Fenestration of the middle cerebral artery in a patient who presented with transient ischemic attack

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Abstract
Cerebral artery fenestrations are usually detected incidentally during angiography, have a reported incidence ranging from of 0.03% to 1%, and rarely cause neurological symptoms. They can, however, be associated with aneurysmal dilatation at the proximal or distal end of the fenestration, cerebral arteriovenous malformations, or (rarely) ischemic symptoms. We present a case of a 54-year-old obese woman who presented with a large convex-lens-like fenestration of the right middle cerebral artery (MCA) at the M1 segment (distal to the origin of the temporal pole) associated with a transient ischemic attack. The MCA fenestration caused a local change in hemodynamic blood flow, which leads to cerebral ischemia. Magnetic resonance angiography (MRA) also revealed an associated small slit-like fenestration of the basilar artery (BA), hypoplasia of the A1 segment of the right anterior cerebral artery, bilateral fetal posterior cerebral arteries, and bilateral absence of the posterior communicating arteries. To our knowledge, this is the sixth reported case of MCA fenestration with an associated ischemic attack. In our case, fenestrations of the MCA, the BA, and hypoplasia of the A1 segment of ACA were not associated with any aneurysms.

Keywords: fenestration, middle cerebral artery, transient ischemic attack.

Introduction
A vascular fenestration is a partial duplication, or luminal division, within a vessel segment that results in two distinct endothelium-lined channels with surrounding muscularis tunica [1]. Fenestrations of the cerebral arteries most commonly occur in the anterior communicating artery (ACoA) [2], the vertebrobasilar system [3, 4], the anterior cerebral artery (ACA) [5], the middle cerebral artery (MCA) [6–9], and the posterior cerebral artery [10].

Cerebral artery fenestrations are generally detected incidentally during angiography [7] and have a reported angiographic incidence ranging from 0.03% to 1% [10]. Patients who have cerebral fenestrations rarely have associated neurological symptoms. Cerebral fenestrations can, however, be associated with aneurysmal dilatation at the proximal Uchino et al. [9] or distal [3] end of the fenestration. Additionally, fenestrations may also be associated with cerebral arteriovenous malformations [11] or ischemic symptoms [7]. According to Jeong et al. [7], if a fenestration is detected in an artery relevant to the vascular system, it should be carefully considered as an additional or causative factor for the symptomatology. Finding a causative relationship between a cerebral artery fenestration and presenting clinical vascular symptoms remains a continuing challenge [9].

In the majority of cases of fenestration of the MCA with associated neurological symptoms, there is an associated aneurysm. From 1962 (the first case report of MCA fenestration) to 2010, there were 67 reported cases of MCA fenestration associated with aneurysms [12]. Furthermore, the documented cases of MCA fenestration associated with cerebral ischemia are exceedingly rare [7].

In the present case report, we present a patient who presented a large convex-lens-like fenestration of the right MCA with a transient ischemic attack. Magnetic resonance angiography (MRA) revealed (i) an associated small slit-like fenestration of the basilar artery (BA), (ii) bilateral absence of the posterior communicating arteries, and (iii) hypoplasia of the A1 segment of the ACA (Figure 1). To our knowledge, this is the sixth reported case of MCA fenestration with associated ischemic attack. The following case will delve into the relevant anatomy, embryology and clinical significance of this anatomic variation.

Case report
We report a case of a 54-year-old obese woman with a past medical history of hypertension, diabetes mellitus, and dyslipidemia who presented to our neurosurgical clinic for an evaluation of an episode of transient left hand clumsiness and dysarthria lasting “a few minutes” one day ago. This was preceded by a sub-acute onset of neck

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CASE REPORT

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stiffness and vomiting five days prior. The patient’s social history was remarkable for a 15 pack-year smoking history and daily intake of non-steroidal anti-inflammatory drugs. Clinical examination was completed using MRA performed at the Neuromed Diagnostic Imaging Center (Timișoara, Romania) using a 1.5-tesla MR scanner. Measurements of the length and diameter were performed using multi-planar reconstruction (MPR) technique. Continuity between the arterial lumens and fenestration arms was performed using shaded surface display (SSD) (virtual arterial endoscopy).

Magnetic resonance imaging (MRI) angiography revealed a large, convex, lens-like fenestration of the M1 segment of the right MCA, just distal to the origin of the temporopolar artery (TPA) (Figure 2). The MCA fenestration had a length of 8.0 mm and a width of 6.6 mm located at the proximal third of the fenestration. The upper arm of the fenestration had a length of 12.8 mm and a diameter of 1.2/0.8 mm in the middle third. The lower arm had a length of 10.5 mm and was flat in cross section and a diameter of 0.9/0.4 mm in the distal third. Additionally, fenestration of the MCA was associated with a small slit-like fenestration of the BA, a hypoplastic A1 segment of the right ACA, bilateral origin of PCA from ICA (Figure 3), and bilateral absence of the PCoA. No other anatomical variants were detected.

