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Case report

Association of tetralogy of Fallot with multiple variations of the cerebral arteries diagnosed by magnetic resonance angiography

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ARTICLE INFO

Article history:

Received 6 December 2019

Revised 5 January 2020

Accepted 7 January 2020

Keywords:

Carotid-anterior cerebral artery anastomosis

Cerebral arterial variation

Nonbifurcating cervical carotid artery

Tetralogy of Fallot

Vertebral artery-anterior inferior cerebellar artery anastomosis

ABSTRACT

Tetralogy of Fallot very rarely manifests with variations of the cerebral arteries, but we detected an extremely rare combination comprising 3 such variants –bilateral carotid–anterior cerebral arteries anastomoses, the vertebral–anterior inferior cerebellar arteries anastomosis (proximal basilar artery duplication) and a nonbifurcating cervical carotid artery– in a 19-year-old man during magnetic resonance angiography to evaluate syncope. Recognition and correct diagnosis of these variations are crucial prior to surgical or radiological cerebrovascular intervention. Both partial maximum-intensity-projection images and source images are useful in identifying these vessels on MR angiography.

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Introduction

Tetralogy of Fallot (TOF) is commonly associated with vascular variations or malformations of the aortic arch or supracar-

diac major venous systems but very rarely with variants of the cerebral arteries. To our knowledge, such association was first reported in 2011 in magnetic resonance angiography of a 2-year-old boy that demonstrated a right persistent hypoglossal artery [1]. The second reported case had 3 variations—a right

Funding: This research received no specific grant from any funding agency in the public, commercial, or not-for-profit sectors.

Acknowledgments: The authors thank Rosalyn Uhrig, M A, for editorial assistance in the preparation of this manuscript.

Declaration of Competing Interest: The authors declare no potential conflicts of interest with respect to the research, authorship, and/or publication of this article.

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<https://doi.org/10.1016/j.radcr.2020.01.008>

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persistent hypoglossal artery, a left carotid–anterior cerebral artery (ACA) anastomosis, and a left accessory middle cerebral artery arising from the A1 to A2 junction of the left ACA [2]. We report a third case demonstrating 3 different cerebral artery variants diagnosed by MR angiography.

Case report

We evaluated a 19-year-old man for syncope he experienced after hitting his toes on the corner of a desk. Born at 36 weeks' gestation and weighing 1996 g, he was diagnosed with TOF within a few days of birth after cardiologic evaluation for a systolic murmur and cyanosis and underwent intracardiac repairs. After this intervention, he appeared in good general health, grew normally, was fairly active, and received regular outpatient follow-up study at our pediatric cardiology department.

After his syncopal event, he underwent magnetic resonance (MR) imaging and MR angiography that utilized a standard 3-dimensional time-of-flight technique, both acquired using a 3-T scanner. No brain lesion was apparent, but imaging demonstrated multiple variations of his cerebral arteries (Fig. 1), including bilateral carotid–ACA anastomoses (Fig. 1b and c), nonbifurcating left cervical carotid artery (Fig. 1d), and anastomosis of the left vertebral (VA) and anterior inferior cerebellar (AICA) arteries (or duplication of the proximal basilar artery [BA]) (Fig. 1e).

We clinically diagnosed that he had hyperventilation syndrome. He experienced no further syncopal attack and underwent regular follow-up study thereafter at our pediatric cardiology department.

Discussion

Numerous reports associate TOF with vascular variations or malformations of the aortic arch or supracardiac major venous system, but variations or malformations of the cerebral arteries are very rarely reported [1,2].

The aortic arch and its branches normally develop from ventral and dorsal arteries, and abnormalities result when structures that should persist regress and those that should regress persist. Possible reasons for anomalies of arch development include (1) deficient growth factors, (2) extrinsic compression, in which mechanical compression from the outside restricts or prevents blood flow, or (3) intrinsic position of the heart that yields preferential streaming of blood. More than 1 factor usually contributes to the anomalous development [3].

