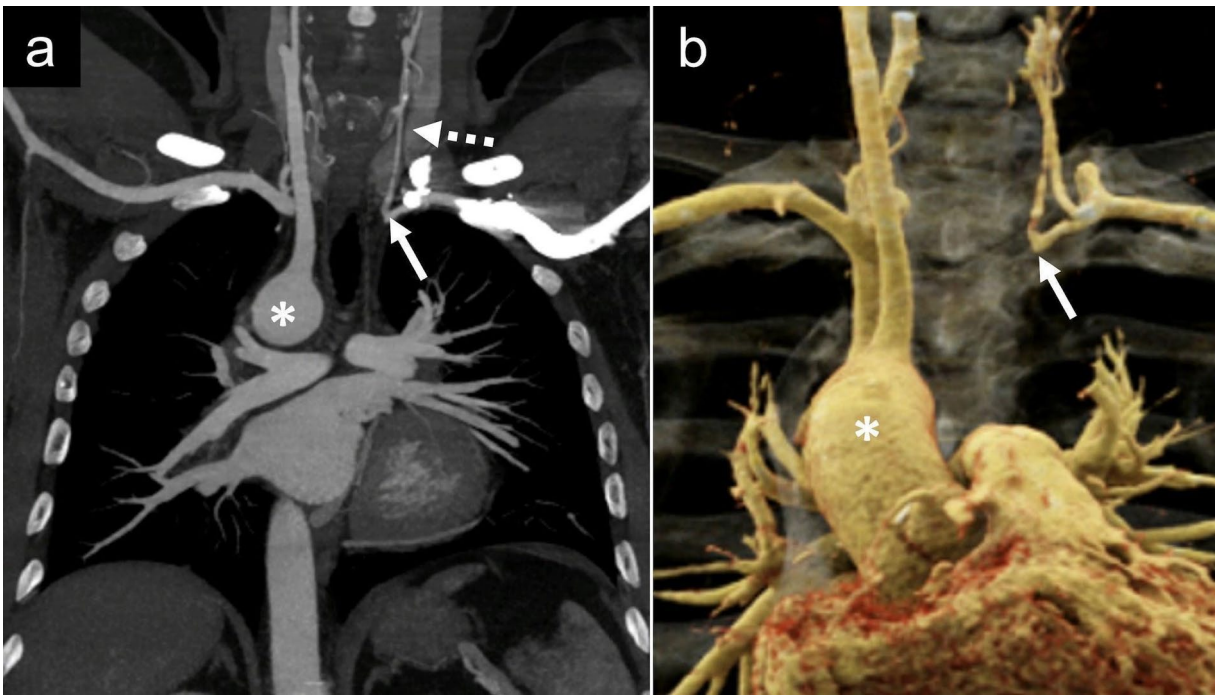

Fig. 1 Coronal maximum intensity projection (MIP) CT image (**a**) and 3D cinematic VRT (volume rendering technique) image (**b**) demonstrate the right-sided aortic arch (a, b, asterisk) with the isolation of left innominate artery (a,b, arrows). The hypoplastic left common carotid artery is also shown (a, dashed arrow)

