

Foramen Caecum Vein Involved in Dural Arteriovenous Fistula Fed by Sphenopalatine Arteries: A Case Report

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Abstract

Anterior cranial fossa (ACF) dural arteriovenous fistulas (ACF DAVFs) are an infrequent subtype of cranial DAVFs that are usually fed by the anterior ethmoidal artery (AEA) and ophthalmic artery (OA) branches. Due to the lack of dural venous sinus in the ACF, they directly drain into cortical veins, resulting in high bleeding tendency. For this reason, ACF DAVFs have to be treated regardless of whether they are symptomatic or not. A 74-year-old man, with hypertension history, came to our attention because of ear pain, dizziness, and impaired hearing for 2 months. No other pathological conditions have been found in his medical history. The patient underwent brain magnetic resonance imaging (MRI) scan and subsequently second level diagnostic investigations with digital subtraction angiography (DSA), showing a foramen caecum (FC) patency and a persistent, enlarged, arterialized FC vein (FCV) involved in an incidental ACF DAVF (Cognard IV), fed mainly by sphenopalatine arteries (SPAs) branches bilaterally. The patient underwent open surgery performed by small high frontal craniotomy with DAVFs complete closure and without any complications. To the best of our knowledge, this is the first case ever described of FCV persistence with involvement in an intracranial vascular malformation. It has been managed by surgical intervention that can be considered, despite the large skin incision, a minimally invasive approach with an excellent cosmetic result and minimal risk of complications.

Keywords: foramen caecum vein, foramen caecum, dural arteriovenous fistula, anterior cranial fossa, frontal

Introduction

Dural arteriovenous fistulas (DAVFs) are vascular lesion characterized by a pathologic shunt between the arterial and the venous system. DAVFs asses only for 10%–15% of intracranial vascular malformation and only 10% of all DAFVs are located in anterior cranial fossa (ACF).^{1,2} Usually these DAVFs subset are mainly fed by anterior ethmoidal artery (AEA) and ophthalmic artery (OA) branches¹ and, due to the lack of dural venous sinus in the ACF, they directly drain into cortical veins, resulting in high bleeding tendency. For this reason, ACF DAVFs have to be treated regardless of whether DAFVs are

symptomatic or not³) and they are usually managed with surgical disconnection rather than endovascular treatment (EVT).⁴ The bleeding risk could be dramatically increased if the venous drainage is mostly due to a usually absent or hypoplastic vein such as the foramen caecum vein (VFC). In fact, VFC is a rare venous collector that, during the embryologic development, connects the nasal mucosa to the superior sagittal sinus (SSS) through foramen caecum (FC) and its persistence in adulthood is reported only in two cases.^{5,6} We described, to the best of our knowledge, the first case of incidental ACF DAVF mainly fed by sphenopalatine artery (SPA) bilaterally and drained via VFC with point of fistula localized in correspondence of a patent FC.

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Case Description

A 74-year-old man, with hypertension history, came to our attention because of ear pain, dizziness, and impaired hearing for 2 months. He had no history

