

Extensive basal ganglia edema caused by a traumatic carotid-cavernous fistula: a rare presentation related to a basal vein of Rosenthal anatomical variation

Case report

ISABELLE RACT, M.D.,¹ AURÉLIE DRIER, M.D.,² DELPHINE LECLERCQ, M.D.,²
NADER SOUROUR, M.D.,² JOSEPH GABRIELI, M.D.,³ MARION YGER, M.D.,⁴
AURÉLIEN NOUET, M.D.,⁵ DIDIER DORMONT, M.D.,² JACQUES CHIRAS, M.D.,²
AND FRÉDÉRIC CLARENÇON, M.D.²

¹Department of Radiology, Pontchaillou Hospital, CHU Rennes; Departments of ²Neuroradiology, ⁴Vascular Neurology, and ⁵Neurosurgery, Pitié-Salpêtrière Hospital, Paris VI University, Paris, France; and

³Department of Radiology, University of Padova, Italy

The authors report a very rare presentation of traumatic carotid-cavernous fistula (CCF) with extensive edema of the basal ganglia and brainstem because of an anatomical variation of the basal vein of Rosenthal (BVR). A 45-year-old woman was admitted to the authors' institution for left hemiparesis, dysarthria, and a comatose state caused by right orbital trauma from a thin metal rod. Brain MRI showed a right CCF and vasogenic edema of the right side of the brainstem, right temporal lobe, and basal ganglia. Digital subtraction angiography confirmed a high-flow direct CCF and revealed a hypoplastic second segment of the BVR responsible for the hypertension in inferior striate veins and venous congestion. Endovascular treatment was performed on an emergency basis. One month after treatment, the patient's symptoms and MRI signal abnormalities almost totally disappeared.

Basal ganglia and brainstem venous congestion may occur in traumatic CCF in cases of a hypoplastic or agenetic second segment of the BVR and may provoke emergency treatment.
(<http://thejns.org/doi/abs/10.3171/2014.1.JNS132016>)

KEY WORDS • traumatic carotid-cavernous fistula • MRI • basal ganglia • basal vein of Rosenthal • edema • vascular disorders

ADIRECT carotid-cavernous fistula (CCF) is an abnormal communication between the internal carotid artery (ICA) and the cavernous sinus leading to a high-flow shunt.¹¹ The mechanisms that may be responsible for a direct CCF are traumatic conditions and spontaneous rupture of an aneurysm located on the cavernous segment of the ICA. Clinically, patients with a direct CCF usually present with pulsating proptosis, corneal edema, and chemosis that are related to engorgement of the superior ophthalmic vein draining into the cavernous sinus.¹² Rarely, CCFs are responsible for brainstem edema due to reflux into a superior petrosal sinus (SPS) draining the lateral mesencephalic vein.⁷

We describe a rare presentation of traumatic CCF

with edema of the basal ganglia (BG) due to an anatomical variation of the basal vein of Rosenthal (BVR).

Case Report

History and Examination. A 45-year-old woman was emergently admitted to our hospital for dysarthria, left hemiparesis, and headache rapidly evolving to a comatose state. Right ocular trauma from a thin metal rod had recently occurred. Brain MRI (3 T) was emergently performed (Fig. 1). Three-dimensional time of flight (TOF) acquisition showed hyperintense signal within the right cavernous sinus; these findings were suggestive of a direct CCF. Both FLAIR and T2-weighted images showed a hyperintense signal in the right side of the pons, the mesencephalon extending to the right middle cerebellar peduncle, the right BG, and the medial aspect of the right temporal lobe. Since apparent diffusion coefficient values were not decreased in these areas, the T2-weighted hyperintense signals were suggestive of vasogenic edema.

Abbreviations used in this paper: BG = basal ganglia; BVR = basal vein of Rosenthal; CCF = carotid-cavernous fistula; DSA = digital subtraction angiography; ICA = internal carotid artery; SPS = superior petrosal sinus; SWAN = susceptibility-weighted angiography; TOF = time of flight.

