Case Report

Basilar vascular system supplied by only right proatlantal intersegmentary artery type 1 with aneurysm and left internal carotid occlusion: a case report and review from the literature

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Abstract

Persistence of proatlantal artery (PA) is a rare condition. More than 40 cases were described in the literature. Aneurysm may involve the PA itself in approximately 2% of cases, most arising from the internal carotid artery (ICA) side of PA. This case was particular because the PA showed a saccular aneurysm on the posterior wall, probably due to atherosclerosis disease and other alterations: plaque ulcerative of ICA, occlusion of left ICA, and aberrant right VA.

Introduction

In the vascular anomalies of the skull base are included anomalies of the circle of Willis, carotid–basilar and carotid–vertebral (VA) anastomoses. Various methods of imaging are available for their detection such as computed tomography angiography, magnetic resonance angiography, and digital subtraction angiography [1,2]. Different studies reported the incidence of primitive carotid–basilar anastomoses as between 0.1% and 1%, and their findings is usually uncommon [3]. These anastomoses are physiological system during the embryogenesis and the failure of their involution contribute to vascular anomalies in the adults [4]. The anastomotic channel between the carotid and VA basilar system are: trygeminal, otic, hypoglossal, intersegmentary proatlantis, and intersegmentalis cervicalis arteries [5,6]. More than 40 cases were described for the persistence of proatlantal artery (PA) [1,7].

Case report

Came to our attention, at interventional radiology of our hospital, a man of 65-year old, who had coronaries artery...
disease, vertigo, balance disorders, and a long history of transient ischemic attacks; he was already examined by color Doppler (CD) ultrasonography in another institute with a finding of left internal carotid artery (ICA) occlusion and stenosis of the contralateral ICA. When he came in our institute, we first decided to repeat the CD examination: it confirmed the occlusion of ICA and showed a VA hypoplasia on the left side, on the right side was described an atheromatous plaque of the ICA defining a stenosis of less than 50%, non-hemodynamically significant, and was detected a vascular trunk with an atheromatous plaque inside that originated from the posterior wall of the ICA with a flow similar to it. The course of this artery was not clear, and it was hypothesized to be an anomaly of the origin of the VA from the ICA (Fig. 1). For further diagnostic assessment and to exclude other vascular anomalies, the patient performed computed tomography angiography of the neck and skull base. It confirmed the atheromatous plaque of the right ICA, showing a little ulceration too, and the unusual vessel that originated anteromedially from the right ICA. This vessel had also a saccular aneurysm on the posterior wall that was not visible at CD examination (Fig. 2). Coronal and sagittal reconstruction showed that a muscular branch originated from it. The right VA was not visible. On the left side, the IC was occluded with an upward rehabilitation, and the VA was hypoplastic terminating directly in the left posterior cerebellar artery. No others abnormalities of intracranial circulation were found.

For his coronary artery disease, during the coronary angiography, the patient also performed a digital subtraction angiography of the epiaortic vessels that confirmed the diagnosis of PA type 1: the anomalous vessel rising from the right internal carotid at the level of C3 (Fig. 3A) curved dorsally at the level of C1 in the occipitoatlantal space, and then, it entered the skull through the foramen magnum (Fig. 3B); here, it proceeded horizontally and dorsally until it joined the basilar system (Fig. 3C).

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Discussion

Embriology and PA description

The first anatomic description of PA was made by Gottshau in 1885, and the carotid basilar and VA anastomoses were authoritatively demonstrated by Congdon (1922) and Paget (1948). These anastomoses originated at the 4-mm embryo stage at development. In this stage, the ICAs extend from the paired dorsal aortae and anastomose with the longitudinal neural arteries at 3 major sites: the trigeminal ganglion, the otic vescicle, and the rootlets of the hypoglossal nerve. These longitudinal arteries are also connected to VA system by cervical intersegmental arteries, and the PA is the most caudal of these arteries. It became the dominant anastomose in the embryo at 5- to 6-mm stage (28-30 days). Two groups classification of PA are described: type I arising from the internal and type II from the external carotid artery. The PA type 1 corresponds to Paget’s PA and accounts for the 57% of cases, the type II to the first cervical intersegmental artery and represents the 38% of cases. There is another variant, the most uncommon, in which the PA originates from common artery bifurcation [7]. The PA originated anteriorly and medially from the ICA at the level of C2-C4 vertebra. It ascends medially along the anterior aspect of the VA bodies up to the suboccipital area, where it curves dorsally toward the atlant, and then enters the skull through the foramen magnum. When PA is large, the VA is usually hypoplastic, and the ipsilateral may
be absent. Hypsilateral or bilateral VA absence or hypoplasia appears in about 50% of cases [7].

**Imaging findings**

Our case presents a large right PA with absence of hypsilateral VA, controlateral VA hypoplasia that terminates in the posterior cerebellar artery. So the only supply artery for the posterior circulation system was the persistent PA. Other common vascular anomalies in the PA persistence were anomalies of circle of Willis and intracranial aneurysms. The anomalies include arterovenous malformation, carotid-cavernous fistula, anomalies due to moyamoya disease and arterial anomalies. Cerebrovascular abnormalities were found in 59% of patients and intracranial aneurysm in 10% of patients [1,8]. Most aneurysms involve the posterior communicating arteries [3]. Recently was described the association with fusiform subclavian aneurysm [8]. Aneurysm may involve the PA itself in approximately 2% of cases, most arising from the ICA side of PA [3,8]. This case was particular because the PA showed a saccular aneurysm on the posterior wall, probably due to atheroscleroses disease and other alterations: plaque ulcerative of ICA, occlusion of left ICA, and aberrant right VA. The association of multiple atheroscleroses stenoses, along with the persistent PA is consistence with data found in the literature [7,9]. Some studies show that the cases of

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**Fig. 3** – DSA of the supraortic trunk in the lateral view (A, B) and frontal view on the right side (C). The first image (A) shows the characteristic course of the PA type 1 (long orange arrow) rising from the right internal carotid (red arrowhead) at the level of C3. In the second image (B), the PA (long orange arrow), curved dorsally at the level of C1 in the occipitoatlantal space, and then, it entered the skull through the foramen magnum. The internal carotid is also visualized (red arrowhead); in the third image, (C) PA (long orange arrow) proceeds horizontally and dorsally until it joined the basilar system (violet arrowhead).
anomalous carotid VA anastomosis associate with ICA ste-
nosis may be successfully treated by carotid endarterectomy
with routine shunting applications. This approach is based
with the risk of cerebral ischemia after carotid clamping and
the perfusion of the ipsilateral hemisphere and subtentorial
structures depend on the controlateral carotid artery [9,10].
The numerous vascular anomalies made very difficult the
surgical treatment in our patient. In fact if the anomalous
vessel is the only source for posterior circulation and if there is
stenosis or occlusion in the carotid system, a posterior cere-
bral ischemia may ensue.

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