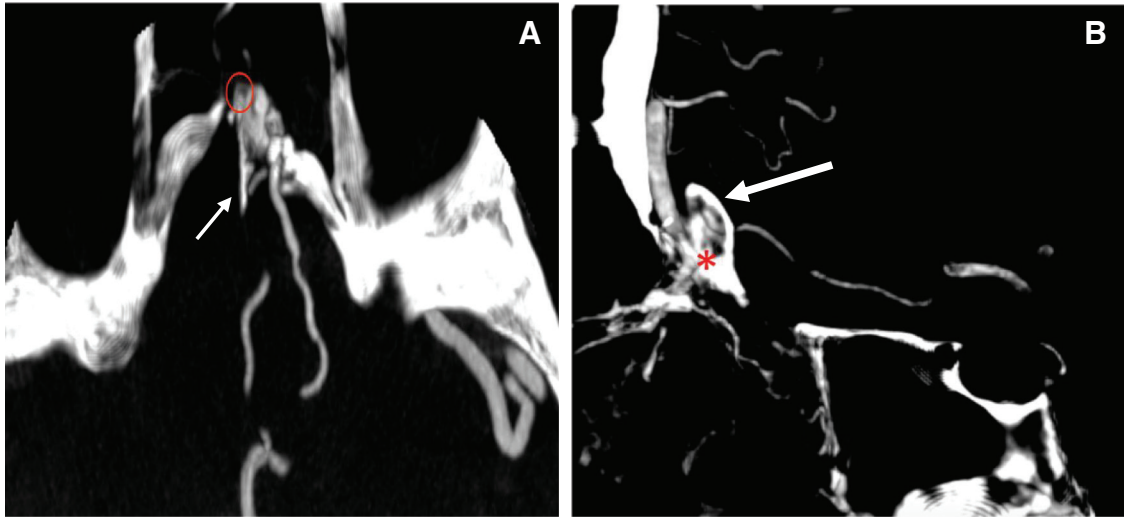


Fig. 1 Preoperative Angio-CT scan (A: axial view; B: sagittal view) shows ACF DAVF involving ectatic VFC and its origin at fistular point in correspondence of the FC (red circle in A and * in B). Arrow: crista galli. ACF DAVF: anterior cranial fossa dural arteriovenous fistula, CT: computed tomography, FC: foramen caecum, VFC: foramen caecum vein.

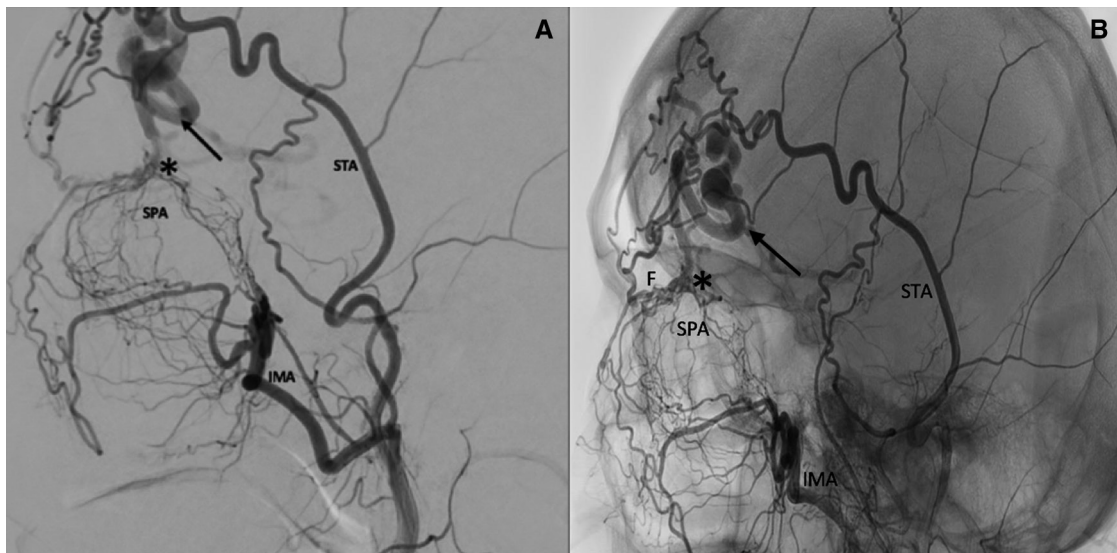


Fig. 2 Preoperative DSA, arterial phase, lateral views of bone subtraction (A) and without bone subtraction view (B) of ACF DAVF involving VFC (arrow) and its origin at fistular point (*). ACF DAVF: anterior cranial fossa dural arteriovenous fistula, DSA: digital subtraction angiography, F: frontal sinus, IMA: internal maxillary artery, SPA: sphenopalatine artery branches from internal maxillary artery (main feeders), STA: superficial temporal artery, VFC: foramen caecum vein.