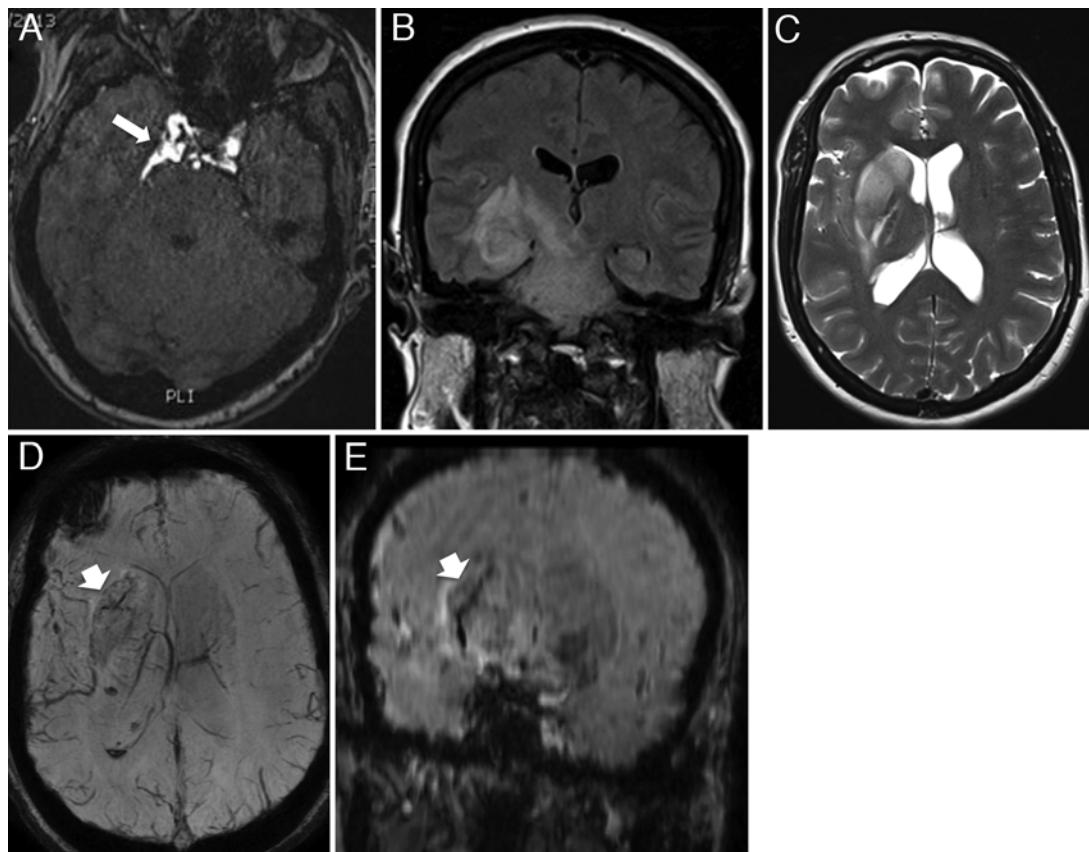


Fig. 1. A 45-year-old woman presented with left hemiparesis, dysarthria, and headache rapidly evolving to a comatose state because of right ocular trauma from a thin metal rod. Three-dimensional TOF image (A) showing hyperintense signal within the right cavernous sinus, which is bulging laterally. These findings are suggestive of a direct CCF. Coronal FLAIR (B) and axial T2-weighted (C) images showing a hyperintense signal involving the brainstem, the medial aspect of the right temporal lobe, and the right BG. Three-dimensional axial SWAN acquisition (D) and MinIP SWAN coronal reconstruction (E) showing a dilated vein close to the BG (white arrows). Note the presence of an intraventricular hemorrhage in the occipital horn of the right lateral ventricle (D), probably related to the direct trauma.

