A1-A2 anterior cerebral artery fenestration. A case report of a rare anatomical variant

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SUMMARY

Cerebral vascular anatomical variations are not uncommon in the human population. Their prevalence is not exactly known, as most of them are incidentally diagnosed on angiography or postmortem dissections. We present a rare fenestration of the A1 segment of the anterior cerebral artery presenting with a ruptured saccular aneurysm.

A 42-year-old hypertensive patient presented unconscious following a ruptured saccular aneurysm. The computer tomography angiography showed a fenestration of the right A1 segment of the anterior cerebral artery (ACA). The medial segment of the A1 was communicating with the left ACA via the Anterior communicating artery, while the lateral segment was directly joining the A2 segment of the same side. Intraoperatively, the two segments were identified as separate vascular structures not sharing adventitia, and of equal caliber. The aneurysm arising from the bifurcation was clipped. The patient recovered with no neurological deficits. Many vascular anomalies like fenestrations and bifurcations are underdiagnosed, as many of them remain asymptomatic and are discovered incidentally on postmortem dissection or angiography for other pathologies. This has led to a paucity of cases to determine the prevalence in the human population. Good knowledge of the vascular anatomy variations and associated risk of aneurysm is important for the vascular neurosurgeons.

Keywords: Anterior cerebral artery – Fenestration – Bifurcation – Anatomical variants – Vascular anatomy

INTRODUCTION

Many different vascular anomalies have been described in the brain, some of which can be found in the arteries that form the circle of Willis and are related to congenital disorders (Enyedi et al., 2021; Dumitrescu et al., 2021). One of these rare variants is fenestration, or partial duplication.

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Submitted: May 8, 2022. **Accepted:** June 14, 2022

https://doi.org/10.52083/LXNM1800

Fenestrations, according to the literature, are more frequent in the posterior circulation (Nyasa et al., 2021; Tanaka, 2017). They are usually silent and asymptomatic, and as a result are most commonly discovered incidentally on cadaver dissections and angiography for other pathologies (Fredon et al., 2021; Krystkiewicz et al., 2021). Fenestration, or partial duplication of the anterior cerebral artery, is almost always accompanied by the presence of an aneurysm. Fenestrations have been described in the A1 segment of the anterior cerebral artery, although the most frequently affected segment of the anterior circulation is the Anterior communicating artery (Acom) (Mamadaliev et al., 2019; Trandafilovic et al., 2021). As with all other vascular disorders, cerebral angiography is invaluable in their accurate diagnosis (Mahajan et al., 2020).

The incidence of arterial fenestrations in the brain ranges from 0.3% to 0.9% in angiography and 0.14% in cadaver dissections, according to the literature (Guo et al., 2018; Iqbal, 2013). Its prevalence in the A1 region is 0-4%, as described in anatomical studies, and 0.058%, as described in angiographic studies (Makowicz et al., 2013). The distal A1 segment is described as the most frequently associated with fenestrations in dissections of cadavers. Various theories have been reported to explain the association between aneurysms and fenestrations, including: Vascular wall weakness (Iwabuchi et al., 2018), and failure of fusion of the plexiform multi-channel network of vessels that develop into major arteries during fetal life (Krystkiewicz et al., 2021; Kwon et al., 2013; Mahajan et al., 2020; Makowicz et al., 2013).

In this article, we present a case of a rare A1 fenestration terminating in the A2 segment with a proximal saccular aneurysm. To the best of our knowledge, this type of fenestration has been described once in a cadaver dissection without an associated aneurysm. The case report below is presented according to the CARE checklist.

CASE DESCRIPTION

A 42-year-old female patient presented to the emergency department with sudden loss of consciousness following a severe headache. She was a known hypertensive on medication, with a history of recurrent headaches. On examination, her GCS was 9, BP 165/100mmHg. She had left hemiparesis. An urgent CT showed intracerebral and intraventricular hemorrhage with no midline shift (Fig. 1A). CTA showed a fenestrated left A1 with a saccular aneurysm at the proximal end of the fenestration (Fig. 1B).