of past or recent trauma. Otorhinolaryngology examination did not show anatomical changes and neurological examination did not show neither cranial nerve deficit nor sensory-motor impairment. The patient underwent brain magnetic resonance imaging (MRI) scan with and without contrast medium, showing a high flow, abnormal and hypertrophic vascular structure in the left ACF. Second

level diagnostic investigations with digital subtraction angiography (DSA) showed a DAVF fed mainly by SPAs bilaterally and, to a lesser extent, by left superficial temporal artery (STA) branches (Figs. 1 and 2). AEAs were not involved in the arterial blood supply. The feeders converged in front of the Crista Galli, in correspondence of the FC, in the VFC (Fig. 1). The arterialized and ectatic VFC continued as

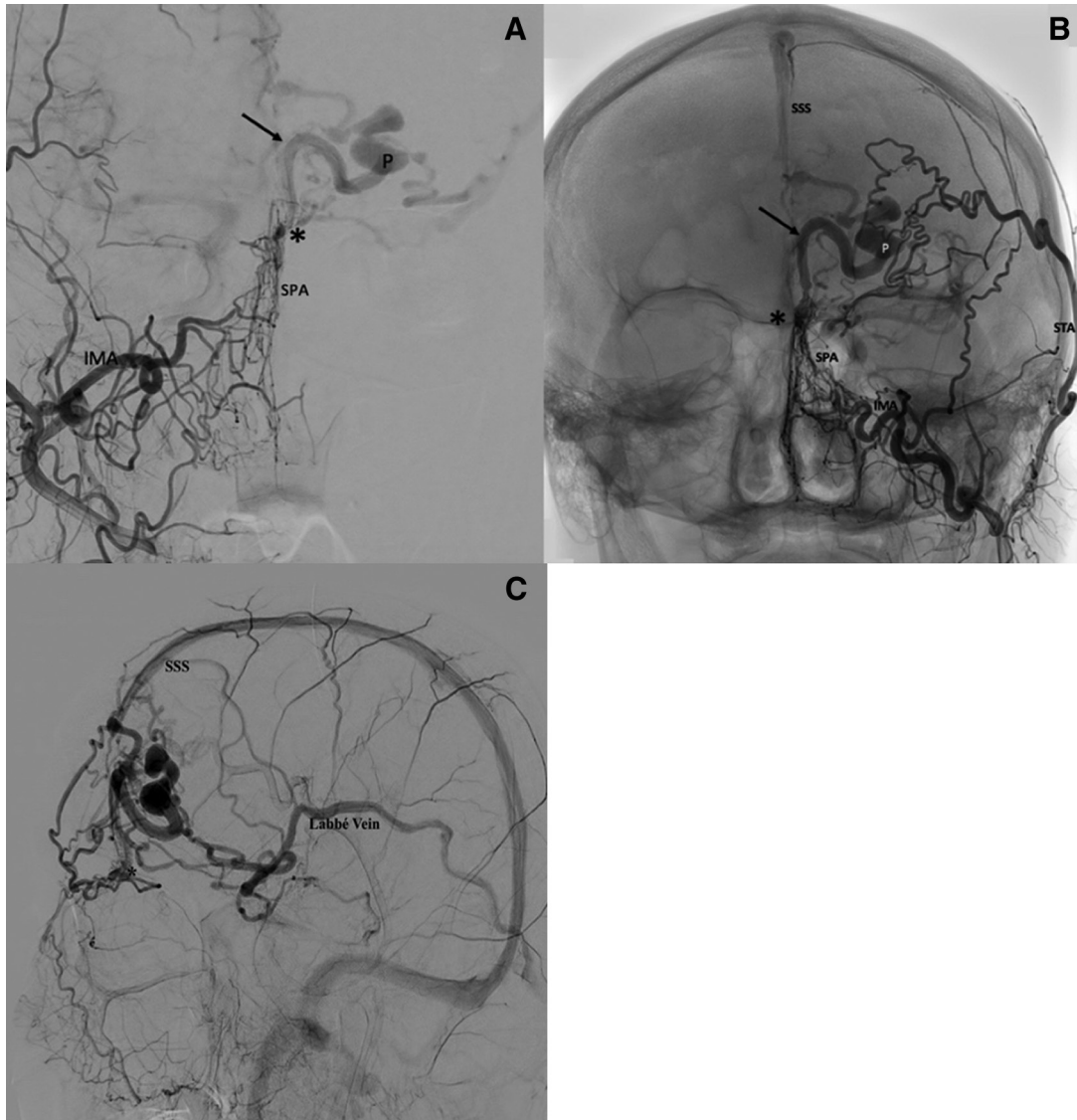


Fig. 3 Preoperative DSA shows ACF DAVF involving VFC (arrow) and its origin at fistular point (*). Anteroposterior view, arterial phase from right (A) and left (B) ECA, P (venous pseudoaneurysm). VFC (C) preoperative DSA, sagittal view showing ACF DAVF and its main drainage in SSS. The first portion of SSS seems to be hypoplastic and not homogeneously filled as possible result of previous sinus thrombosis; it is possible to identify posterior drainage via Labbè vein. ACF DAVF: anterior cranial fossa dural arteriovenous fistula, DSA: digital subtraction angiography, ECA: external carotid artery, IMA: internal maxillary artery, SPA sphenopalatine artery branches from IMA (main feeders), SSS: superior sagittal sinus, STA: superficial temporal artery, VFC: foramen caecum vein.

