

## CASE REPORT



## Fenestration of the cervical internal carotid artery misdiagnosed as dissection

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### Abstract

Internal carotid artery (ICA) anatomical variations are relatively rare occurrences during diagnostic imaging procedures. Their presence can have important prognostic consequences in the evaluation of vascular neurological diseases. It is therefore important to have a good knowledge about these variations, in order to avoid unwarranted medical interventions. We present the case of a patient harboring a right ICA fenestration in the cervical segment, misdiagnosed as a dissection on computed tomography angiography, admitted in the Department of Neurology and treated accordingly. The possible pathological and embryological origins of arterial fenestrations are discussed, and a brief review of the literature related to ICA fenestrations is presented.

**Keywords:** internal carotid artery, arterial fenestration, dissection, fibromuscular dysplasia.

### Introduction

Internal carotid artery (ICA) fenestrations are extremely rare anatomic variants, to the best of our knowledge only 12 cases [1–6] being reported in the literature so far. Fenestrations can be mistaken for dissections, pseudo-aneurysms or floating thrombi on imaging studies [7], therefore recognition of their presence is essential, in order to avoid unnecessary medical interventions.

### Aim

We present the case of a patient harboring a right ICA fenestration in the cervical segment, misdiagnosed as ICA dissection on a computed tomography angiography (CTA), admitted in the Department of Neurology and treated accordingly.

### Case presentation

A 54-year-old female patient, known to have a chronic maxillary and ethmoid sinusitis, presented in the Department of Otorhinolaryngology complaining of chronic hemicrania for two months, exacerbated three days ago. Head magnetic resonance (MR) examination showed no imaging signs of sinusitis, or other intracranial pathological changes (not shown). In order to rule out a vascular disease, a CTA of the head and neck was performed on a Siemens (Siemens, Erlangen, Germany) Somatom Definition 64-channel scanner: a fusiform dilatation with a diameter of 5.6 mm, in which a hypodense linear filling defect with a length of 8.5 mm was discovered in the distal part of the right cervical segment of the ICA (Figure 1, a and b), at the level of the first cervical vertebra. Concomitantly,

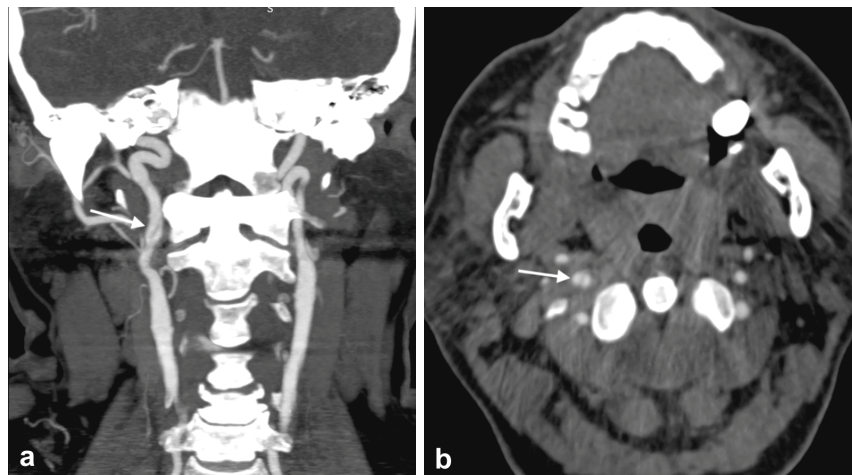
both ICA's had marked tortuosities and wall irregularities were evident on the left vertebral artery, suggestive of fibromuscular dysplasia (FMD) (Figures 1 and 2). Clinical and otorhinolaryngological examinations were normal with a heart rate 68 beats/minute and blood pressure of 110/65 mmHg. Neurological exam revealed a conscious patient, without meningeal signs, cranial nerve deficits, or other focal deficits, preserved bilateral osteotendinous reflexes and coordination. Laboratory findings were within normal limits: leukocyte count 5810/mm<sup>3</sup>, hemoglobin 13.8 g/dL, platelet count 297 000/mm<sup>3</sup> and erythrocyte sedimentation rate of 7 mm/h.

