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Case Report

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Fenestration in proximal internal carotid and dissection: A case with unusual location report and relevant literature review

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Abstract

Vascular fenestration is a rare congenital variant, in which a segment of vessels divides into two distinct channels, covered by a layer of endothelium. It is usually asymptomatic, but is sometimes accompanied by other structural abnormalities, such as intracerebral vascular aneurysms.

Fenestration may be detected deliberately or accidentally by imaging modalities such as 3D angiography. Such detection can even occur in post-mortem autopsies.

Relevant literature demonstrates that three-quarters of fenestrations occur, anatomically, in the posterior circulation, and the most involved artery are basilar and vertebral arteries (with prevalence varying between 0.3% to 28.0% in different articles and methods). Overall, fenestration, more commonly happens in intra-cranial vessels, and less frequently in the extracranial ICA. Among the intracranial vessels, the internal carotid artery fenestration is the least frequent. In articles reviewed to date, about 10 similar cases have been reported.

The patient represented in this study, presented with a chief complaint of transient blurred vision (amaurosis fugax) and was diagnosed with Transient Ischemic Accident (TIA). As her case was suspicious of dissection, she went under Digital Subtraction Angiography (DSA). Following the investigations, proximal internal carotid artery (ICA) fenestration, as well as dissection in the petrous part of the ICA were observed, which are both rare findings and in unusual locations. Also, proximal ICA was duplicated just after bifurcation, uniting back before the petrous part.

Besides reporting these unusual findings, we will review relevant literature about anatomical variants of fenestration. Paying special attention to angiographic findings, along with a good knowledge of rare findings such as ICA fenestration, can lead to timely and precise detection of such abnormalities.

Introduction

Vascular fenestration is a rare congenital variant, in which a segment of vessels divides into two distinct channels, covered by a layer of endothelium surrounding the muscularis tunica, that merge back together at the end of the affected portion [1-4].

Clinically, fenestration is usually asymptomatic or has little clinical significance. But sometimes fenestrations are accompanied by other structural abnormalities, such as intracerebral vascular aneurysms. These conditions may happen due to secondary defects in the media layer near the fenestration site. Also, turbulent flow resulting from defects in the proximal and distal ends of the fenestrated media, can lead to aneurysm formation [5]. The aneurysm may occur distal or proximal to the site of anomaly site.

Fenestration may be detected deliberately or accidentally by imaging modalities such as 3D angiography. Such detection can even occur in post-mortem autopsies.

From the anatomical point of view, multiple studies have examined the distribution of fenestration locations in cerebral arteries, with relatively similar results. A high-quality meta-analysis demonstrated that three-quarters of fenestrations occurred in the posterior circulation, and the most involved artery was the basilar artery [6].

In general, fenestration is more common in the intracranial vessels, and most cases of intracranial fenestration have been reported in the anterior cerebral artery, basilar artery, vertebral artery, and anterior communicating artery, varying each of them from 0.3% to 28.0% in different articles and methods [7-9].

Among the intracranial vessels, fenestration in the internal carotid artery is the least common with the extracranial portion having the lowest occurrence. In articles reviewed to date, about 10 similar cases have been reported.

The patient represented in this study, presented with a chief complaint of transient blurred vision (amaurosis fugax) and was diagnosed with Transient Ischemic Accident (TIA). As her case was suspicious of dissection, she went under Digital Subtraction Angiography (DSA). Following the investigations, proximal internal carotid artery (ICA) fenestration, as well as dissection in the petrous part of the ICA were observed, which are both rare findings and in unusual locations. Also, proximal ICA was duplicated just after bifurcation, uniting back before the petrous part.

Case presentation

A 45 years old woman presented to our clinic with transient blurred vision (amaurosis fugax) and left hemiparesis. The neurologic examination was unremarkable. Basic laboratory tests, along with an MRI were performed, with normal results (Figures 1 &2). Carotid Doppler Ultrasonography showed damping flow in the proximal part of left ICA. Brain MRA failed to show an ICA fenestration clearly.

As the patient was young, and stroke was improbable, cerebral DSA was performed to rule out a suspected dissection. DSA showed an ectopic area and dissecting pattern in the distal petrous area of ICA (Figures 3A-3B), which could justify patient's symptoms. Proximal ICA became duplicated just after bifurcation, and united back before the petrous segment.

The interesting point was that in the proximal ICA, a fenestration was observed, which has rarely been reported in the literature. Based on the findings above, the patient underwent standard medical treatment and became relieved of her symptoms.

