CASE REPORTS

Duplicate PICA and anomalous origin of posterior meningeal artery – an uncommon association with a paediatric cerebellar AVM

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Abstract

Duplication of the posterior inferior cerebellar artery (PICA) is a rare anatomical variation and its presence in association with an arteriovenous malformation (AVM) is not yet documented. We report a right-sided, duplicated PICA in association with a cerebellar AVM, clinically presented as repeated primary third and fourth ventricular bleed in a 7 years old female. Considering very early age of presentation, repeated bleeding episodes, presence of intranidal aneurysm and involvement of brainstem structures, therapeutic embolization was done in two sessions. After the first session of targeted embolization carried out through the anatomically distinct and hypertrophied distal PICA segment which was able to substantially reduce the AVM flow, a proximal vessel became distinct. This vessel arose about 0.5 cm proximal to the rostral PICA and divided into a prominent posterior meningeal artery (PMA) characterised by its vertical disposition directed postero-laterally. The other division ran a medial course aligned towards the main PICA trunk. Literature search reveals that till to-date only 5 cases has been documented and such vessels have a right sided predilection of occurrence. The medullary feeder seems to occur most often from the rostral vessel which in our case was the dominant AVM feeder requiring very cautious and controlled embolization technique. We achieved significant obliteration of the AVM at short term follow-up without any significant neurodeficit.

INTRODUCTION

Duplication of the posterior inferior cerebellar artery (PICA) has rarely been documented by angiography in peer-reviewed literature and was incidental in most reports. An association between such anatomic variation and an arteriovenous malformation (AVM) has not been documented. We report here a right-sided, duplicated PICA in association with a cerebellar AVM. The clinical presentation was due to repeated primary third and fourth ventricular bleed in a 7 year old female.

CASE REPORT

A 7-year-old girl presented with two similar secondary headaches, of which the first one resolved with supportive treatment 5 months apart. Each episode was associated with severe headache, recurrent vomiting and neck stiffness. Clinical examination was unremarkable except for neck rigidity. She remained afebrile and did not report any visual symptoms or lateralised deficits. Computed Tomography (CT) brain was suggestive of bleed into the third and fourth ventricles. (Figure 1) Magnetic resonance imaging (MRI) brain with angiography revealed a large posterior fossa AVM arising out of the vertebrobasilar system. It had a deep location involving the vermis and pons. (Figure 2) Routine laboratory workup and markers for other congenital anomalies like hereditary hemorrhagic telangiectasia (HHT) were negative.

Digital subtraction angiography (DSA) revealed fairly large right cerebellar and pontine AVM with compact nidus and dominant feeders from right anterior inferior cerebellar artery (AICA) and posterior inferior cerebellar artery.
Figure 1. Axial computed tomography (CT) brain showing blood in the fourth (A) and third (B) ventricles.

Figure 2. Source image of magnetic resonance angiography (3T) showing arterio-venous malformation (AVM) involving the brainstem (A) and the cerebellar vermis close to fourth ventricle (B). Time of flight sequences lateral (C) and antero-posterior (D) view showing the extent of AVM (pre-procedural MRI images).
Venous drainage was predominantly into right sigmoid and transverse sinus. Intraneurysms were documented, suggesting a potential source of recurrent rupture. Left vertebral angiogram revealed retrograde filling of AVM from right anterior inferior cerebellar artery. (Figure 3) Left vertebral angiography angiogram revealed a hypertrophied PICA and another distinct but faintly opacified vessel arising proximally from V4 segment of vertebral artery, raising curiosity of either a double origin Right PICA or, more rarely, a duplicated PICA. (Figure 4)

She was managed with differential diagnosis of duplicate PICA, double origin PICA, or fenestrated PICA. Considering the very early age of presentation, repeated bleeding episodes, intraneurysms and involvement of brainstem structures, a decision was made for multi-stage therapeutic embolization. After detailed counselling and obtaining informed consent, two sessions of targeted embolization were carried out. We achieved obliteration of the AVM at short-term follow-up. Mild lower motor neuron type right facial palsy was observed after the second setting, which recovered completely. (Figure 5)

**DISCUSSION**

We gained better insight into the proximal vessel under discussion after the first session of targeted embolization carried out through the anatomically distinct and hypertrophied distal PICA segment, which substantially reduced the AVM flow with significant clarity of the vascular anatomy. (Figure 4) This vessel arose about 0.5 cm proximal to the rostral PICA and divided into a prominent posterior meningeal artery (PMA) characterised by its vertical disposition directed posterolaterally. The other division ran a medial course aligned towards the main PICA trunk. The origin of the PMA is typically from the vertebral artery. It has
Figure 4. Microcatheter within rostral posterior inferior cerebellar artery (PICA) depicting the typical curves of the vessel (A). The cursor is pointing towards the origin of caudal PICA (B); magnified view of origin of proximal PICA and its branching (C); the posterior meningeal artery marked out with black arrows (D) (post-procedural digital subtraction angiography (DSA) images).

Figure 5. Gradient echo (GRE, 3T magnetic resonance imaging) sequence showing embolic cast in the AVM (A) and (B). Magnetic resonance angiography time of flight sequences (3T) revealing significant diminution of arteriovenous malformation (AVM) flow (C) & (D) (post-procedural MRI images)
also been described rarely from the occipital artery, neuro-meningeal trunk (hypoglossal artery) of the Ascending pharyngeal artery and the PICA. To date, six such anomalous origins of PMA have been described, of which on four occasions it arose from the lateral medullary segment of PICA. It is challenging to differentiate between the entity of duplicated PICA and double origin of PICA. We choose the term duplicated PICA in our subject based on the existing knowledge that in double origin of pica the two vessels should converge. However, in a high flow AVM setting, such convergence would merit both origins to be hypertrophied rather than one. Origin of the PMA from the caudal vessel also lends credence to this fact. The prevalence of duplicated PICA is hard to establish. A literature search reveals that to date, 6 cases have been documented, including ours. (Table 1) Curiously, such vessels have a right-sided predilection of occurrence. The medullary feeder seems to occur most often from the rostral vessel, which in our case was the dominant AVM feeder requiring a very cautious and controlled embolization technique.

In conclusion, duplicated PICA is a very rare anatomical variant. This variation has right-sided predilection in the documented cases so far. It is difficult but possible to differentiate between a duplicated PICA and double origin of PICA. PMA originating from PICA is an uncommon vascular variation. The presence of such a congregation of vascular variation in a subject with AVM possibly points out to some inherent issues of vascular differentiation occurring at an early embryologic period.

**DISCLOSURE**

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**REFERENCES**