Carotid–ACA anastomosis or infraoptic course of ACA

The anomalous ACA arises from the internal carotid artery (ICA) at the level of the ophthalmic artery, pass between the

bilateral optic nerves, and finally anastomoses with the A1–A2 junction of the ACA. The infraoptic course of our patient's bilateral ACAs below the ipsilateral optic nerve and ascension between the optic nerves (Fig. 1b and c) represents a rare variation, with reported prevalence by MR angiography of only 0.086%, that demonstrates right-side predominance [4]. Wong et al [5] reviewed 42 reported cases of carotid–ACA anastomosis and classified them into 4 types. In Type I, the infraoptic A1 acts like a collateral artery, and in Type II, the ICA bifurcates at the level of the ophthalmic artery, and there is an infraoptic A1 but no supraoptic A1. Type III is similar to Type II, but the contralateral A1 is also absent, and Type IV includes an accessory ACA. Types I and II can occur bilaterally, and Wong's group reported 8 bilateral cases among the 42. Ours was a bilateral case of Wong's Type II. Also, they reported that 59% of patients with this variation had associated cerebral aneurysms. Especially, aneurysms are found at the anterior communicating artery complex, probably resulting from hemodynamic stress caused by anomalous blood flow from this variation. Our patient had no aneurysm.

Nonbifurcating cervical carotid artery

The common carotid artery usually bifurcates at the level of the third and fourth cervical vertebrae. Some arteries arise from the proximal segment of the external carotid artery (ECA) just above the bifurcation, but no arteries branch from the cervical segment of the ICA [6,7]. In the very rare case that the cervical carotid artery is nonbifurcating, some branches of the ECA arise directly from the common carotid artery without forming a carotid bifurcation, and the ICA has no physiological dilatation at its origin (carotid bulb). This variation has a reported prevalence by MR angiography of 0.21% and shows no significant laterality [8]. Our patient demonstrated a nonbifurcating left cervical carotid artery with proximal-to-distal order of branching from the lingual, then facial artery, and then distal trunk of the ECA, and the occipital artery arose from the trunk of the ECA (Fig. 1d). Configuration of this variation is agenesis of the proximal main trunk of the ECA, however, agenesis of the proximal segment of the ICA forms this rare variation [8].

VA–AICA anastomosis (proximal BA duplication)

Anastomosis of our patient's left VA with the AICA can also be regarded as duplication of the proximal BA (Fig. 1e). Duplication can occur when 2 embryologically different vessels fail to fuse normally during early embryonic development [9]. Fenestration or duplication of the BA is a common congenital variation that can form when the primitive longitudinal neural arteries fail to fuse during embryologic development at the fifth week of gestation [9,10].