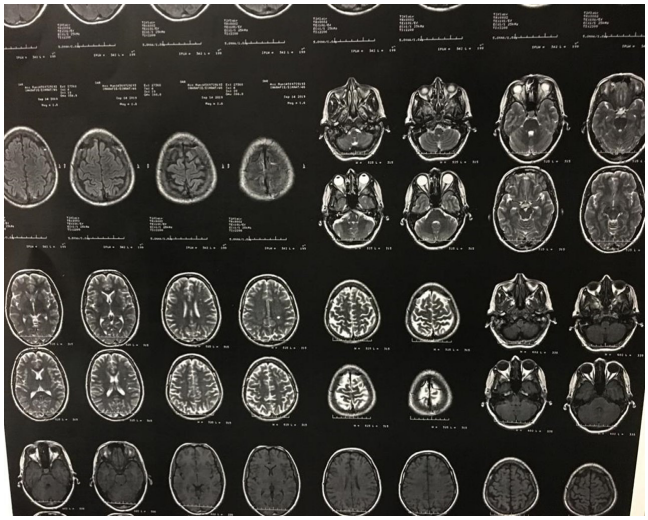

Figure 1: Normal MRI findings

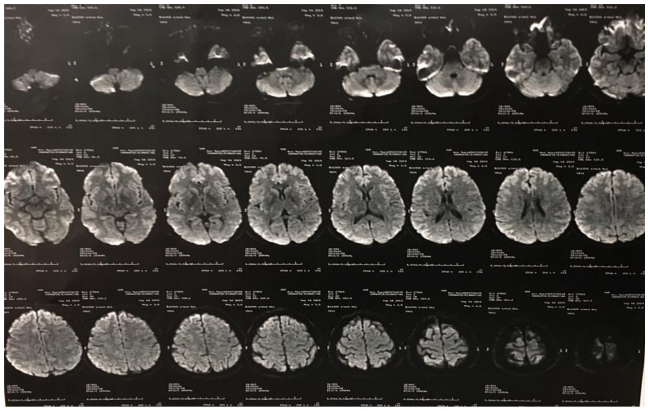

Figure 2: Normal MRI findings

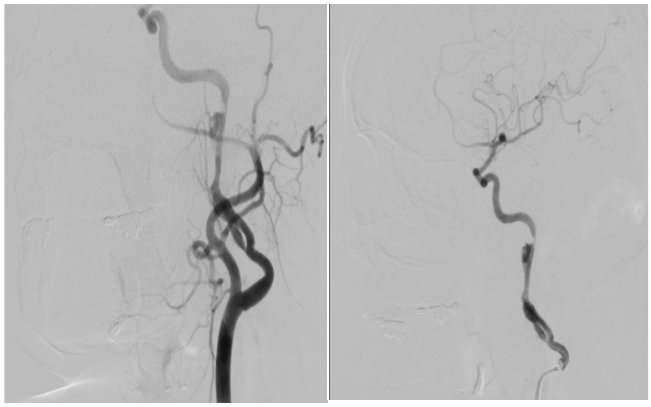

Figure 3A&B: Cerebral DSA: fenestration of the proximal ICA, and stenosis of the distal Petrus part with a dissecting pattern. Proximal ICA became duplicated just after bifurcation, and united back before the petrous segment.

Discussion

Cerebral artery fenestration is an anatomic variant, in which a segment of an artery is divided into at least two distinct channels, with their own endothelial and muscular layers [3,28,29].

Occasionally, fenestration is also mentioned as segmental duplication of the arteries. But it should be noted that in duplication, there are two distinct arteries of independent origin that remain separate in their course [9].

Fenestration, in general, is relatively rare and is caused by incomplete fusion of primary embryonic vessels. Depending on the degree of embryological fusion, the severity of this abnormality varies [1].

There are various opinions about carotid artery fenestration from the embryological point of view. For example, the fenestration may occurs due to aberrant fusion of the primitive carotid plexus, or when the cranial ductus caroticus persists, or an abnormal connection is formed between the third aortic arch and the dorsal aorta [10,11].

Different diagnostic methods have been used for detecting fenestrations, resulting in varied reported prevalence. Methods such as autopsy, surgical dissection, Digital Subtraction Angiography (DSA), Computed Tomography (CT), and Magnetic Resonance (MR) imaging have been used resulting in a prevalence of 0.7 to 60% [6,9,10,12-14,30]. This wide range may be due to small sample sizes, selection biases, retrospective design of the studies, and different sensitivity of detection techniques. All in all, detection rate of fenestration, has improved significantly using 3D rotational angiography [10,31].

