



# Dural carotid-cavernous fistula presenting with confusion and expressive aphasia

Sean Woolen<sup>b</sup>, Joseph J. Gemmete<sup>a,b,c,\*</sup>, Aditya S. Pandey<sup>a,b</sup>, Neeraj Chaudhary<sup>a,b</sup>

<sup>a</sup>Department of Neurosurgery, Room UH B1 D328, 1500 East Medical Center Drive, Ann Arbor, MI 48109, USA

<sup>b</sup>Department of Radiology, University of Michigan Hospitals, Ann Arbor, MI, USA

<sup>c</sup>Department of Otolaryngology, University of Michigan, Ann Arbor, MI, USA

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## ABSTRACT

We report a patient who presented with confusion and expressive aphasia at the onset of a cortical venous reflux (CVR) related to a known dural carotid-cavernous fistula (DCCF). CVR and venous congestion are rare findings in a DCCF, however, it is important to recognize these findings early to reduce the associated risks of venous hemorrhage and infarction. To our knowledge, no previous report has described an association of expressive aphasia with CVR from a DCCF. This patient highlights the potential for venous congestion secondary to CVR from a DCCF presenting with a constellation of symptoms, including confusion and expressive aphasia with corresponding MRI findings. Clinicians should consider a DCCF in the differential diagnosis of this clinical presentation, with corroborative imaging findings.

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## 1. Introduction

Cortical venous reflux (CVR) and venous congestion are rare findings in a dural carotid-cavernous fistula (DCCF), however, it is important to recognize these findings early to reduce the associated risks of venous hemorrhage and infarction [1–4]. We report a patient who presented with confusion and expressive aphasia at the onset of CVR from a DCCF.

## 2. Case report

A 68-year-old woman was referred for evaluation and treatment of a DCCF. One year prior to treatment, she experienced double vision, primarily affecting her left eye, with no other reported symptoms. She was diagnosed with a DCCF that was treated conservatively, with resolution of clinical symptoms at 3 months. She then developed symptoms in her right eye 6 months after the initial diagnosis, consisting of difficulty with eye movement, blurred vision, diplopia, and proptosis. The diplopia was corrected by refractory lens and timolol maleate ophthalmic solution eye drops (Timoptic; Merck, Whitehouse Station, NJ, USA).

While traveling on an airplane 1 year after her initial diagnosis, she developed acute mental status changes, confusion, and expressive aphasia with no other neurological deficits. Brain MRI was obtained, and when compared to the prior MRI, it demonstrated a new hyperintense fluid-attenuated inversion recovery (FLAIR) signal abnormality within the left frontal operculum (Fig. 1A). An ophthalmology examination confirmed persistent proptosis of the right eye with an increase in intraocular pressure and gaze palsy. A decision was made to perform embolization of the DCCF.

A cerebral angiogram was performed to define the extent of the fistula. Transarterial access was obtained in the right common femoral artery, and a 4-French sheath was placed. A 4-French vertebral catheter was used to selectively catheterize the right internal carotid artery (ICA), right external carotid artery (ECA), right vertebral artery, left ICA, left ECA, and left vertebral artery. An angiogram of each of the selected vessels was performed in the anteroposterior and lateral projections.

The cerebral angiogram identified bilateral DCCF. The right ICA angiogram demonstrated early filling of the right cavernous sinus from the meningohypophyseal trunk, with drainage anteriorly into the ophthalmic vein and contralateral filling of the left cavernous sinus, with CVR in the left cerebral hemisphere (Fig. 1B). The left ECA angiogram demonstrated early filling of the left cavernous sinus with CVR into the left superficial middle cerebral vein (SMCV) and left superior petrosal sinus (SPS), with partial filling of the right cavernous sinus (Fig. 1C). The left ICA angiogram demonstrated early filling of the left cavernous sinus from branches of the meningohypophyseal trunk.

The bilateral DCCFs were treated via a transvenous approach through the inferior petrosal sinus by placing multiple microcoils into the left and right cavernous sinuses. Selective venograms were performed by injecting contrast into the left cavernous sinus, which demonstrated CVR with filling of the left SMCV and left SPS (Fig. 2A, B). A cone beam CT scan showed the engorged venous channels within the left frontal operculum corresponding to the hyperintense FLAIR signal abnormality within the left frontal operculum (Fig. 2C). The final cerebral angiogram showed complete occlusion of the fistula (Fig. 3A, B).