cortical vein, representing the main venous cortical drainage in the anterior one-third of SSS. It presented a venous pseudoaneurysm (Figs. 3A and 3B) while collateral left frontal vein branches complete the anterior DAVF drainage into the anterior one-third of the SSS, distally to the VFC. The DAVF also presented a posterior venous drainage into the left transverse sinus (TS) via Labbè vein (Fig. 3C) and was classified in DAVF type IV according to Cognard Classification.⁷⁾ After a multidisciplinary board with interventional neuroradiologists, considering the

high risk of bleeding and the anatomical features of the DAVF, it was decided to perform neurosurgical intervention. A cerebrospinal fluid (CSF) lumbar drainage was placed preoperatively to achieve dura mater detension to allow a natural retraction of the frontal lobe, avoiding excessive brain manipulation. After a bicoronal skin incision, a neuro-navigated (Brainlab AG, Heimstetten, Germany) small high frontal craniotomy above the left frontal sinus was performed. The dura was opened and the VFC was identified in the context of engorged frontal cortical

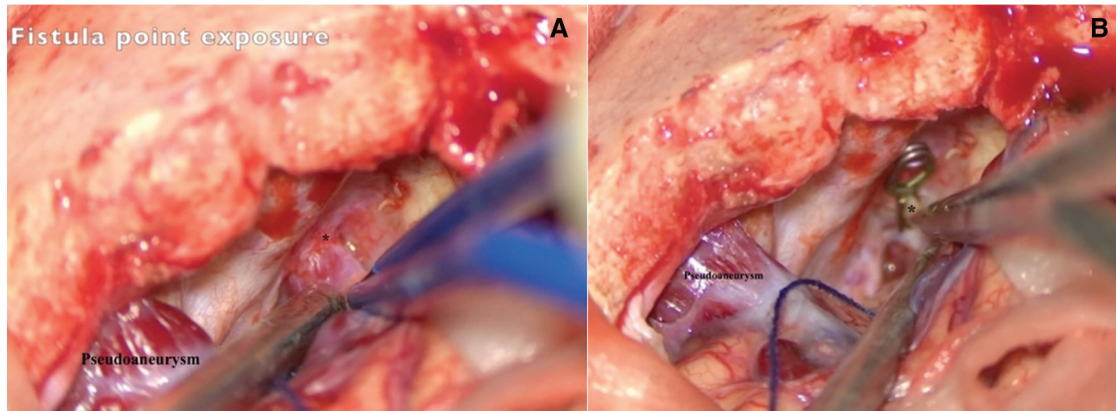


Fig. 4 Intraoperative pictures showing fistular point exposure (*) and pseudoaneurysm (A) and draining vein clipping (B).