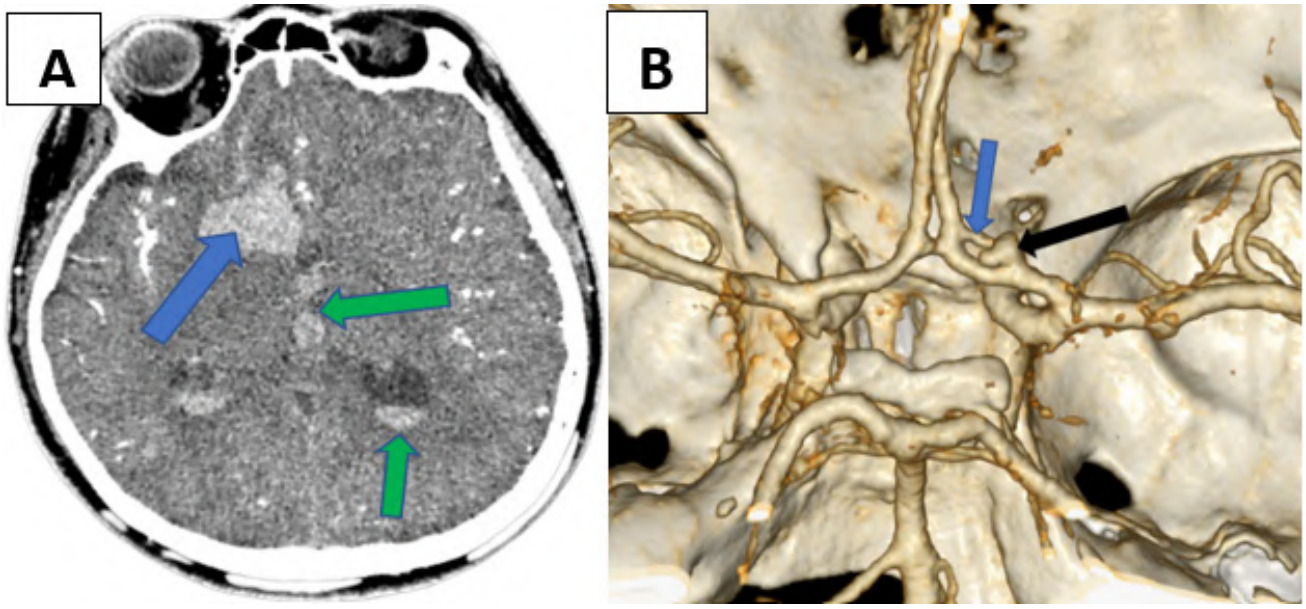

Fig. 1.- A- Preoperative CT brain showing a right frontal intracerebral hematoma (blue arrow) and Fischer grade 4 subarachnoid hemorrhage. **B**- Preoperative CTA shows a fenestrated right A1 connecting to the right A2 (blue arrow) with a saccular aneurysm (black arrow) arising from the origin of the fenestration.

The patient was stabilized in the intensive care unit for 48 hours. On the second day post admission, she underwent aneurysm clipping. Intraoperatively, the partial duplication of the right A1 was visualized (Fig. 2A) with an aneurysm arising at the origin of the partial duplication (Fig. 2B).

The aneurysm was successfully clipped with no intraoperative complications (Fig. 2C).

The post-operative period was uneventful, and the patient was discharged on the fourteenth day post-operative with no neurological deficit. At 3-month follow-up, the patient remained asymptomatic without any restriction to daily activities.

There were no diagnostic challenges faced in this patient, as the neuroimaging performed was adequate.

Endovascular services at the time the patient presented were not available at the institution. As a result, an endovascular approach was not an option.

DISCUSSION

Thomas Willis in 1664 described the complete cerebral vascular circuit, the so-called circle of Willis, and described it as a network of arterial vascular anastomosis, located at the base of the skull (Nyasa et al., 2021; Dumitrescu et al., 2021).

The cerebral arterial circulation is divided into an anterior and posterior system. The anterior circulation includes the internal carotid arteries, which give rise to the anterior cerebral arteries, which are connected via the anterior communicating artery, and the middle cerebral arteries. The posterior circulation includes the paired vertebral arteries, which merge giving rise to the basilar artery. The basilar artery gives rise to the anteroinferior and superior cerebellar arteries, and the posterior cerebral arteries (Tanaka, 2017). The posteroinferior cerebellar arteries arise directly from the vertebral arteries before they form the basilar artery. The anterior and posterior systems communicate through the posterior communicating arteries (Makowicz et al., 2013).

The anterior cerebral artery (ACA) presents both clinically significant and incidental variations in its anatomy. Previous studies have found a significant difference in the prevalence of these anatomical variants amongst different ethnic groups (Jiménez Sosa et al., 2017).