Based on the symptoms and imaging findings, a diagnosis of cervical ICA dissection was established, the patient was admitted in the Department of Neurology and started on double antiplatelet therapy, with 75 mg Aspirin and 75 mg Clopidogrel daily for two weeks. During the hospitalization, the clinical status of the patient remained unchanged and the headache decreased in intensity.

For a better assessment of the dissected segment and its hemodynamic impact, catheter angiography was performed after two weeks on a Siemens (Siemens, Erlangen, Germany) Artis Zee biplane system: digital subtraction angiography (DSA) and three-dimensional (3D) rotational angiography images were obtained (Figure 2, a–c). Findings were similar to the CTA: marked tortuosities of both ICA's, vertebral artery FMD and a short filling defect in the right cervical ICA, which was difficult to depict with DSA images alone, due to the narrow “gap” in the artery. This limitation was easily overcome with the acquisition of 3D reconstructed rotational angiography images, where the “gap” in the artery became obvious (Figure 2, a–c). The angiographic findings were characteristic for an

arterial fenestration, consequently the diagnosis was changed, and the antiplatelet therapy was stopped. The

patient was discharged with symptomatic treatment for her headache.



**Figure 1 – (a) Coronal MIP CTA image: linear hypodense filling defect in the distal cervical ICA, at the level of the first cervical vertebra; (b) Axial CTA image: filling defect “splitting” the vascular lumen in two channels. MIP: Maximum intensity projection; CTA: Computed tomography angiography; ICA: Internal carotid artery.**



**Figure 2 – (a) Oblique DSA of the ICA shows the “slit”-like defect in the artery; (b) 3D reconstructed rotational angiography depicts the fenestration; (c) AP DSA image of the left vertebral artery shows irregularities of the V3 segment, with ectasia and stenosis, consistent with the diagnosis of FMD (white arrows). DSA: Digital subtraction angiography; ICA: Internal carotid artery; 3D: Three-dimensional; AP: Anteroposterior; FMD: Fibromuscular dysplasia.**

## Discussions

Arterial fenestrations are short divisions of the vascular lumen in two vascular channels, each with its own tunica intima, media and adventitia, although a shared adventitia is possible [8]. They are relatively common incidental, asymptomatic variants in the intracranial arterial system, most frequently encountered at the level of the anterior cerebral-communicating artery complex. Other frequent sites of occurrence include the middle cerebral artery and the vertebrobasilar system [9].

The ICA seems to be the rarest artery supplying the brain affected by this variant, with only 12 cases reported in the literature so far [1–6] (Table 1). The underlying mechanisms that can lead to the development of fenestrations are incompletely understood, although some theories were proposed.

During the early stages of embryonic growth, the ICA's develop through fusion of a plexiform network of arterial channels into a single vessel in the 4–5 mm crown-rump length embryo [10]. A merging defect during

this process could lead to an unfused vascular segment, similar to the well-recognized fenestrations in the vertebrobasilar arterial territory, where multiple longitudinal neural arteries coalesce into the adult arterial system. This theory is reinforced by the existence of fenestrations located on other ICA segments, like supraclinoid [10] or intracavernous [11]. Other possible embryological explanations are the persistence of the carotid duct, a vascular structure that connects the 3<sup>rd</sup> and 4<sup>th</sup> aortic arches, until the 12 mm crown-rump length embryo, when it normally regresses [5].

One interesting observation is that the majority of ICA fenestrations encountered so far are located in the distal cervical segment (Table 1), as is our own case. This location is compatible with the origin of hypoglossal artery or type 1 proatlantal artery, two primitive anastomoses between the ICA and the vertebral arteries present in the embryo. They regress early in its development; therefore, one can assume that defects during the embryonic period are good theoretical candidates as to the origin of ICA fenestrations.