In a retrospective 2013 study, about 11,000 patients who underwent digital subtraction angiography between 1992 and 2011 were studied. Of these, 228 unique cases with fenestration were found. Therefore, this study reported a prevalence of fenestration of 2.1%, which was 0.7% higher than that of DSA, and also lower than other modalities such as MR (2.8 - 3.0%), DSA (28.9%-22.9%), and CT (3.5-12.9%) and specially much less than those reported from the autopsy series [15-17,32].

Different medical centers have reported a different prevalence of fenestration in various cerebral vessels, most of whom show the lowest incidence of fenestration in the internal carotid artery. For example, in the previously mentioned study, three-quarters (73.2%) of all fenestrations occurred in the posterior circulation, with the Basilar artery being the most common location (52.6%). Following that, were other arteries such as the middle and posterior cerebral arteries (4.4% and 0.4%), with the internal carotid artery being the least common site [18].

Fenestration may be detected accidentally, or may have low clinical significance. On the other hand, it may be associated with other complications, such as aneurysms, arterial-venous malformations, ischemic strokes, dissections, dural fistula, and other cerebrovascular anomalies [19, 20]. These complications may occur due to changes in microvascular flow dynamics (14). Thinning or lack of media layer is also observed around the fenestration site. Therefore, some believe that fenestration is not a benign finding. Thus, multiple studies have investigated concomitant incidence of aneurysms in association with fenestrations, with results ranging from 2.5 to 17% [8,13].

In a study by van Rooij et al. in 2015, 179 patients with subarachnoid hemorrhage who had been hospitalized for two years were studied. Of these, 140 patients underwent 3D cerebral angiography, resulting in finding a total of 210 aneurysms. There were also 45 fenestrations, most of which were distributed in the anterior communicating artery (69%), anterior cerebral artery (9%), middle cerebral artery (9%), and basilar artery (4%). Fenestration was not reported in the carotid arteries. Of the 210 aneurysms, 14 were located at the site of fenestration. The study results showed a 24% prevalence of intracranial fenestrations in patients with suspected rupture of aneurysms. This study had the limitation of investigating exclusively patients with suspected subarachnoid hemorrhage, in whose population the rate of the aneurysms is higher than the general population [21].

There are reports of fenestration in various places, such as the distal ICA. But the novelty of this report is that this fenestration occurred in the cervical carotid region of the ICA, which is very rare in terms of location and very few cases have been reported. Therefore, it has gained scientific importance for reporting.

Previously, two cases of internal carotid artery fenestration were reported by Hasegawa T et al [22].

One was of a 47-year-old woman with a saccular aneurysm in the right middle cerebral artery, who was found to have a fenestration of the right internal carotid artery at the site of the atlas. The Second case was of a 51-year-old man hospitalized for grand mal seizures, which was found to be due to a malignant glioma in the right temporal lobe. A fenestration of the right internal carotid artery in the atlantoaxial joint was also detected, with the vessel dilated in diameter at the level of C1-C2.

As recently as 2019, a study by Uchino A et al reported a patient with unilateral hypoplasia of the internal carotid artery. The findings of this patient were associated with ipsilateral anomalous posterior cerebral artery, and fenestrated P1 segment. This article suggested that partial maximum-intensity-projection imaging helps identify rare arterial variations on MR angiography [23].

Another study in 2019 by Shiozaki E et al. reported a rare case of fenestration in the cavernous segment of the left internal carotid artery, with an aneurysm proximal to the fenestrated portion. The case in study was a 44-year-old woman with a dull headache, who underwent CT angiography and 3D-rotational angiography; no aneurysms were found in the right ICA, but an intra-cavernous fenestration and aneurysm were found in the left ICA. The aneurysm was small (5 X 3 mm), with a wide neck arising from the origin of the ventral limb of the duplicated vessels. According to this study, only three cases of this type of ICA fenestrations had been reported by 2019, and this was also the first report of an ICA fenestration with an unbroken aneurysm at the fenestrated segment [24].

Other studies have addressed the supra-clinoid ICA fenestration (SIF). This is important because it can mimic an intracranial aneurysm, an intraluminal thrombus, or a focal dissection on non-invasive imaging. For example Sgreccia A et al (2018) reported a 55-year-old patient with a right SIF and a wide-necked aneurysm, located on the limb of the fenestration. Also a second small aneurysm was found, distal to fenestration site [25].