The clinical follow-up visits at 1 month and 1 year showed resolution of the clinical signs and symptoms of confusion, expressive aphasia, right eye proptosis, restriction of gaze movements, and diplopia. The intraocular pressure normalized in both eyes. The MRI showed resolution of the hyperintense FLAIR signal abnormality in the left frontal operculum due to normalization of the venous congestion (Fig. 3C). The follow-up cerebral angiogram, 6 months after the embolization, demonstrated continued occlusion of the fistula.

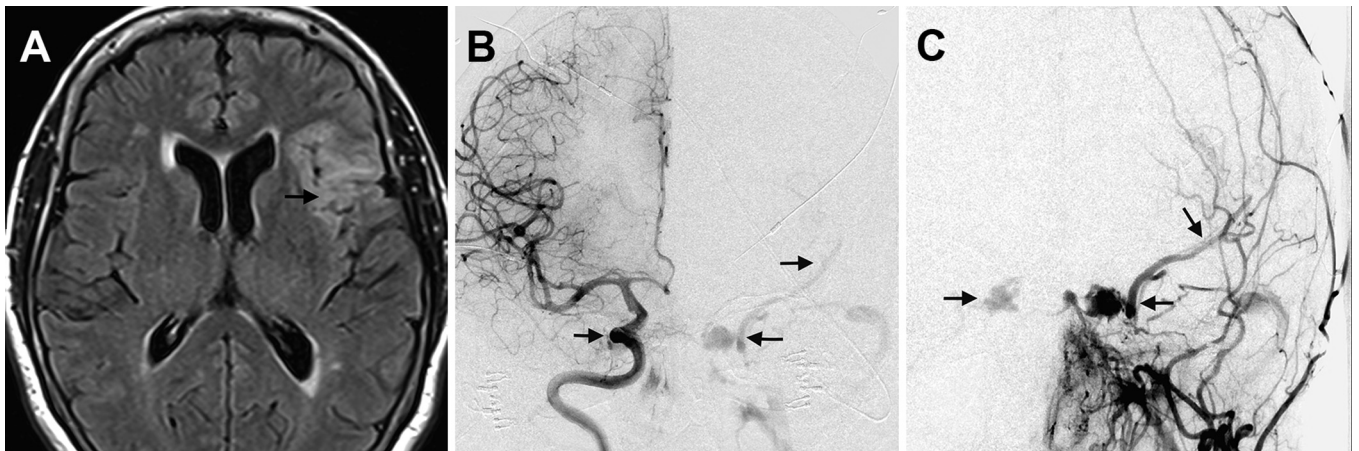
## 3. Discussion

DCCF have a slow, progressive course, with the majority of patients presenting with progressive glaucoma, proptosis, ophthalmoparesis, or conjunctival injection [2,5]. The natural history of a DCCF is variable, and the literature reports that spontaneous resolution without treatment occurs in 10–60% of patients, possibly due to spontaneous thrombosis of the involved segment of the cavernous sinus [6].