Our patient had multiple vascular risk factors including: hypertension, obesity, diabetes mellitus, dyslipidemia and smoking. The association between MCA fenestration [with a significantly decreased lumen of the lower arm (Figure 4)] and cerebral ischemia suggests that the fenestration disturbed blood flow and caused cerebral ischemia. After an extensive evaluation of the clinical symptoms and imaging examinations, the patient was put on specific medications.
Figure 3 – Three-dimensional magnetic resonance angiography (3D MRA) imaging of the cerebral circulation. (A) Left medial view, with the origin of the fetal type of right posterior cerebral artery, and the presence of a large convex-lens-shaped fenestration of the M1 segment of the right middle cerebral artery. (B) Right medial view, with the origin of the fetal type of left posterior cerebral artery. ACA, anterior cerebral artery; F, fenestration; ICA, internal carotid artery; L, left; LA, lower arm; MCA, middle cerebral artery; PCA, posterior cerebral artery; R, right; UA, upper arm.

Figure 4 – The morphological appearance of the large convex-lens-shaped fenestration of the M1 segment of the right middle cerebral artery at different levels of section. (A) Multiplanar reconstruction (MPR) technique images of two first two segments (M1 and M2) of the right middle cerebral artery with the middle cerebral artery fenestration, with marking of origin (B) and ending (I) level of fenestrations, and the proximal third (C and D), middle (E and F) and distal (G and H) thirds of upper and lower arms of fenestration. (B–I) Shaded surface display (SSD) (virtual arterial endoscopy) images of endoluminal aspects of the upper and lower arms of the fenestration. Flat aspect in cross section of the lower arm of fenestration, with a significantly decreased of the lumen diameters.

Discussion

The MCA is a lateral extension of the internal carotid artery (ICA) and is the largest and most complex of the cerebral vessels [13]. The MCA is divided into four segments: M1 (sphenoidal), M2 (insular), M3 (opercular) and M4 (cortical) [14, 15]. The M1 (sphenoidal) segment courses within the sphenoidal compartment of the Sylvian...
fissure, from the point of origin of the MCA to the genu of the insula. It runs posterior and parallel to the sphenoid ridge. The main trunk of the MCA divides in one of three ways: bifurcation (78%), trifurcation (12%), or division into multiple trunks (10%) [14]. The most frequent branching pattern is bifurcation (73–78% of cases) [14]. The point at which the M1 segment of the MCA ends varies among authors. Gibo et al. [14] states that the bifurcation point of the MCA is located in the M1 segment, which is subdivided into pre-bifurcation and post-bifurcation parts. Türe et al. [16] considers the demarcation between M1 and M2 as the bifurcation point of the MCA.

There are four anatomical variations of the MCA: duplication, accessory, early branching, and fenestration. In 1962, Crompton [17] reported the first case of MCA fenestration in 347 MCAs examined at autopsy. The incidence of MCA fenestration varies from 0.02% to 1% according to the methods of investigation. The incidence has been reported as 0.17% to 1% by autopsy [17, 18] and 0.02% to 0.43% in angiographic studies [6, 19, 20].

Contrary to fenestrations located in other arterial cerebral and cervical segments, the mechanism underlying the formation of MCA fenestration remains uncertain [6]. Early report of Padge [21] reveals that during the normal development of the cranial arterial system (35 days of embryo-fetal life), the distal primitive ICAs initially divides into a large branch (the future anterior choroidal artery) and numerous small arterial twigs. These arteries normally coalesce into the definitive MCA. The persistence of more than one of these arterial twigs leads to the appearance of fenestrations. This process is favored by the early branching TPA. It is hypothesized that this process could either interfere with the coalescence of primitive arterial twigs into a common proximal MCA (M1 segment) or maintain the patency of collateral channels that would normally involute [6].

Okudera et al. [22] classified the fenestrations of the M1 segment of the MCA into three distinct types based on their location: the proximal type at the level of proximal portion of M1; the intermediate type at the level of the central portion of the M1; and the distal type at the level of the distal portion of M1, just prior to bifurcation. Fenestrations of the proximal type of M1 are the most common, comprising anywhere from 60% [7] to 100% of cases [8]. The right side is most commonly affected, with a reported incidence between 50% [9] to 80% of cases [7].

Gailloud et al. [6] reported five cases of MCA fenestrations. In all five cases, there was early branching of the TPA form the inferior limb of the fenestrated segment. Uchino et al. [9] analyzed six cases of MCA fenestrations and reported early branching of the TPA in four out of six cases (67%). In two (33%) cases, however, the TPA arose from the fenestrated segment. In our case, the M1 MCA fenestration was located just distal to the origin of the TPA.