A study by Makowicz et al. (2013) revealed that the presence of these atypical variants of cerebral vessels is associated with an increased risk of ischemic event and/or aneurysm formation. The typical complete variant of ACA is seen in only 30-57 % of the population (López-Sala et al., 2020).

Almost all the variants of ACA and its branches have been described in the literature. Several authors have classified variants of ACA and its branches: A1, Acom, and A2 (Jiménez Sosa et al., 2017; Kayembe et al., 1984). Some of the described variants are illustrated in Fig. 3. We have added

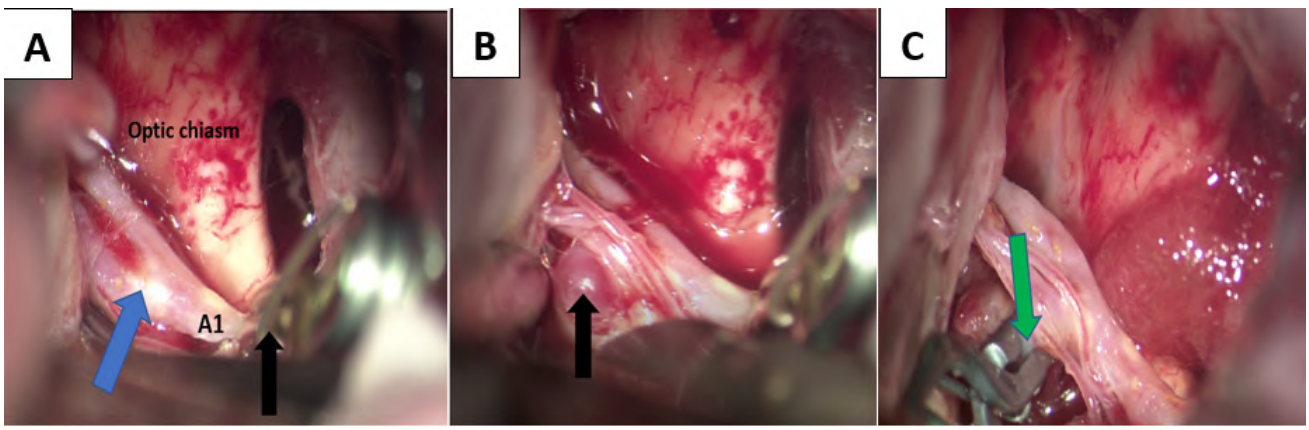

Fig. 2.- A- Intraoperative exposure of the A1 segment after placement of temporary clip (black arrow). Notice the abnormal bifurcation of A1 (blue arrow). **B**- The saccular aneurysm (black arrow) seen arising posteriorly at the A bifurcation. **C**- Shows the view after aneurysm clipping with the 2 branches of A1 clearly visible.

the variant described in this case report: i.e., A1-A2 fenestration.

The variations seen in the ACA include: reduction in caliber (Hypoplasia), complete absence (aplasia), the same artery with different and independent origin (Duplication, triplication), and fenestration, which occurs when vessels duplicate forming two channels corresponding to a single path, with each channel having its own endothelium and tunica media but possibly sharing the tunica adventitia and, most importantly, reuniting distally (López-Sala et al., 2020). Fenestration is also referred to as partial duplication (Guo et al., 2018).

A duplication occurs with two distinct arteries with separate origin that do not converge distally. It is most often seen in MCA variants. Duplication of A1 segment of ACA occurs in up to 4% of normal subjects in cadaveric studies (Perlmutter and Rhoton, 1976). ACA Trifurcation is the occurrence of 3 segments and prevalence in A2 with the third branch arising from Acom in 2-13% of cases. Hypoplasia of ACA and Aplasia both account for 31.2% and 10.6% respectively (López-Sala et al., 2020; Makowicz et al., 2013).

We describe a relatively rare variant of ACA, complete fenestration or partial bifurcation of the right A1. The term bifurcation is used cautiously instead of fenestration, as the two segments did not share an adventitia and connected to the A2 segment, as opposed to the classic fenestration, which merges with the same arterial segment and shares an adventitia.

Fenestration of the right A1 resulted in two separate A1 branches, a medial and lateral segment, with the medial segment connecting to the left A2 via the Acom and the lateral segment directly connecting into the A2 on the same side. Sonda and Basso (2015) reported a similar type of partial bifurcation of A1 in a cadaveric dissection, but in their case it was associated with a duplication of Acom. The association of fenestration with aneurysms has been described in literature. Makowicz et al. (2013) attribute this to a focal smooth muscle defect in arterial intima media at a bifurcation site, a so-called “medial” defect. This, in addition to hemodynamic turbulences at the bifurcation site, increases the risk of aneurysm formation and subsequent rapture.