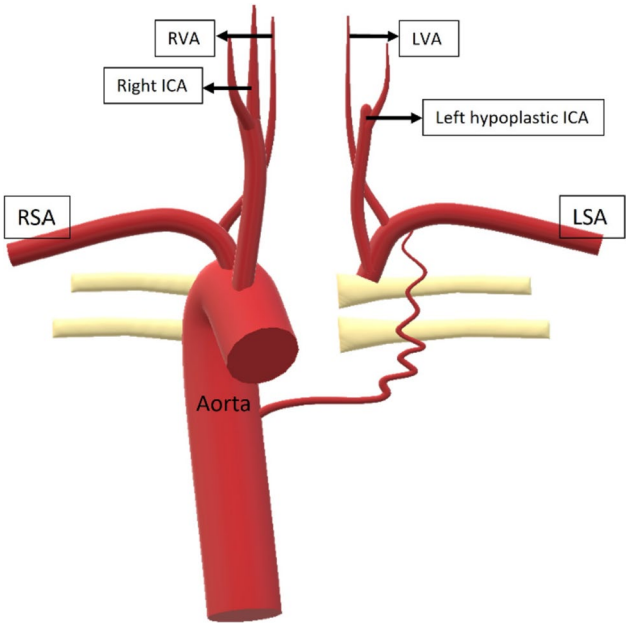

Fig. 2 Schematic arrangements of the vessels of the neck and demonstration of the right aortic arch with isolation of the left innominate artery. The left innominate artery is atretic and had no connection with the aortic arch. A tortuous collateral vessel arising from the descending thoracic aorta supplies the left subclavian and hypoplastic left common carotid artery. ICA internal carotid artery, LSA left subclavian artery, RSA right subclavian artery, LVA left vertebral artery, RVA right vertebral artery

less common types are the aberrant left subclavian artery, isolation of left subclavian artery, aberrant left brachiocephalic artery, and isolation of left brachiocephalic artery, which are explained by the embryological interruption and remodeling of Edward's hypothetical double arch [9].

An isolated left brachiocephalic artery with the right-sided aortic arch can be explained by interruption at two sites in the double aortic arch, in the left anterior arch proximal to the left common carotid artery and the left posterior arch distal to the ductus arteriosus [3]. Persistence or regression of the 6th aortic arch can explain the connection of the isolated left brachiocephalic artery to the pulmonary artery [8]. Isolated left innominate artery may be seen with tetralogy of Fallot, ventricular septal defect, atrioventricular septal defect, pulmonary stenosis, double-outlet right ventricle, coarctation of the aorta, aortic atresia, Down's syndrome, Goldenhar syndrome, CHARGE association and 22q11 microdeletion [2]. The blood supply to the isolated left innominate artery occurs via mediastinal and vertebral pathways [10]. It may be asymptomatic or may show the symptoms of subclavian steal syndrome, such as episodic dizziness, vertigo, and left upper limb claudication due to vertebrobasilar ischemia [4].

Internal carotid artery dysgenesis manifesting as hypoplasia, aplasia, and agenesis is a very rare congenital anomaly occurring in less than 0.01% of the population. Abnormal regression, atresia, or involution of the first and third aortic arches and distal portion of the dorsal aorta lead to internal carotid artery dysgenesis [1, 6]. The term "hypoplasia" is used when a portion of the artery is present, the initial segment of the artery is regular in size or even slightly enlarged proximal to its abrupt narrowing [7]. Hypoplasia or agenesis of the internal carotid artery is associated with intracranial vascular anomalies. Intracranial aneurysms associated with agenesis or aplasia have been reported as 25–43%, much higher than the general population, 2 to 4% [7]. Agenesis or hypoplasia of the internal carotid artery may be asymptomatic. However, it can present with headaches, symptomatic epilepsy, cerebral ischemia, hemiplegia, or intracranial hemorrhage [7, 10]. Left internal carotid artery agenesis associated with the isolation of the left innominate artery has been previously reported, which presents with subarachnoid hemorrhage due to intracranial arterial aneurysm rupture [10].

Conclusion

We reported a case of hypoplasia of the internal carotid artery with the isolation of the left innominate artery in a right-sided aortic arch. The co-occurrence of these anomalies is extremely rare. Isolation of the left innominate artery is associated with structural congenital heart defects and subclavian steal syndrome. In our case, the patient had a history of surgical closure of membranous ventricular septal defect in childhood but did not show the symptoms of the subclavian steal syndrome. Internal carotid artery dysgenesis is associated with intracranial arterial aneurysms. Although not in the scan field of our CT scan, an elective CT angiography of the intracranial vasculature for the presence of arterial aneurysms should be undertaken in these patients.