Uchino et al. [11] described two types of fenestrations of the BA by using MRI angiography: small slit-like fenestration and large convex-lens-like fenestration. In 2006, Uchino et al. [9] analyzed six fenestrations of the MCA, five of which were at the proximal segment of M1 and one was at the middle segment. All the fenestrations were of the slit-like type. In our case, there was a large, convex-lens-like fenestration of the M1 segment of the right MCA, just distal to the origin of the TPA.

Bharatha et al. [23] analyzed a total of 504 sequential cerebral CT angiography studies, which revealed a total of 53 fenestrations (10.5%), including BA 2.4% (two cases) and MCA 0.4 (two cases). The difference in frequency between the fenestrations of BA and MCA has also been reported in other studies [3, 4, 6, 7, 9, 18, 19, 24].

Most of the reports of intracranial arterial fenestrations have been that of single arterial fenestrations, with a minority of cases containing double fenestrations. Uchino et al. [4] reviewed MR angiographic images of 3327 patients and revealed 92 patients with 93 fenestrations of BA (2.80%). According to Bayrak et al. [24], the fenestrated segments of BA are generally lying in close proximity to the vertebrobasilar junction. The fenestrated BA in our case appeared similarly.

Koh et al. [25] has described multiple fenestrations (arrows) resembling plexiform network of the distal A1 right ACA. Aktüre et al. [26] reported two cases of bilateral A1 fenestration, while Makowicz et al. [27] has reported a case of bilateral fenestration of the ACoA. Bayrak et al. [24], analyzing 395 cases evaluated by CT angiography, reported a total of 53 intracranial arterial fenestrations which were identified in 51 patients. In two patients, the fenestration of the BA was associated with fenestration of the ACoA. In our case, there was an associated large convex-lens-like fenestration of the M1 segment of the right MCA with a small vertebral artery (VA) fenestration located close to the vertebrobasilar junction.

While analyzing the morphology of the ACA, Given & Morris [28] revealed some degree of asymmetry of anterior arteries present in 80% of patients. Hypoplasia is a common variant of ACAs, which is found in 10% of post-mortem examinations [29] and 3% of MRA examinations [9]. According to Makowicz et al. [27], this variant increases the risk and extent of ischemia within the frontal lobe. Common variants of ACAs include aplasia or hypoplasia of the A1 segment. Hypoplasia has been found in 10% (aplasia in 1–2%) of post-mortem examinations [29], while angio-MR has demonstrated hypoplasia of the A1 segment and A2 segment in 3% and 2% of cases, respectively [9]. In these cases, the contralateral A1 segment of ACA is usually dilated. In this way, the ACoA supplies the entire area usually covered by the variant or absent ACA. An early report by Dimmick & Faulder [29] demonstrated, by MDCT angiography, a case of a hypoplasic right A1 segment with associated fenestrated right A2 segment. Furthermore, Uchino et al. [9] reported that 14% of ACoA aneurysms are associated with a hypoplasic A1. The authors postulated that the ACoA aneurysms frequently occur in patients with an asymmetric A1 segment due to hemodynamic stresses. In our case, there was associated hypoplasia of the A1 segment of the ACA with the fenestration of MCA and VA.

Fenestrations of the MCA are usually associated with an aneurysm [9] and rarely with cerebral arterio-venous malformation [11]. In our case, the two fenestrations (MCA and VA) and hypoplasia of A1 segment of ACA were not associated with any aneurysms.
In agreement with Jeong et al. [7], we postulate that the MCA fenestration caused a local change in hemodynamic blood flow, which lead to cerebral ischemia in our patient.

In our case, fenestrations (MCA and VA) and hypoplasia of A1 segment of ACA were not associated with any aneurysms. The MCA fenestration and decreased lumen of the lower arm may have caused hemodynamic changes to blood flow, which lead to cerebral ischemia. It is also necessary to mention that our patient has multiple vascular risk factors including hypertension, obesity, diabetes mellitus, dyslipidemia and smoking.

Conclusions

One of the least common vascular anomalies of the cerebral circulation is the large, convex, lens-like fenestration of the right MCA. The association of MCA fenestration with other cerebral fenestrations, transient ischemic attacks, and segmental hypoplasia of the cerebral artery is extremely rare and generally an incidental finding. Bilateral origin of the PCA from the ICA is rare as well and its association with cerebral arterial fenestrations is extremely rare. Bilateral absence of the PCoA disconnects the anterior cerebral circulation of vertebral basilar system, increasing the risk of posterior ischemic attack. Identification of such anomalies should take precedence prior to planning any neuroradiological interventions or surgical procedures to prevent possible risks.

Conflict of interests

The authors declare that they have no conflict of interests.

References


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