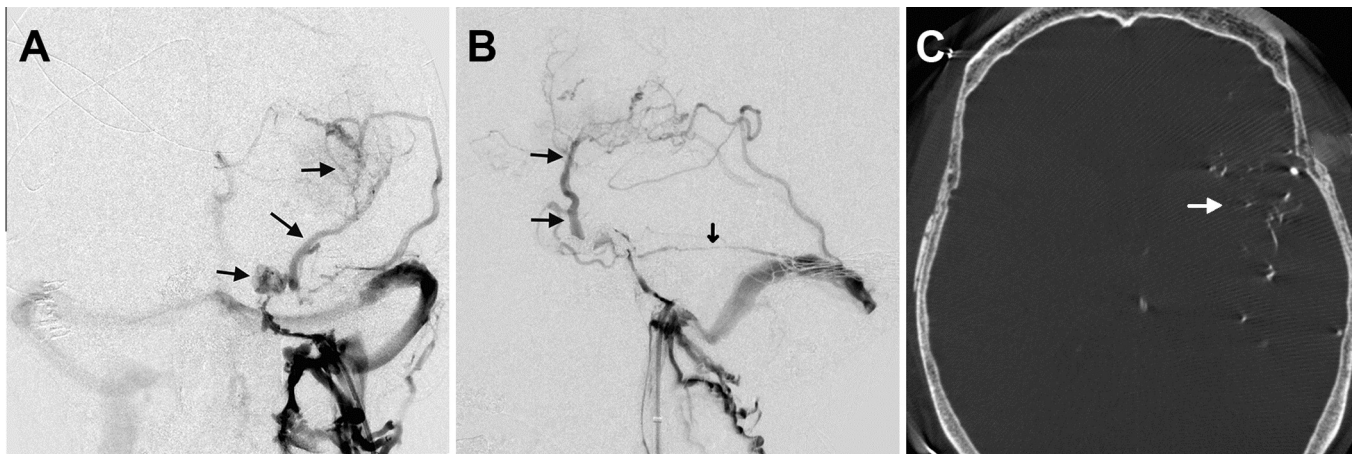
CVR can occur in 27–63% of carotid-cavernous fistula (CCF) patients with a majority of these from direct CCF [3,4]. CCF with

\* Corresponding author. Tel.: +1 734 936 4566; fax: +1 734 764 4230.

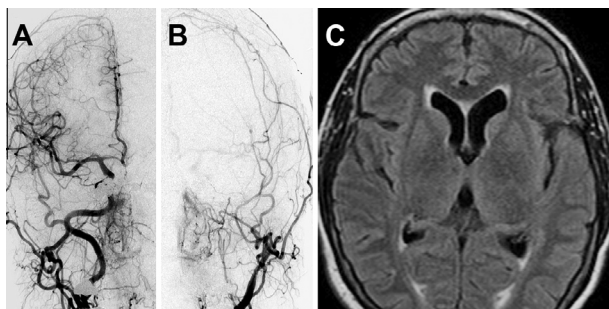
E-mail address: gemmete@med.umich.edu (J.J. Gemmete).



**Fig. 1.** (A) Axial, fluid-attenuated inversion recovery (FLAIR) MRI shows abnormal high signal within the left frontal operculum (black arrow). (B) The frontal right internal carotid angiogram shows earlier filling of both cavernous sinuses and the left superficial middle cerebral vein (SMCV; black arrows). (C) The frontal left external carotid angiogram shows early filling of both cavernous sinuses and left SMCV (black arrows).



**Fig. 2.** Selective venograms of the left cavernous sinus in (A) anteroposterior projection shows filling of the cavernous sinus and left SMCV with venous congestion in the left frontal operculum (black arrows), and (B) lateral projection shows retrograde filling of the left SMCV (black arrows) and superior petrosal sinus (small black arrow). There is engorgement of cortical venous structures within the left frontal operculum. (C) The axial cone beam CT scan from contrast injected into the left cavernous sinus shows engorgement of the cortical venous structures (white arrow) within the left frontal operculum corresponding to the abnormal high signal within the left frontal operculum, seen in [Figure 1A](#).



**Fig. 3.** (A) The frontal right common carotid angiogram after coil embolization of both cavernous sinuses shows no early filling of the cavernous sinuses and left superficial middle cerebral vein (SMCV). (B) The frontal left external carotid angiogram after coil embolization of both cavernous sinuses shows no early filling of both cavernous sinuses and venous structures in the left cerebral hemisphere. (C) Axial, fluid-attenuated inversion recovery MRI at 1 month after embolization shows interval resolution of the abnormal high signal within the left frontal operculum.

CVR are considered to be at increased risk for venous hemorrhage or infarction (30–40%) compared to those without CVR [1,2]. Therefore, CVR on digital subtraction angiography is an indication to perform urgent endovascular treatment [2,7]. However, to our knowledge, no previous report discusses the change in clinical symptoms that can occur as a DCCF starts to develop CVR.

Our patient presented with the typical slow progression of symptoms with diplopia, conjunctival injection, and proptosis. Her conservative treatment resulted in symptom progression, including confusion and expressive aphasia associated with the development of CVR. This correlated with a hyperintense FLAIR signal abnormality in the frontal operculum.