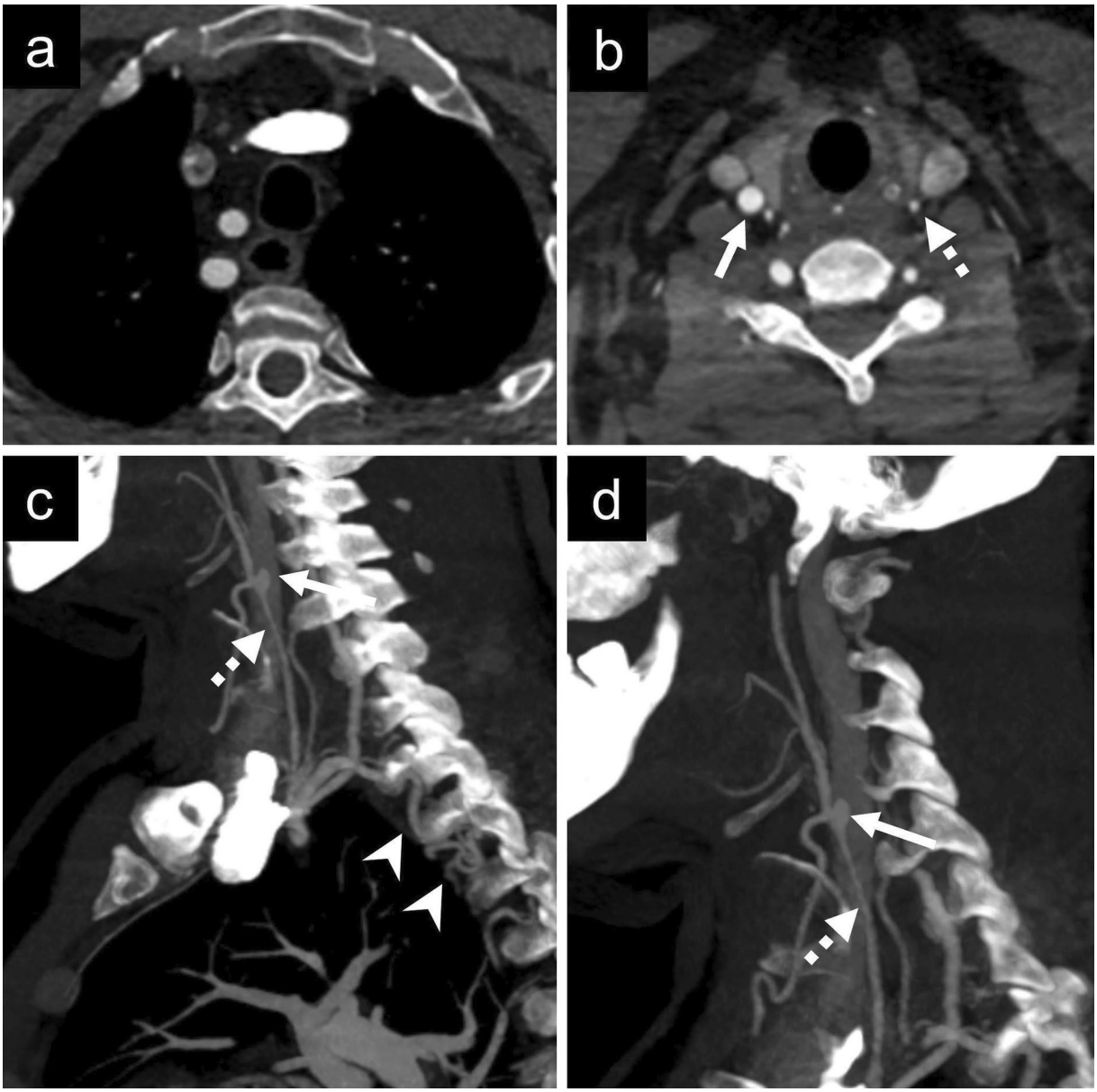

Fig. 3 Axial CT image (**a**) above the aortic arch level shows the right common carotid artery and the right subclavian artery, the two vessels arising from the right aortic arch. In axial CT image at the level of the thyroid gland (**b**), the hypoplastic left common carotid artery (dashed arrow), and the normal right common carotid artery (solid arrow) are seen. Sagittal oblique maximum intensity projection (MIP) images (**c**, **d**) demonstrate the hypoplastic left internal carotid artery (c, d, arrows), hypoplastic left common carotid artery (c, d, dashed arrows), and the tortuous collateral vessel supplying the left subclavian artery (c, arrowheads)

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Declarations

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Ethical approval and consent to participate Not applicable.

Consent for publication The patient has consented to the submission of the case report to the journal.

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