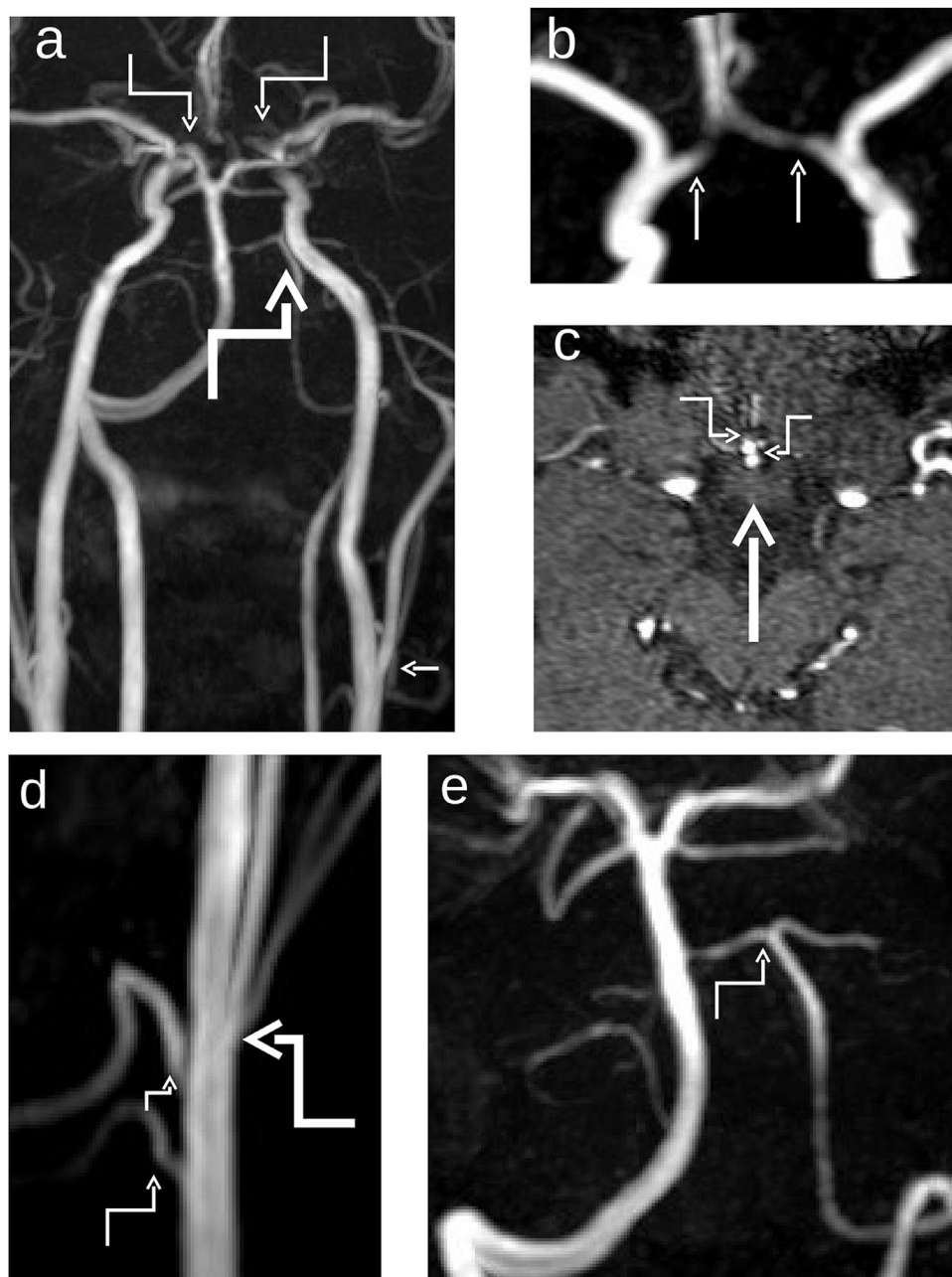


Fig. 1 – (a) Anteroposterior projection of magnetic resonance (MR) angiography shows poor visualization of bilateral A1 segments of the anterior cerebral artery (long arrows). Slightly high position of the left carotid bifurcation is suspected (short arrow). The terminal segment of the left vertebral artery takes an extreme lateral course (large arrow). **(b)** Partial maximum-intensity-projection (MIP) image of the carotid system shows anastomosis of the carotid and anterior cerebral arteries bilaterally (arrows). **(c)** MR angiographic source image shows bilateral anomalous arteries (arrows) are ascending in front of the optic chiasm (large arrow). **(d)** Lateral partial MIP image of the left carotid system shows nonbifurcating cervical carotid artery. The proximal-to-distal branching order the lingual (long arrow), then facial artery (short arrow), and the distal trunk of the external carotid artery (large arrow), from which the occipital artery arises. There is no carotid bulb. **(e)** Partial MIP image of the vertebrobasilar system shows anastomosis between the left vertebral artery and the left anterior inferior cerebellar artery (arrow). This variation also can be regarded as duplication of the proximal basilar artery.

Conclusions

We present what we believe is the third report associating TOF with an extremely rare combination of 3 types of variation of the cerebral arteries that differed from those previously described in the literature. Our 19-year-old patient demonstrated bilateral carotid–ACA anastomoses, a nonbifurcating cervical carotid artery, and anastomosis of the VA and AICA (proximal BA duplication) in MR angiography. Though these findings were incidental, may foreshadow aneurysm formation at their junctions. Recognition and correct diagnosis of these variations are crucial prior to surgical or radiological cerebrovascular intervention. Both partial maximum-intensity-projection images and source images are useful in identifying these vessels on MR angiography.

Ethical standards

We declare that all human and animal studies have been approved by the Melbourne Health Research Ethics Committee and have therefore been performed in accordance with the ethical standards laid down in the 1964 Declaration of Helsinki and its later amendments.

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