DOI:10.4274/tnd.2023.93992
Turk J Neurol 2023;29:299-301

A Possible Cause for the Development of Spontaneous Carotid Cavernous Fistula: Wegener's Granulomatosis

Spontan Karotis Kavernöz Fistül Gelişiminde Olası Bir Neden: Wegener Granülomatosiz

Remzi Emre Şahin[1], Kezban Aslan-Kara[1], Hasan Bilen Onan[2], Şebnem Bıçakcı[1]
[1]Cukurova University Faculty of Medicine, Department of Neurology, Adana, Türkiye
[2]Cukurova University Faculty of Medicine, Department of Radiology, Adana, Türkiye

Keywords: Wegener granulomatosis, carotid cavernous fistula, Barrow classification
Anahtar Kelimeler: Wegener granülomatoz, karotis kavernöz fistül, Barrow sınıflaması

Dear editor,

Here, the spontaneous carotid cavernous fistula (CCF) accompanying the diagnosis of Wegener's granulomatosis (WG) and the accompanying idiopathic intracranial hypertension (IIH) are discussed. The 65-year-old female patient with WG had been in remission with immunosuppressive treatment for 7 years. Approximately 3 months before her admission, severe bilateral headache, which was predominant in the left forehead, started and worsened daily for a month. We discovered that the headache was compressive, and since hypertension was detected at the time, antihypertensive treatment was provided, and her blood pressure was under control. The severity of the headache gradually increased, and in the last 10 days, complaints of nausea and vomiting, redness in the left eye, a droopy eyelid, and blurred vision in both eyes but predominant in the left eye were added to the headache. No history of trauma was described. During a physical examination, the patient's body mass index was 35.16 and arterial blood pressure was 130/80 mmHg. In the neurological and neuroophthalmological examinations, there was limitation of her outward gaze on the left, proptosis, chemosis, decrease in bilateral visual acuity (0.6, 0.4 L>R), and apparent obliteration of optic disc borders on the left (grade 1, 2). Direct and indirect light reflexes were bilaterally normal (DIR/IDIR: ++/++), relative afferent pupillary defect was negative, and color vision was found to be preserved (Figure 1). Routine laboratory tests including a thyroid function test were normal. Magnetic resonance (MR) venography imaging was normal. In the cerebral–orbital MR imaging, the left cavernous sinus was wider than the right, there were many tortuous vascular structures at this level and contrast enhancement on the walls of the left cavernous sinus, the superior ophthalmic vein was dilated, and it was interpreted as CCF due to drainage toward the facial vein (Figure 2A, B). Cerebrospinal fluid (CSF) opening pressure was 580 mm H_2O, and CSF biochemistry and cytology were normal. In the digital subtraction angiography (DSA), there was a high-flow (A-V) fistula between the left external carotid artery (ECA), the meningeal media artery arising from the internal maxillary artery branch and the accessory meningeal artery and the cavernous sinus, as well as between the left (ICA), the meningeal branches at the level of the cavernous segment and the cavernous sinus. There was a 4x3 mm incidental

Figure 1. Chemosis and conjunctival hyperemia in the left eye at the time of the patient's admission

Address for Correspondence/Yazışma Adresi: Dr. Remzi Emre Şahin, Cukurova University Faculty of Medicine, Department of Neurology, Adana, Türkiye
Phone: +90 505 527 76 71 E-mail: dr.emresahin91@hotmail.com ORCID: orcid.org/0000-0003-0813-8752
Received/Geliş Tarihi: 30.08.2022 **Accepted/Kabul Tarihi:** 13.01.2023

saccular aneurysm in the left ICA paraophthalmic segment, and another high-flow A-V fistula between the internal maxillary branch of the right ECA and the cavernous sinus (Figure 3A, B). It was angiographically accepted as type-D CCF according to the Barrow classification due to the rapid filling of the cavernous sinus through the fistula and the involved vascular structures. Since the inferior petrosal sinus was closed and there was tortuosity in the superior ophthalmic vein, the fistula could not be reached with the DSA procedure, and endovascular treatment could not be performed. As medical treatment, acetazolamide (1.500 mg/day) was prescribed for high intracranial pressure and gabapentin (300 mg/day) was prescribed for pain. During the follow-up, a significant decrease in the headache and chemosis in the left eye were observed. The patient was discharged with outpatient follow-up planned; however, 1 month later, we learned that she had been hospitalized due to infection and had died.