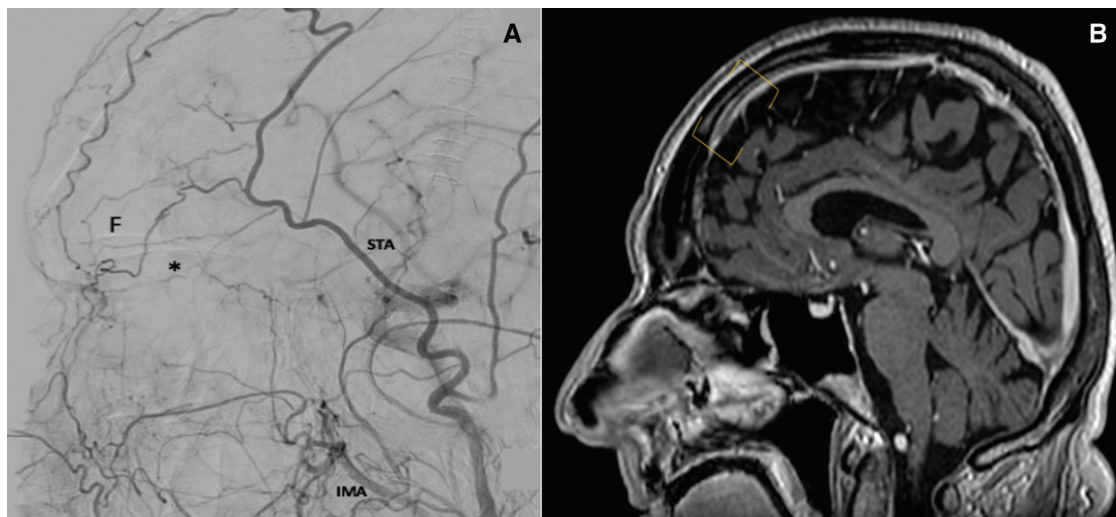


Fig. 5 (A) Postoperative DSA, lateral view, arterial phase, from left ECA; F: frontal sinus; (*): fistular point is not visible anymore. (B) Preoperative post contrast MRI, sagittal view. The first portion of SSS (Square brackets) seems to be extremely hypoplastic and not regularly perfused as result of previous thrombosis. DSA: digital subtraction angiography, ECA: external carotid artery, IMA: internal maxillary artery, MRI: magnetic resonance imaging, SSS: superior sagittal sinus, STA: superficial temporal artery.

veins and followed to the fistular point, located in FC. The fistular point and the draining vein were checked with microscopic indocyanine green (ICG) angiography and closed with a vascular straight clip (Fig. 4). The draining veins became dark and less tense and the disappearance of fistulous flow was verified by a second ICG angiography. Finally, coagulation and section of the venous collector and adjacent pseudoaneurysm were performed. After the surgical procedure, the patient did not show the onset of new neurological deficit, and the postoperative computed tomography (CT) scan did not revealed any postoperative complication. The postoperative DSA showed the complete DAVF closure (Fig. 5A), and patient was discharged after 4 days.

Discussion

ACF DAVFs represent a particular subset of all DAVFs because there is no dural sinus in this location; hence, fistulas drain directly into the frontal cortical veins, making them more prone to bleed.⁸⁾ The natural history of cranial DAVFs is largely predicated on the presence of cortical venous drainage as the most important predictor factor of the patient's clinical presentation. On this basis, the Cognard classification is one of the most used system for categorizing DAVFs.⁹⁾ The high bleeding risk and the associated neurological morbidity require treatment regardless of whether DAVFs are symptomatic or not.³⁾ Due to its rarity, the literature concerning

