Anatomical variations of the posterior circulation: case reports and a review of literature

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Summary

Introduction: The intracranial vascular anatomical variations, although rare, represent an interesting field of research, since many anomalous variants are possible and in most cases they remain asymptomatic. The capability of the cerebral circulation to adapt to several flow changes is confirmed by the fact that in several cases these anatomical variation compensate for an eventual unsuccessful development of the normal circulation, especially in the posterior section of cerebral circulation.

Materials and Methods: A comprehensive review of PubMed literature was performed and three clinical cases have been analyzed.

Results: Several angiographic and MR-angiography reports have been evaluated, regarding general and specific anatomical variants of the posterior circulation.

Discussion: Although rare, the anatomical variations of the posterior intracranial circulation represent an interesting field of investigation in order to achieve a better comprehension of the embryological development of the circulatory system.

Key words
Anatomical variation; intracranial arteries; posterior circulation.

Introduction

The vertebral artery comes from the longitudinal vascular anastomoses between seven cervical intersegmental arteries (the first is the pro-atlantal). The proximal portion last intersegmental artery gives the subclavian and initial vertebral artery, while the other six naturally involve (Komiyama et al., 1999). Normally, after the origin from the subclavian artery, the vertebral artery enters the sixth or seventh transverse foramen, then proceeds upward vertically within the transverse foramen until C1, where it crosses the posterior arch of the atlas before entering the foramen magnum. The vertebral artery has been divided into four segments: the first is between the longus colli and the scalenus anterior muscles (V1 segment), the second is through the transverse processes from C6 to C2 (V2 segment), the third is from C2 to the medial side of rectus capitis lateralis muscle (V3 segment), and the fourth (V4 segment) is that
which becomes intracranial after the perforation of the dura mater and from which the postero-inferior cerebellar artery and the anterior spinal artery arise. At the lower border of the pons, the two vertebral arteries combine to form the basilar artery (Idowu et al., 2010). In a large number of cases, the calibre of the two vertebral arteries is asymmetric and the left one is dominant (Albayrak et al., 2007). The basilar artery is a median vessel that arises from the confluence of the two vertebral arteries, is contained in the pontine cistern within the sulcus basilaris. The basilar artery may be further subdivided into three portions: proximal, middle and distal. The proximal segment gives rise to the antero-inferior cerebellar artery, the middle portion is responsible for the vascularization of the brainstem (short pontine branches) and the distal part terminates into the right and left posterior cerebral arteries and right and left superior cerebellar arteries (Krabbe-Hartkamp et al., 1998). Some studies have described the anatomical variations of the vertebro-basilar circulation (Graves et al., 1996). The dysembryogenetic events that lead to these anatomical variations are not yet well explained. The absence of screening protocols focusing on vascular anatomical variations does not allow a precise evaluation of the incidence of these events and the only available data come from autopsy studies.

**Materials and Methods**

A comprehensive review was assessed by consulting PubMed (http://www.ncbi.nlm.nih.gov/pubmed/) and dedicated books in order to evaluate the incidence and the anatomical features of three specific anomalous conditions concerning the posterior circulation: a persistent trigeminal artery (PTA) associated to basilar hypoplasia, the fenestration of the vertebro-basilar junction and a hypoglossal artery. Furthermore, three cases observed at our institution have been reported, for which informed consent was obtained.

**Results**

Most of the available data are provided by autopsies. The studies reported in literature show that the anatomical variations of the posterior circulation represent a rare condition. Program screenings are obviously not exploitable because there is no expensive, reproducible and not invasive test to evaluate a large number of subjects. We have focused our research on three anatomical variations of the posterior circulation, represented by three clinical cases: a persistent trigeminal artery, the fenestration of the vertebro-basilar junction and a hypoglossal artery.

**Case report 1: persistent trigeminal artery (PTA)**

A 32-year-old woman, asymptomatic, underwent a magnetic resonance (MR) and MR-angiography as a healthy volunteer in a case-control study conducted at our institution for another study, after an informed consent was obtained. A 1,5 Tesla MR-angiography exam performed with TOF (time of flight) and phase contrast sequences showed marked hypoplasia of the basilar artery with normal aspect of both posterior
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cerebral arteries and the presence of a persistent trigeminal artery (Fig. 1a). The PTA originated from the left carotid siphon and supplied the posterior circulation in association to the ipsilateral posterior communicating artery (Fig. 1c-d). Perforating arteries directed to the brainstem and the antero-inferior cerebellar arteries were probably provided by the residual part of the basilar artery. A persistent trigeminal artery is the most common carotid-vertebrobasilar anastomosis and the incidence reported in angiographic and MR-angiographic series varies between 0.1% and 0.6% (Temizöz et al., 2010; Meila et al., 2011; Uchino et al., 2011). Although the reported incidence is about 0.2%, considering the cases not diagnosed and not reported the impact may probably reach 3% (Pereira et al., 2009). The PTA originates from the internal carotid artery (ICA) where the vessel leaves the carotid canal and penetrates the cavernous sinus. The artery passes through the cavernous sinus medial to the ophthalmic branch of the trigeminal nerve via Meckel’s cave and reaches the posterior fossa. The PTA may either run laterally to the dorsum sellae (lateral type) or have a middle course through or over the dorsum sellae to communicate with the basilar artery (medial type) (Meila et al., 2011). The lateral and medial types of PTA have been considered to occur with equal frequency but the greater rarity of the medial type was recently reported with a ratio of lateral to medial types of 11:1 (Uchino et al., 2011).