Susceptibility-weighted angiography (SWAN) images showed a focal hematoma in the right side of the pons associated with a faint intraventricular hemorrhage probably from the direct trauma caused by the thin metal rod. Additionally, a tube-shaped structure suggestive of a dilated vein was seen close to the right BG (Fig. 1).

Treatment. Digital subtraction angiography (DSA) was performed with the patient under general anesthesia for diagnostic and therapeutic purposes (Fig. 2). These imaging studies confirmed a high-flow direct CCF in the right cavernous portion of the ICA with reflux toward the contralateral cavernous sinus. Precise analysis of the venous network on the DSA studies revealed the presence of a hypoplastic second segment of the BVR; the first segment and its branches mainly drained into the cavernous sinus via the uncal vein. Thus, at the arterial phase, both the right cavernous sinus and the first segment of the BVR and its branches were opacified via the torn right ICA. It is noteworthy that the right SPS did not appear patent on DSA.

Because there was no filling of the supraclinoid branches of the right ICA, the circle of Willis was functional, and the tear on the ICA was large, the decision was

made to close both the fistula and the right ICA endovascularly with coils via a double-lumen balloon (Ascent 6–9 mm, Micrus, Johnson & Johnson). The procedure was performed under full heparinization (activated clotting time between 2- and 3-fold greater than baseline) without any technical difficulty.

Posttreatment Course. At the 1-month clinical follow-up, the patient had a satisfactory recovery, with only slight residual left hemiparesis. On follow-up MRI (3 T), the T2 and FLAIR signal abnormalities had almost totally disappeared (Fig. 3).

The angiography findings (first segment of the BVR draining into the cavernous sinus) and the resolution of MRI signal abnormalities on follow-up after shunt occlusion confirmed the initial diagnosis of engorgement of the veins draining into the first segment of the BVR due to retrograde filling related to the direct CCF.

Discussion

Carotid-cavernous fistulas can be classified into 4 groups according to Barrow et al.¹ The first group, Type A, the most severe type, is a direct connection between