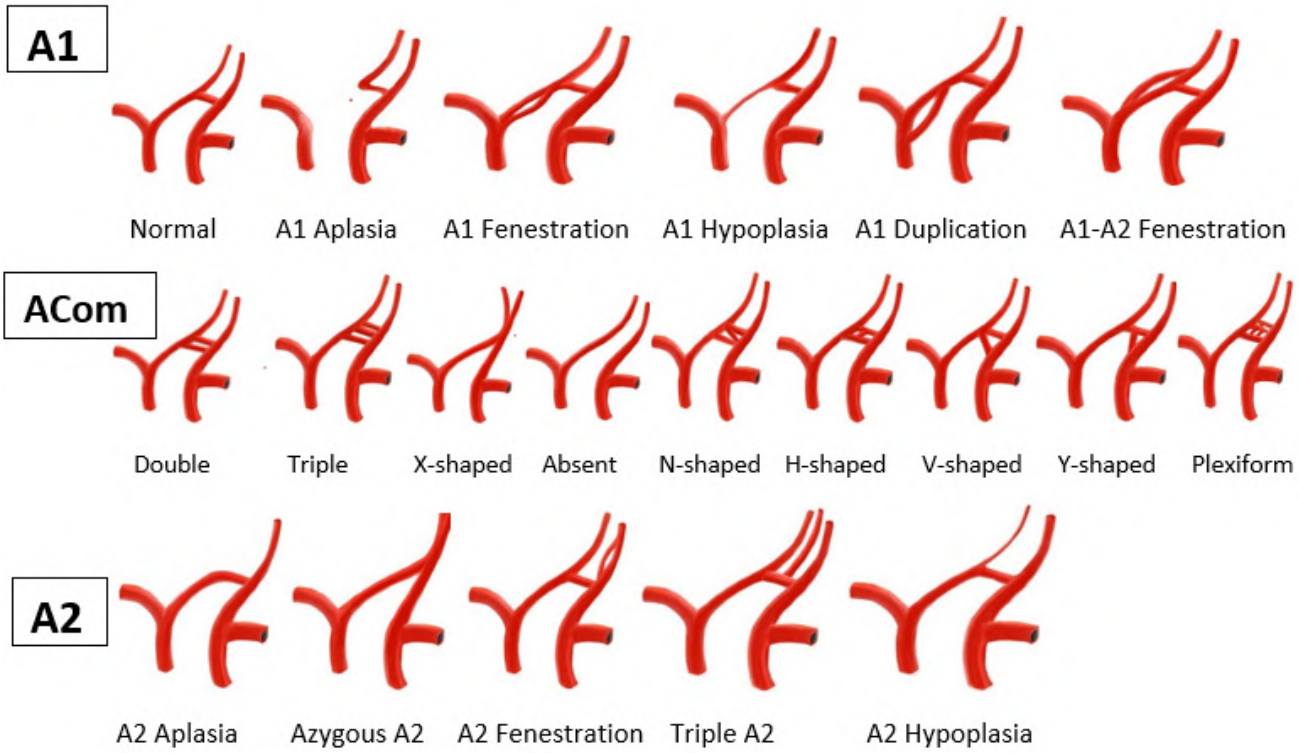

Fig. 3.- Pictural presentation of the many anatomical variations of the Anterior Cerebral Arteries (ACA). They have been grouped into A1, ACom, and A2 (Drawings by Dr. Musa Gerald).

Many cases of fenestrations are asymptomatic and go undiagnosed. Our patient presented with a raptured saccular aneurysm, which was managed with standard microsurgical techniques with very good neurological outcome.

CONCLUSION

Many vascular anomalies like fenestrations and bifurcations are underdiagnosed, as many of them remain asymptomatic and are discovered incidentally on postmortem dissection or angiography for other pathologies. This has led to paucity of cases to determine the prevalence in the human population. A good knowledge of the vascular anatomy variations and associated risk of aneurysm is important for the vascular neurosurgeons.

ACKNOWLEDGEMENTS

The publication was carried out with the support of the Peoples Friendship University of Russia (RUDN) Strategic Academic Leadership Program.

ETHICS APPROVAL AND CONSENT TO PARTICIPATE

Written informed consent was obtained from the patients' caregivers. Consent for publication of this case report and accompanying images was obtained from the patients' caregivers.

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