Saltzman et al. (Saltzman et al., 1959) described three different subtypes of morphological variations of PTA. Type 1 PTA joins the basilar artery between the superior cerebellar arteries and the anterior inferior cerebellar arteries. The basilar artery proximal to the junction is usually hypoplastic and the posterior communicating arteries are absent or poorly visualized. The trigeminal artery supplies both the posterior cerebral arteries and the superior cerebellar arteries. Saltzman type 2 PTA also joins the basilar artery between the superior cerebellar arteries and the anterior inferior cerebellar arteries but the posterior communicating arteries are present and supply the posterior cerebral arteries. Type 3 PTA is a trigeminal artery variant in which the latter directly joins a cerebellar artery [9]: in Saltzman type 3a PTA ends in the superior cerebellar arteries, type 3b PTA ends in the antero-inferior cerebellar artery and type 3c PTA ends directly in the postero-inferior cerebellar artery (Meila et al., 2011). In the early development of the embryo three longitudinal artery systems exist bilater-
ally, two ventral systems i.e. the ventral and dorsal aorta while the third, dorsal one is located on the midline. The dorsal system corresponds to the paired longitudinal neural arteries. The trigeminal artery develops in the embryo at the fourth month stage, connects the distal dorsal aorta with the paired longitudinal neural artery system, and regresses at 7-12 months post natal (Meila et al., 2011).

Case 2: fenestration of vertebro-basilar junction

A 32-year-old woman was admitted to our institution because of frequent episodes of cephalalgia with acute onset and associated dysequia. The patient underwent a MR exam that showed a large aneurysm at the vertebro-basilar junction and a peri-saccular hyperintense signal in T2-weighted sequences revealing mass effect on the brainstem (Fig.2a-b). A digital subtraction angiography confirmed the presence of a large (2x1,5 cm) aneurysm localized on a fenestration of the vertebro-basilar junction that was visualized with injections from both vertebral arteries, that were symmetrical. The 3D angiographic reconstruction showed that the base of the aneurysm was positioned on both branches of the proximal segment of the fenestration (Fig.2c-d).

Fenestration of vertebro-basilar junction is an uncommon anatomical variation secondary to the embryological development of two branches at the junction of the

![Figure 2](image-url)
two vertebral arteries that meet together after a variable distance of a few millimetres. The assessment of the real frequency of fenestration of the basilar artery is difficult since the data vary according to type of series. However, the incidence of fenestration of the basilar artery is reported to be 1.3% to 6% in autopsy series and 0.02% to 0.6% in angiography series and the variety of this range may depend on cases in which fenestrations are very thin and difficult to visualize at angiography (Bentura et al. 2010). In another report, fenestrations or partial duplications of the basilar artery were demonstrated in approximately 0.3 to 5.26% of autopsy series, while they were found only in 0.1 to 1.9% of angiography series (Eustacchio et al., 1997). The basilar artery is the second most frequent site of fenestration of intracranial arteries after vertebral artery (Osborn and Kirk, 1987; Picard et al., 1993); it is reported in 0.6% angiographs and in about 5% of some autopsy series (Wollschlaeger et al., 1967; Takahashi et al., 1973), and may be found along the entire course of the vessel, even if it seems to be more frequent in the proximal third, at the level of the junction of the two vertebral arteries (Yoon et al., 2004). At the fourth month of gestation the bilateral longitudinal neural arteries are connected laterally at multiple locations with the primitive hindbrain plexus. In this period two fusions occur: one at the ponto-mesencephalic sulcus, that leads to the caudal branches of internal carotid arteries, while the other one determines the vertebro-basilar maturation. These processes are frequently completed during the ninth month. The fusion of the neural arteries associated with the regression of the bridging vessels might be a possible explanation for the basilar artery fenestration. This condition is usually observed in the proximal segment of the vessel because of incomplete fusion of the longitudinal arteries, which proceeds in cranio-caudal direction, while the distal segment is rarely interested. Furthermore, it was supposed that vertebro-basilar junction fenestration might be secondary to the persistence of the rostral part of a primitive lateral vertebrobasilar anastomosis. Angioarchitectural characteristics have been studied and described, such as focal defects at both ends of the medial walls of the fenestration, localized absence of the tonaca media, discontinuous elastic laminae, thickening of sub-endothelium in the distal part and thinning in the proximal part (Bentura et al., 2010). The development of an aneurysm at the junction of a fenestrated basilar artery is thought to be due to both alterations in blood flow and anatomical defects of the vessel wall (Yoon et al., 2004; Albanese et al., 2009).