Atypical traumatic carotid-cavernous fistula

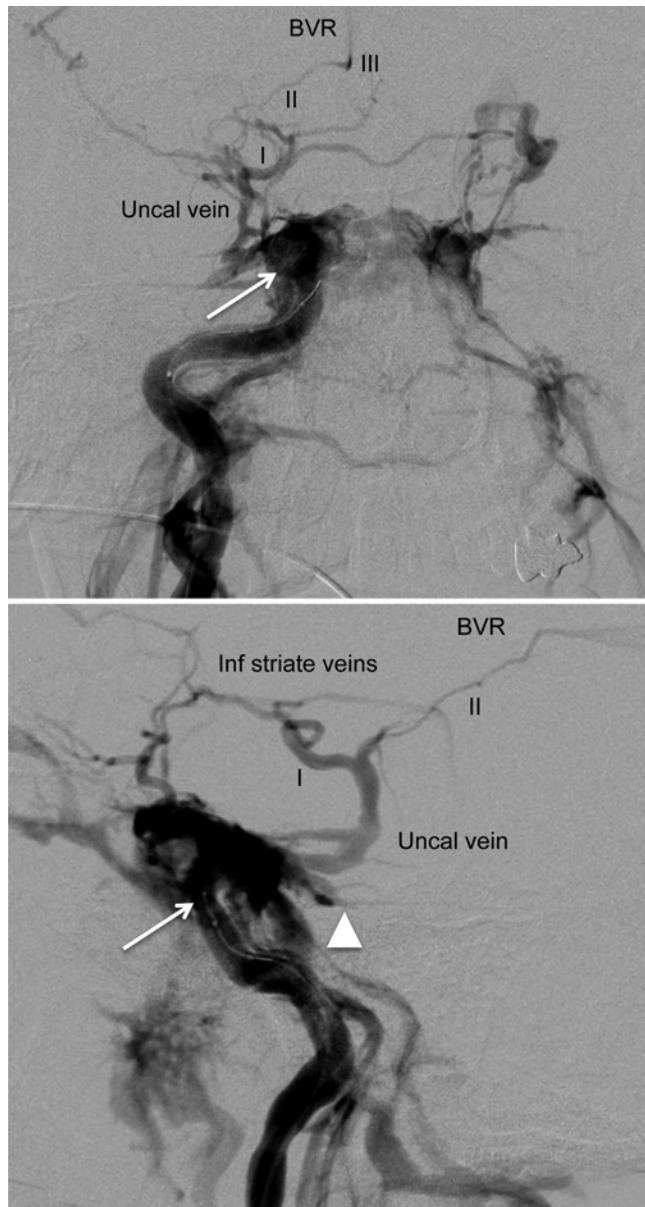


FIG. 2. Right ICA digital subtraction angiograms, anteroposterior (upper) and lateral (lower) projections. Opacification of both cavernous sinuses is seen at the arterial phase, confirming the diagnosis of a right direct CCF (arrows). On the lateral projection, a dilated first segment of the BVR appears as well as the connection between the BVR and the cavernous sinus via the uncal vein. The second segment of the BVR appears hypoplastic. Note the absence of opacification of the SPS (arrowhead). Inf = inferior; I, II, and III = first, second, and third segments of the BVR.

the ICA and the cavernous sinus. The other groups, Types B, C, and D, are each an indirect dural communication between dural arteries and the cavernous sinus. Our patient had a posttraumatic Type A direct CCF.

The clinical presentation of CCFs depends on the degree of shunting and the venous drainage pathways. The cavernous sinus receives drainage anteriorly from the ophthalmic veins and superiorly from sphenoparietal sinus and cortical veins. Then blood flow into the cavernous sinus drains posteriorly into the inferior petrosal

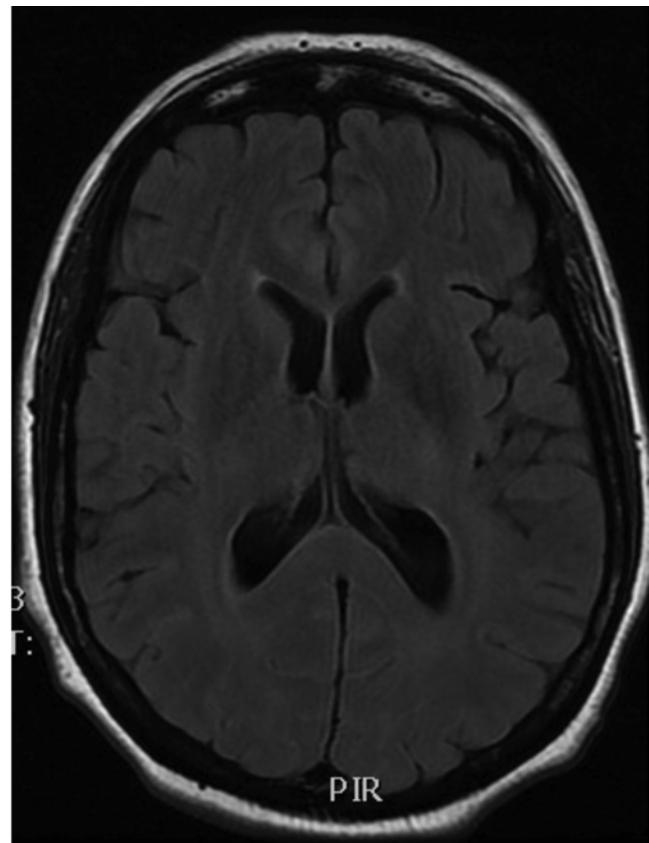


FIG. 3. Axial FLAIR image obtained at the 1-month follow-up, showing total disappearance of the signal abnormalities.

sinus and the SPS and inferiorly into the pterygoid plexus via emissary veins.^{7,11}