**Table 1 – Review of cases with fenestrations of the cervical ICA published in the literature so far**

No. of patient	Authors (year) [Ref. No.]	Cases	Age [years]	Gender	Location	Associated anomalies
1.	Tanaka & Matsumoto (1982) [1]		58	M	Distal	Intracranial aneurysm
2.	Hasegawa <i>et al.</i> (1985) [2]	Case No. 1	47	F	Distal	Intracranial aneurysm
3.		Case No. 2	51	M	Distal	Glioblastoma
4.	Nakamura <i>et al.</i> (1993) [3]		73	M	Proximal	n/a
5.	Ahn <i>et al.</i> (2003) [4]		49	F	Distal	Fusiform dilatation of a limb
6.	Gailloud <i>et al.</i> (2004) [5]	Case No. 1	59	F	Distal	n/a
7.		Case No. 2	52	M	Distal	DVA
8.		Case No. 3	51	F	Proximal	Intracranial aneurysm, FMD
9.		Case No. 4	74	M	Distal	FMD
10.		Case No. 5	41	F	Distal	FMD
11.		Case No. 6	46	F	Distal	n/a
12.	Liang <i>et al.</i> (2016) [6]		60	M	Distal	none
13.	Our case		54	F	Distal	FMD

ICA: Internal carotid artery; M: Male; F: Female; DVA: Developmental venous anomaly; FMD: Fibromuscular dysplasia; n/a: Not available.

A different approach to the origin of ICA fenestrations was proposed by Gailloud *et al.* [5], who described six cases with short ICA fenestrations, suggesting that they are a consequence of dissections. It is our opinion that, with the exception of Cases Nos. 5 and 6, the first four are very similar to the rest of cases reported in other papers, also similar to ours, we therefore consider those cases as fenestrations, not post-dissection changes. Moreover, the fact that three of them had associated signs of FMD seems to point to a generalized tendency to vascular anomalies.

One important finding in our review is the association of ICA fenestration with other vascular pathologies (Table 1). Three patients have intracranial aneurysms [1, 2, 5], three patients from Gailloud *et al.* paper (Cases Nos. 3, 4 and 5) and our own have FMD. These findings highlight a possible underlying congenital vascular vulnerability. In this context, it is mandatory to carefully evaluate vascular imaging studies of the head and neck, in order to exclude devastating pathologies like intracranial aneurysms.

ICA fenestrations can mimic serious vascular disorders: dissections, pseudo-dissections, floating thrombi or pseudo-aneurysms [5, 7, 12]; not recognizing their appearance on imaging studies can lead to unwarranted treatments with potential harm to the patient. Our patient complained of acute onset headache with no neurological deficits at clinical examination. MR imaging study was normal, prompting a vascular imaging study to exclude pathologies like dissections or aneurysms. The CTA appearance was mistaken for a dissection and the patient was started on dual antiplatelet therapy for secondary ischemic stroke prevention. Besides the increased risk of hemorrhagic complications associated with dual antiplatelet therapy [13], there are also additional costs related to hospitalization and further imaging studies for the surveillance of the “false disease”. It is crucial for physicians who take care of patients with carotid artery disease, to be aware of this extremely rare vascular anomaly.

Imaging findings in ICA fenestrations are characteristic. The fenestrated arterial segment is usually short and slightly dilated compared to the rest of the ICA. Inside this fusiform dilation, a linear filling defect “splitting” the vascular lumen can be well seen on axial CTA or MR

angiography images. The fenestration extends over a short distance of a few millimeters, as opposed to dissections, which usually involve longer segments of the artery. Although fenestrations have typical features on CTA images, sometimes catheter angiography is helpful in the differential diagnosis. DSA images in multiple projections can depict the two separate channels and the “gap” in the artery and with the help of 3D rotational reconstructed images, the diagnosis becomes certain.

## Conclusions

ICA fenestrations are extremely rare anatomical variants that can mimic serious vascular diseases like dissections, floating thrombi and pseudoaneurysms. Their recognition on imaging studies is essential in order to prevent unnecessary medical interventions. Fenestrations seem to be associated with other vascular disorders, *e.g.*, FMD and intracranial aneurysms; it is therefore extremely important to pay careful attention to the cerebral vasculature when looking at imaging studies of patients with this rare variant.

## Conflict of interests

The authors declare that they have no conflict of interests.

## Informed consent

The Institutional Ethics Committee approved the publication of this case report and waived the requirement to obtain any informed consent.