Case 3: Hypoglossal artery

A 44-year old man, asymptomatic except for some cephalalgic episodes, underwent a MR and MR-angiography exam that rised the suspicion of a vascular malformation; a digital subtraction angiography exam then showed a persistent hypoglossal artery (Fig. 3).

The hypoglossal artery is a vestige of the embryonic carotid-vertebro-basilar anastomoses. Normally, the involution of those anastomoses begins when the posterior communicating arteries start to develop (40th day) and is complete during the 4th-5th month of pregnancy. Although these segmental arteries normally obliterate after the 120th-140th day, sporadically such an artery remains patent resulting in a carotid-basilar anastomosis during adult life (Vlychou et al., 2003). First described by Batujeff in 1889 as an accidental findings during an autopsy (Jukic et al., 2001),
Persistent hypoglossal artery is a rare vascular variation with a frequency of 0.02-0.1% at angiography. In these cases the basilar artery arises from the internal carotid artery (Batuieff, 1889; Brismar, 1976; Ouriel et al., 1988); the hypoglossal artery connects the primitive internal carotid artery and the longitudinal neuronal artery, which later becomes the basilar artery (Janzen et al., 2009). After its origin from the extracranial internal carotid artery at a level between C1 and C3 vertebrae, the persistent hypoglossal artery joins the hypoglossal nerve, enters the posterior cranial fossa via the anterior condylar foramen and the hypoglossal canal, and forms the basilar artery with or without anastomosis with the contralateral vertebral artery. A primary hypoglossal artery does not pass through the foramen magnum. Persistence of this primitive anastomosis may partially compensate for inappropriate development of the vertebro-basilar system (Burger et al., 2007). Definitive diagnosis is based on the recognition of an anomalous artery in the enlarged hypoglossal canal (Parmar et al., 2005; Dimmick and Faulder, 2009). Although conventional angiography has been considered as the reference standard for the detection and evaluation of vascular structures, there are diagnostic alternatives with different advantages. Computerized tomographical (CT) angiography is a non-invasive examination, which has the advantage of multiplanar demonstration of the hypoglossal artery by the use of 3D reconstruction techniques. Therefore, CT-angiography provides excellent anatomic localization of the hypoglossal artery in all its parts and depicts clearly its entrance into the hypoglossal canal (Oelerich and Schuierer, 1997; Wagner, 2001). MR imaging and in particular MR-angiography are able to demonstrate the hypoglossal without any contrast medium. MR-angiography, by using a maximum intensity projection program, is able to show the complete carotid-basilar anastomosis without superimposition of other vessels (Chaljub et al., 1995; Hähnel et al., 2001). Both CT and MR-angiography are diagnostic alternatives, which have already been reported in the literature in cases of hypoglossal artery detection and evaluation of pathologic conditions (Kanai et al., 1992; Fujita et al., 1995; Wagner, 2001). A persistent hypoglossal artery is related with alterations of the anatomy of cerebral circulation and, although it can be observed as an incidental finding in cerebral angiography, its presence has been associated with clinical implications (De Caro et al., 1995). The hypoglossal artery is related to several clinical manifestations and conditions (Guerri-Guttenberg, 2009) such as hypoglossal nerve palsy (Al-Memar and Thrush, 1998) glossopharingeal nerve neuralgia, (Baltsavias et al., 2007); alterations of this vessel may occur in aneurysms, hemodynamic insults such as ischemic stroke which stimulate a compensatory flow through the anterior and posterior circulation (Kempe and Smith, 1969; Huynh-Le et al., 2004; Pasco et al., 2004; Kobayashi et al., 2008; Terayama et al., 2011), moya-moya disease (Komiyama et al., 1999), arterio-venous malformations (Yamanaka et al., 1990), atherosclerosis and thrombosis (Fields, 1980; Conforto et al., 2007; Uzawa et al., 2010).
Discussion

An incidence of about 8.6% variations of the intracranial circulation (Poudel and Bhattarai, 2010) is reported in the literature and most variations are asymptomatic since collateral or atypical circulation supplies a normal blood flow. The embryological alterations that determine these variations are not yet well established. The studies carried out by Streeter (1918) and Kaplan et al. (1961) about the development of the vascular system, although not recent, represent a fundamental basis for further analysis. Although rarely, these events may be associated with pathological conditions such as aneurysms (Graves et al., 1996; Ezaki et al., 2003; Albanese et al., 2009) or artero-venous malformations and fistulas (Rafalowska et al., 1995). Some limitations of this study include the absence of series with standardized criteria. Many studies cited the results about series of patients who were investigated with different techniques (MR-angiography or digital subtraction angiography). The absence of a screening protocol does not allow the evaluation of the real incidence of these anatomical variations, since they are in almost all cases asymptomatic.

Conclusions

The anatomical variations of the posterior circulation represent rare events and in most cases they are asymptomatic, for these reasons no screening protocols are indicated. However, further studies may be useful to investigate the embryological development of intracranial circulation.

References


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