Generally, patients present with the classic clinical triad of proptosis, chemosis, and pulsatile bruits caused by orbital venous congestion. Less commonly, the venous congestion can be posterior (cerebral and brainstem), causing neurological symptoms rather than ocular ones. Rarely, reflux into the cortical veins can be observed and represents a risk factor for focal neurological deficit, seizure, venous infarction, or even intracranial hemorrhage.¹¹

Mechanisms leading to venous infarction caused by venous congestion have been studied for indirect CCFs,^{2,8} but remain poorly understood for traumatic direct CCFs. In indirect dural CCFs, risk factors for venous infarction or intracranial hemorrhage are as follows: exclusive cortical venous drainage, ectasia of the draining vein, agenesis of the second and/or third segments of the BVR, thrombosis of the superior orbital vein or SPS, and distal cortical venous drainage.³⁻⁵ For direct CCFs, venous cerebral congestion has been reported in 6 cases,^{4,6,9,14,15} all of which showed brainstem congestion, including one case associated with congestion of the cervical spinal cord.⁶ In 3 cases, as seen in our patient, the SPS was not patent. However, to our knowledge, there is no case in the literature of BG venous congestion related to flow stagnation of veins drained by the first segment of the BVR and caused by hypoplasia of its second segment.

The BVR is an anastomotic venous channel generally

connecting the cavernous sinus to the great cerebral vein (the so-called vein of Galen). Its course turns around the cerebral peduncle and presents a close relationship with the posterior cerebral artery. Typically, the BVR is divided into 3 segments:^{8,13} The first segment, also called the anterior or striate segment, mainly drains the inferior striate veins, deep cerebral vein, insular veins, and olfactory vein. The second, or peduncular, segment mainly drains the peduncular vein, inferior ventricular vein, inferior choroidal vein, and hippocampal veins. The third, or posterior mesencephalic, segment mainly drains the lateral mesencephalic vein and the posterior thalamic vein.

However, the BVR presents some anatomical variations. Indeed, Suzuki et al.,¹³ after performing 3D CT angiography in 250 patients, reported that the connection between the first and second segments of the BVR was not seen (agenesis or hypoplasia) in 36.9% of the cases.

A simple classification of the potential venous drainages of the BVR has been established according to the 5 drainage pathways formed during the early embryonic stage.¹⁰ Drainage can be toward the vein of Galen, toward the cavernous sinus or sphenoparietal sinus, toward the SPS via the lateral mesencephalic vein, toward the SPS via a peduncular vein, and toward the transverse sinus or straight sinus via the tentorium sinuses.

If the uncal vein, which connects the cavernous sinus to the first segment of the BVR, is well developed, the connection between the first and second segments is usually hypoplastic or agenetic, as observed in our case. Since the first segment of the BVR drains the inferior striate veins and some hippocampal veins, engorgement of this segment by the CCF via the uncal vein explains the signal abnormalities seen on emergency MRI. Finally, the CCF occlusion in our case resolved engorgement of the first segment of the BVR and its tributaries, which was confirmed by the favorable clinical and imaging outcome.

Conclusions

Brainstem and BG venous congestion is a rare but potentially serious complication of traumatic CCF. The association of a large tear on the ICA, a developed uncal vein with a hypoplastic second segment of the BVR, and a thrombosed or hypoplastic SPS may favor this rare complication of a direct CCF. Neurosurgeons as well as interventional neuroradiologists should be aware of this unusual presentation of CCF that provokes emergency treatment.

Disclosure

Dr. Sourour is a consultant for Covidien and Penumbra. The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

Author contributions to the study and manuscript preparation include the following. Conception and design: Clarençon, Ract, Drier, Leclercq, Sourour, Gabrieli, Dormont. Acquisition of data: Clarençon, Ract, Drier, Leclercq, Sourour, Gabrieli, Yger, Chiras. Analysis and interpretation of data: all authors. Drafting the article: Clarençon, Ract, Drier, Leclercq, Sourour, Gabrieli, Nouet, Dormont, Chiras. Critically revising the article: all authors. Reviewed submitted version of manuscript: all authors. Study supervision: Clarençon.