the ACF DAVFs is largely based on case reports and small case series that provide the current features of ACF DAVFs in terms of clinical presentation, anatomy, and treatment. DAVFs typically present with decreased mental function, neuropsychiatric syndromes, seizures, pulsatile tinnitus, bruit or intracranial haemorrhage and the essential feature associated with development of neurological deficit or intracranial haemorrhage is congestion of the brain venous system.²⁾ From the anatomical point of view, ACF DAVFs are usually fed by EA branches (50–100%) and distal internal maxillary artery (IMA) branches (20–66%) with unilateral or bilateral supply, middle meningeal artery (MMA) (22%), cavernous internal carotid artery (ICA) dural branches (7%), and anterior cerebral artery (ACA) (7%) depending on the series.^{2,10–13)} The venous drainage is usually considered anterior when provided by orbitofrontal or frontopolar veins (70%) or posterior via posterior orbitofrontal and olfactory veins (30%). ACF DAVFs with posterior drainage include also those provided by vein of Labbé or Trolard (11%) and those with deep venous drainage in the vein of Rosenthal (19%), which probably account for that cases with first presentation characterized by intracerebral haemorrhage far from the fistular point.^{13–15)} In this report, we describe the first case of FC patency and VFC persistence, associated with a vascular malformation. The FC represents a primitive tract that during embryologic development contains a dural diverticulum which extends from the ACF to the dermal surface of the nose. Normally, the dural diverticulum undergoes complete involution and the FC fills in with fibrous tissue and variably ossifies. In adulthood, less than 1.5% of foramina remain opened and rarely transmits the VFC.¹⁶⁾ The close interconnection between the nasal mucosa and the VFC, in this particular case, is also confirmed by the arterial supply that is strictly provided by SPAs without any involvement of the EAs. Differently from others vascular disorders such as brain capillary telengectasias (BCTs), developmental venous anomalies (DVAs), cavernous malformation (CM), and arteriovenous malformations (AVMs) for which the belonging to a unique congenital spectrum of disease has recently been hypothesized,^{17,18)} DAVFs in adulthood are generally considered acquired conditions. The pathogenetic mechanism underlying DAVFs formation has usually been related to the venous sinus thrombosis while angiocentric growth factors such as vascular endothelial growth factor (VEGF) and basic fibroblastic growth factor (bFGF) promote neovascularization and development of a DAVF; hence, they should not be considered as a direct cause of DAVFs but rather predisposing factor for their

evolution.¹⁹⁾ In the case described, although non-evident signs of sinus thrombosis appear on the DSA, it is possible that the sinus recanalization occurred after a previous thrombosis and the inhomogeneous filling of the anterior one-third of the SSS could be the result of this event (Fig. 5B). This hypothesis, though speculative, is supported by the evidence in literature, of spontaneous dural sinus recanalization with or without associated fistula resolution.^{20,21)} In this limited number of cases, DAVF resolution could anticipate or occur after the sinus recanalization confirming, on the one hand the pathogenetic role of sinus thrombosis and, on the other hand, suggesting that venous hypertension (even in absence of thrombosis) could initiate as well as maintain DAVF patency. Moreover, the only two cases described in literature of nasal mucosa venous drainage into the SSS via VFC, presented, both, a hypoplastic anterior portion of the SSS, replaced by bilateral longitudinal frontal cortical veins.^{5,6)} This finding further validates the theory that venous hypertension could play a role in keeping the VFC patent, preventing its involution and establishing the conditions for DAVF formation in this extremely rare subset of patients.²⁰⁾ ACF DAVFs are usually managed with surgical disconnection rather than EVT because of the difficulty of navigating with EVT numerous or tortuous arterial feeders and risk of inadvertent embolization of the central retinal artery (CRA). In addition, surgical disconnection in ACF DAVFs has the highest complete obliteration rate compared with EVT.⁴⁾ The safety and efficacy of surgical procedures have been improved performing less invasive craniotomies that do not require excessive fronto-basal craniotomy and the frontal sinus opening to directly expose the cribriform plate and the DAVF. We performed, in accordance with a recent review of literature,²²⁾ a small unilateral high frontal craniotomy, with the advantages of less destructive bone work, frontal sinus sparing and less brain retraction; on the same time, the CSF diversion allows a dynamic brain retraction that reduces the risk of unintentional dural opening and the possible superficial veins breaking during craniotomy.^{23,24)}

Conclusion

To the best of our knowledge, this is the first case ever described of VFC persistence and involvement in intracranial vascular malformation. It has been managed by surgical intervention that can be considered, despite the large skin incision, a minimally invasive approach with excellent cosmetic results and minimal risk of complications.

Patient Consent

The patient has consented to the submission of the case report for submission to the journal.

Conflicts of Interest Disclosure

The authors declare that they have no conflict of interest.

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