#### Conflicts of Interest/Disclosures

The authors declare that they have no financial or other conflicts of interest in relation to this research and its publication.

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## Episodic weakness and vacuolar myopathy in hypokalemic periodic paralysis



Diana Basali<sup>b</sup>, Richard A. Prayson<sup>a,\*</sup>

<sup>a</sup> Department of Anatomic Pathology, Cleveland Clinic, Level 25, 9500 Euclid Avenue, Cleveland, OH 44195, USA

<sup>b</sup> Stritch School of Medicine, Loyola University Chicago, Maywood, IL, USA

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### ABSTRACT

We report a 50-year-old woman who presented with a 20 year history of gradually progressive lower extremity weakness, characterized by knee buckling with occasional falls and foot dragging. She also experienced difficulty in lifting her arms above her shoulders. The primary periodic paralyses are rare disorders caused by dysfunctional ion channels in skeletal muscle. The hypokalemic type is generally an autosomal dominant condition, due to missense mutations in the alpha subunits of the skeletal muscle L-type calcium channel genes, CACN1AS, or the skeletal muscle sodium channel gene, SCN4A. The affected patients typically present with episodic weakness. For our patient, the consumption of foods high in carbohydrates seemed to precipitate the episodes of weakness. Her family history was significant for six blood relatives, including three sons and three relatives on the paternal side, who had experienced similar symptoms. A biopsy of the left rectus femoralis muscle showed vacuolar myopathic changes in the scattered muscle fibers, accompanied by occasional degenerating and regenerating muscle fibers. There was no evidence of inflammation on the biopsy. The vacuoles were often associated with increased acid phosphatase staining. An electron microscopic examination showed that the vacuolar changes were due to T-tubule dilation, a characteristic of hypokalemic periodic paralysis. Other metabolic etiologies of vacuolar myopathy, such as acid phosphatase (lysosomal) associated acid maltase deficiency (a glycogen storage disease), need to be considered in the differential diagnosis.

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### 1. Introduction

The primary periodic paralyses are disorders of dysfunctional ion channels and are classified as either hyperkalemic or hypokalemic, based on serum potassium levels and the response to potassium administration. Hypokalemic periodic paralysis (HPP) is a rare disorder, with a prevalence of 1:100,000, and consists of attacks of flaccid paralysis that often occur at night or early in the morning [1]. The attacks can either occur spontaneously, or be provoked by exercise or a high carbohydrate meal. We report a 50-year-old woman with symptoms, family history, and muscle biopsy findings consistent with HPP.

### 2. Case report

A 50-year-old woman who presented with a 20 year history of gradually progressive proximal lower extremity weakness, described episodes of knee buckling with occasional falls, foot

dragging and difficulty in lifting her arms above her shoulders. One year prior, she fractured her left hip and subsequently required a walker for ambulation. She also described episodes of weakness which seemed to be precipitated by eating certain salads, pasta or pizza dinners. Her family history was remarkable for three adult sons, a paternal aunt and two paternal cousins who had experienced similar episodes of weakness, and a father who had similar symptoms and who died of pneumonia at 39 years of age from suspected respiratory weakness. Her general physical examination was unremarkable. The electromyography studies showed a generalized myopathy, predominantly involving the lower extremities, with associated fibrillation potentials and occasional myotonic discharges. Her electrolytes were normal except for low serum potassium. The sedimentation rate, aldolase, vitamin B12 level and thyroid stimulating levels were normal.

A biopsy of the left rectus femoralis muscle was performed and showed vacuolar myopathic changes in scattered muscle fibers (Fig. 1). The occasional vacuole showed acid phosphatase associated staining. Increased periodic acid-Schiff staining, indicative of glycogen accumulation, was not noted, but occasional degenerating and regenerating muscle fibers were observed. A rare focus of chronic inflammation that was comprised primarily of benign

\* Corresponding author. Tel.: +1 216 444 8805; fax: +1 216 445 6967.

E-mail address: [praysor@ccf.org](mailto:praysor@ccf.org) (R.A. Prayson).