Further, Ga Y Lee et al (2018) reported a rare case of fenestration in the right supra-clinoid internal carotid artery. Their patient was a 65-year-old woman with no particular underlying disorder, who had been complaining of headaches and dizziness for a month. DSA was performed on the patient, suggest-

ing a fenestration of the supra-clinoid ICA, combined with a para-clinoid aneurysm [26].

Also, Orru E et al. in 2015, reported a concomitant arteriovenous malformation (AVM) distal to the fenestration site. They presented a 26-year-old woman with a grade I right frontal AVM, in whom a left supra-clinoid ICA fenestration was detected incidentally via cerebral angiography [27].

These reports were the majority of relevant internal carotid fenestration cases, which overall are rare. In our case, specially, fenestration occurs in the proximal site of the internal carotid, which is in an unusual anatomical location, and is associated with dissection.

References

1. Dimmick SJ, Faulder KC. Normal variants of the cerebral circulation at multidetector CT angiography. Radiographics: A review publication of the Radiological Society of North America, Inc. 2009; 29(4): 1027-43.
2. Uchino A, Takase Y, Nomiyama K, Egashira R, Kudo S. Fenestration of the middle cerebral artery detected by MR angiography. Magnetic resonance in medical sciences: MRMS: An official journal of Japan Society of Magnetic Resonance in Medicine. 2006; 5(1): 51-5.
3. Liu H, Hong J, Wang S, Wei L. Anterior communicating artery complex fenestration combined with tandem aneurysm: a case report and literature review. Medicine. 2020; 99(19).
4. Maroufi SF, Rafiee Alavi SN, Abbasi MH, Famouri A, Mahya n, Armaghan S, et al. Comparison of Doppler Ultrasound and Digital Subtraction Angiography in extracranial stenosis. Annals of Medicine and Surgery. 2022; 74: 103202.
5. Sanders WP, Sorek PA, Mehta BA. Fenestration of intracranial arteries with special attention to associated aneurysms and other anomalies. AJNR American journal of neuroradiology. 1993; 14(3): 675-80.
6. Cooke DL, Stout CE, Kim WT, Kansagra AP, Yu JP, Gu A, et al. Cerebral arterial fenestrations. Interventional neuroradiology: journal of peritherapeutic neuroradiology, surgical procedures and related neurosciences. 2014; 20(3): 261-74.
7. Vasović L, Trandafilović M, Jovanović I, Ugrenović S, Antović A, Karadžić R, et al. Human basilar artery abnormalities in the prenatal and postnatal period. World neurosurgery. 2013; 79(3-4): 593.e15-23.
8. van Rooij SB, van Rooij WJ, Sluzewski M, Sprengers ME. Fenestrations of intracranial arteries detected with 3D rotational angiography. AJNR American journal of neuroradiology. 2009; 30(7): 1347-50.
9. Menshawi K, Mohr JP, Gutierrez J. A Functional Perspective on the Embryology and Anatomy of the Cerebral Blood Supply. Journal of stroke. 2015; 17(2): 144-58.
19. Parmar H, Sitoh YY, Hui F. Normal variants of the intracranial circulation demonstrated by MR angiography at 3T. European journal of radiology. 2005; 56(2): 220-8.
11. Priya S, Thomas R, Nagpal P, Sharma A, Steigner M. Congenital anomalies of the aortic arch. Cardiovascular diagnosis and therapy. 2018; 8: S26-s44.
12. Dey M, Awad IA. Fenestration of supraclinoid internal carotid artery and associated aneurysm: embryogenesis, recognition, and management. World neurosurgery. 2011;76(6):592.e1-5.
13. Bozek P, Pilch-Kowalczyk J, Kluczewska E, Zymon-Zagorska A. Detection of cerebral artery fenestrations by computed tomography angiography. Neurologia i neurochirurgia polska. 2012; 46(3):239-44.
14. Mirzaasgari Z, Haghi-Ashtiani B, Refaiean F, Vahedifard F, Homayooni AS, Sobhkhiz M. Diagnostic value of high-frequency ultrasound in carpal tunnel syndrome during pregnancy: A case-control study: Tehran University of Medical Sciences. 2021. 73-7 p.
15. Arraez-Aybar LA, Villar-Martin A, Poyatos-Ruiperez C, Rodriguez-Boto G, Arrazola-Garcia J. Prevalence of fenestrated basilar artery with magnetic resonance angiography: a transversal study. Surgical and radiologic anatomy : SRA. 2013; 35(6):487-93.
16. Bayrak AH, Senturk S, Akay HO, Ozmen CA, Bukte Y, Nazaroglu H. The frequency of intracranial arterial fenestrations: a study with 64-detector CT-angiography. European journal of radiology. 2011; 77(3): 392-6.
17. Dimmick SJ, Faulder KC. Fenestrated anterior cerebral artery with associated arterial anomalies. Case reports and literature review. Interventional neuroradiology: journal of peritherapeutic neuroradiology, surgical procedures and related neurosciences. 2008; 14(4): 441-5.
18. Sun ZK, Li M, Li MH, Li YD, Sun WP, Zhu YQ. Fenestrations accompanied by intracranial aneurysms assessed with magnetic resonance angiography. Neurology India. 2012; 60(1): 45-9.
19. Hudak I, Lenzser G, Lunenkova V, Doczi T. Cerebral arterial fenestrations: a common phenomenon in unexplained subarachnoid haemorrhage. Acta neurochirurgica. 2013; 155: 217-22.
20. Esmaeili S, Mirzaasgari Z, Vahedifard F, editors. Optic Nerve Sheath Ultrasound Predicts The Severity Of Acute Ischemic Stroke. International Journal Of Stroke; 2020: Sage Publications Ltd 1 Olivers Yard, 55 City Road, London Ec1y 1sp, England.
21. van Rooij SBT, Bechan RS, Peluso JP, Sluzewski M, van Rooij WJ. Fenestrations of Intracranial Arteries. American Journal of Neuroradiology. 2015; 36(6): 1167-70.
22. Hasegawa T, Kashihara K, Ito H, Yamamoto S. Fenestration of the internal carotid artery. Surgical Neurology. 1985; 23(4): 391-5.
23. Uchino A, Ehara T, Kurita H. Hypoplasia of the internal carotid artery with associated fenestration and extremely long P1 segment of the ipsilateral posterior cerebral artery diagnosed by MR angiography. Surgical and radiologic anatomy : SRA. 2019; 41(6): 707-11.
24. Shiozaki E, Hiu T, Chikamatsu G, Nakamura H, Ozono K, Honda K, et al. [A Rare Case of an Unruptured Aneurysm Arising from the Proximal End of the Fenestration of the Cavernous Segment of the Internal Carotid Artery]. No shinkei geka Neurological surgery. 2019; 47(2): 217-23.
25. Sgreccia A, Coskun O, Di Maria F, Rodesch G, Consoli A. Fenestration of the supraclinoid segment of the ICA and associated aneurysms: a case report with literature review. Acta neurochirurgica. 2018; 160: 1143-7.
26. Lee GY, Shin GW, Jung HS, Baek JW, Heo YJ, Lee YJ, et al. Fenestration of the supraclinoid internal carotid artery connecting the neck of the paraclinoid aneurysm and the origin of the posterior communicating artery: A case report. Interventional neuroradiology: journal of peritherapeutic neuroradiology, surgical procedures and related neurosciences. 2018; 24(3): 274-6.
27. Orru E, Wyse E, Pearl MS. Fenestration of the supraclinoid internal carotid artery in a patient with a concomitant intracranial arteriovenous malformation. BMJ case reports. 2015; 2015.