## References

- [1] Tanaka M, Matsumoto S. [Fenestration of the internal carotid artery]. *Neurol Med Chir (Tokyo)*, 1982, 22(4):291–294. <https://doi.org/10.2176/nmc.22.291> PMID: 6179001
- [2] Hasegawa T, Kashiwara K, Ito H, Yamamoto S. Fenestration of the internal carotid artery. *Surg Neurol*, 1985, 23(4):391–395. [https://doi.org/10.1016/0090-3019\(85\)90214-9](https://doi.org/10.1016/0090-3019(85)90214-9) PMID: 3975829
- [3] Nakamura H, Yamada H, Nagao T, Fujita K, Tamaki N. Fenestration of the internal carotid artery associated with an ischemic attack – case report. *Neurol Med Chir (Tokyo)*, 1993, 33(5):306–308. <https://doi.org/10.2176/nmc.33.306> PMID: 7687037
- [4] Ahn JY, Kim OJ, Joo YJ, Joo JY. Fenestration of the internal carotid artery associated with arterial dissections. *J Clin Neurosci*, 2003, 10(2):257–260. [https://doi.org/10.1016/s0967-5868\(02\)00331-4](https://doi.org/10.1016/s0967-5868(02)00331-4) PMID: 12637067

- [5] Gailloud P, Carpenter J, Heck DV, Murphy KJ. Pseudo-fenestration of the cervical internal carotid artery: a pathologic process that simulates an anatomic variant. *AJNR Am J Neuroradiol*, 2004, 25(3):421–424. PMID: 15037466
- [6] Liang Z, Xiu C, Liu Z, Sun X, Yu G, Tao M, Li W. Misdiagnosis of a patient with internal carotid artery fenestration: a case report and literature review. *Ann Vasc Surg*, 2016, 30:309.e1–309.e4. <https://doi.org/10.1016/j.avsg.2015.07.031> PMID: 26522583
- [7] Tourdias T, Berge J, Menegon P, Sibon I. Fenestration of the internal carotid artery mimicking floating thrombus on CT and MR angiography. *Neurology*, 2011, 76(21):1846. <https://doi.org/10.1212/WNL.0b013e31821ccd28> PMID: 21606460
- [8] Hakim A, Gralla J, Rozeik C, Mordasini P, Leidolt L, Piecho-wiak E, Ozdoba C, El-Koussy M. Anomalies and normal variants of the cerebral arterial supply: a comprehensive pictorial review with a proposed workflow for classification and significance. *J Neuroimaging*, 2018, 28(1):14–35. <https://doi.org/10.1111/jon.12475> PMID: 28945289
- [9] van Rooij SBT, van Rooij WJ, Sluzewski M, Sprengers MES. Fenestrations of intracranial arteries detected with 3D rotational angiography. *AJNR Am J Neuroradiol*, 2009, 30(7):1347–1350. <https://doi.org/10.3174/ajnr.A1563> PMID: 19439481
- [10] Ichikawa T, Miyachi S, Izumi T, Matsubara N, Naito T, Haraguchi K, Wakabayashi T, Koketsu N. Fenestration of a supraclinoid internal carotid artery associated with dual aneurysms: case report. *Neurosurgery*, 2011, 69(4):E1005–E1008; discussion E1009. <https://doi.org/10.1227/NEU.0b013e318223b613> PMID: 21572363
- [11] Uchino A, Nomiyama K, Takase Y, Kohata T, Kudo S. Intracavernous fenestration of the internal carotid artery. *Eur Radiol*, 2006, 16(7):1623–1624. <https://doi.org/10.1007/s00330-005-0104-2> PMID: 16408202
- [12] Filep RC, Bajkó Z, Simu IP, Stoian A. Pseudo-dissection of the internal carotid artery in acute ischemic stroke. *Acta Neurol Belg*, 2020, 120(2):469–472. <https://doi.org/10.1007/s13760-018-1025-6> PMID: 30251236
- [13] Algra A, de Schryver EL, van Gijn J, Kappelle LJ, Koudstaal PJ. Oral anticoagulants *versus* antiplatelet therapy for preventing further vascular events after transient ischaemic attack or minor stroke of presumed arterial origin. *Cochrane Database Syst Rev*, 2006, (3):CD001342. <https://doi.org/10.1002/14651858.CD001342.pub2> PMID: 16855967

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