References

1. Barrow DL, Spector RH, Braun IF, Landman JA, Tindall SC, Tindall GT: Classification and treatment of spontaneous carotid-cavernous sinus fistulas. *J Neurosurg* **62**:248–256, 1985
2. Blanc R, Maia Barros AD, Brugieres P, Méder JF, Gaston A: [Cavernous sinus dural arteriovenous fistula complicated by edematous cerebral lesions from venous etiology.] *J Neuro-radiol* **31**:220–224, 2004 (Fr)
3. Brown RD Jr, Wiebers DO, Nichols DA: Intracranial dural arteriovenous fistulae: angiographic predictors of intracranial hemorrhage and clinical outcome in nonsurgical patients. *J Neurosurg* **81**:531–538, 1994
4. Bussière M, Lownie SP, Pelz DM, Nicolle D: Direct carotid-cavernous fistula causing brainstem venous congestion. *J Neuroophthalmol* **29**:21–25, 2009
5. Cognard C, Gobin YP, Pierot L, Bailly AL, Houdart E, Casasco A, et al: Cerebral dural arteriovenous fistulas: clinical and angiographic correlation with a revised classification of venous drainage. *Radiology* **194**:671–680, 1995
6. Herrera DA, Vargas SA, Dublin AB: Traumatic carotid-cavernous fistula with pontomesencephalic and cervical cord venous drainage presenting as tetraparesis. *J Neuroimaging* **21**:73–75, 2011
7. Kiyosue H, Hori Y, Okahara M, Tanoue S, Sagara Y, Matsumoto S, et al: Treatment of intracranial dural arteriovenous fistulas: current strategies based on location and hemodynamics, and alternative techniques of transcatheter embolization. *Radiographics* **24**:1637–1653, 2004
8. Miyamoto N, Naito I, Takatama S, Shimizu T, Iwai T, Shimoguchi H: Clinical and angiographic characteristics of cavernous sinus dural arteriovenous fistulas manifesting as venous infarction and/or intracranial hemorrhage. *Neuroradiology* **51**:53–60, 2009
9. Murata H, Kubota T, Murai M, Kanno H, Fujii S, Yamamoto I: Brainstem congestion caused by direct carotid-cavernous fistula—case report. *Neurul Med Chir (Tokyo)* **43**:255–258, 2003
10. Padget DH: The cranial venous system in man in reference to development, adult configuration, and relation to the arteries. *Am J Anat* **98**:307–355, 1956
11. Ringer AJ, Salud L, Tomsick TA: Carotid cavernous fistulas: anatomy, classification, and treatment. *Neurosurg Clin N Am* **16**:279–295, viii, 2005
12. Shownkeen H, Bova D, Origitano TC, Petruzzelli GJ, Leonetti JP: Carotid-cavernous fistulas: pathogenesis and routes of approach to endovascular treatment. *Skull Base* **11**:207–218, 2001
13. Suzuki Y, Ikeda H, Shimadu M, Ikeda Y, Matsumoto K: Variations of the basal vein: identification using three-dimensional CT angiography. *AJNR Am J Neuroradiol* **22**:670–676, 2001
14. Teng MM, Chang T, Pan DH, Chang CN, Huang CI, Guo WY, et al: Brainstem edema: an unusual complication of carotid cavernous fistula. *AJNR Am J Neuroradiol* **12**:139–142, 1991
15. Turner DM, Vangilder JC, Mojtahedi S, Pierson EW: Spontaneous intracerebral hematoma in carotid-cavernous fistula. Report of three cases. *J Neurosurg* **59**:680–686, 1983

Manuscript submitted September 23, 2013.

Accepted January 13, 2014.

Please include this information when citing this paper: published online February 14, 2014; DOI: 10.3171/2014.1.JNS132016.

Address correspondence to: Frédéric Clarençon, M.D., Department of Neuroradiology, Pitié-Salpêtrière Hospital, 47 Blvd de l'Hôpital, Paris 75013, France. email: fredclare5@msn.com.