28. Esmaeili S, Abbasi F, hossein Abbasi M, Eshraghi A, Mirzaasgari Z. Impact of pre-stroke metformin use on prognosis of Ischemic stroke; A pilot study. Journal of Bioengineering Research. 2019; 1(4): 45-9.

29. Acton EK, Abbasi MH, Kasner SE. Evaluating Age, Sex, Racial, and Ethnic Representation in Acute Ischemic Stroke Trials, 2010 to 2020: A Systematic Review and Meta‐ Analysis. Journal of the American Heart Association. 2022; 11(8): e024651.

30. Raeesmohammadi L, Esmaeili S, Abbasi MH, Mehrpour M, Mirzaasgari Z, Baradaran HR, et al. Transbulbar B-mode sonography in multiple sclerosis without optic neuritis; clinical relevance. Brain Research. 2020; 1734: 146723.

31. Vahedifard F, Sadeghniiat Haghighi, A. The role of Neuroradiology in Neuroplasticity: New advancements. World Journal of Advanced Research and Reviews. 2022; 14(02): 156-60.

32. Esmaeili S, Mirzaasgari Z, Vahedifard F. Optic Nerve Sheath Ultrasound Predicts The Severity Of Acute Ischemic Stroke. Ininternational Journal Of Stroke 2020 Nov 1 (Vol. 15, No. 1_ Suppl, Pp. 310-310). 1 Olivers Yard, 55 City Road, London Ec1y 1sp, England: Sage Publications Ltd.