WG is a systemic inflammatory disease characterized by vasculitis of small- and medium-sized vessels, resulting in idiopathic, systemic necrotizing granulomatous inflammation. Central nervous system involvement, peripheral polyneuropathy, cranial neuropathy (II, VI and VII), ischemic stroke, seizure, pachymeningitis, and cerebritis can be seen (1). CCF is an abnormal shunt from the carotid artery to the cavernous sinus (2). Spontaneous CCFs are generally idiopathic and constitute 20%–25% of all CCFs (2,3). Aneurysm rupture is known to be caused by hypertension or genetic conditions that predispose to vascular injuries such as Ehlers–Danlos syndrome or fibromuscular dysplasia. Type D CCF causes indirect fistulas. It is most commonly

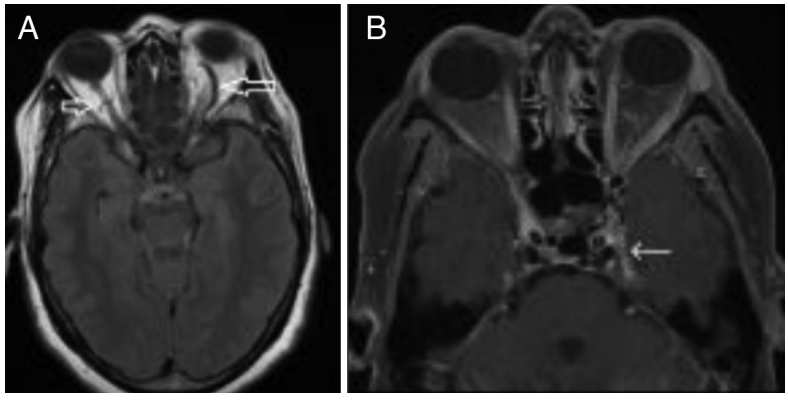

Figure 2. (A) Dilated superior vena ophthalmic, tortuous structure (white arrow), (B) contrast on the cavernous sinus wall (white arrow)

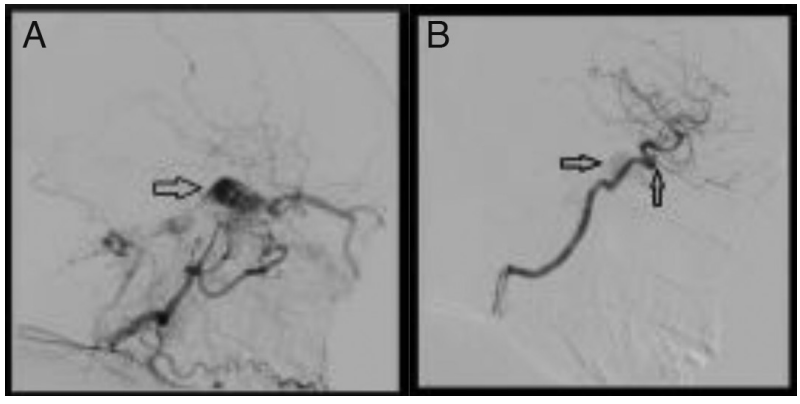

Figure 3. (A) High-flow A-V fistula between the left ECA, the meningeal media artery arising from the internal maxillary artery branch and the accessory meningeal artery, and the cavernous sinus (area shown with an arrow). (B) High-flow A-V fistula between the meningeal branches at the level of the left ICA cavernous segment and the cavernous sinus (horizontal arrow), as well as a 4x3 mm incidental saccular aneurysm (vertical arrow) in the left ICA paraophthalmic segment

ICA: Internal carotid artery, ECA: External carotid artery

Table 1. Barrow classification	
Type-A	Spontaneous or traumatic direct fistula between the ICA and cavernous sinus
Type-B	Dural shunt between the ICA meningeal branches and cavernous sinus
Type-C	Dural shunt between the ECA meningeal branches and cavernous sinus
Type-D	Dural shunt between both the ECA and ICA meningeal branches and cavernous sinus
ICA: Internal carotid artery, ECA: External carotid artery	

seen in older women (2,3). In indirect CCFs, IIH is detected due to increased cavernous sinus pressure (4). This rate has been reported as 8.7% in direct CCF and 3.6% in indirect CCF (5). Barrow type-D subtype generally includes low-flow, indirect CCFs (4). In traumatic CCF, there is a known history of trauma in this region. It is thought that there are congenital developmental defects in the vascular structures in congenital patients. The DSA method is the gold standard in diagnosis and classification of CCF (6). Barrow classification is used according to angiography findings (Table 1) (2,3,4,5,6), and today, endovascular treatments are at the forefront in the treatment of CCF (7). Standard practice is to close the fistula with a detachable balloon; however, it is not recommended in the presence of an aneurysm. The most important factor determining the success of the treatment is the fistula flow rate (3). The success rate of endovascular treatment is 89% (6).

Our patient was accepted as having hemodynamically high-flow, type D CCF, considering the CCF on both sides and the patient's age, and the indirect, spontaneous, angiographic, and clinical findings predisposed by WG and hypertension leading to endothelial damage in etiology. Although it is type-D according to Barrow classification, its high flow leads to suspicion of the presence of mixed types in these patients. Additionally, obesity and long-term corticosteroid use were thought to be facilitating factors for IIH detected in the patient. As a result, regardless of its severity, CCF in the presence of proptosis and chemosis, without a history of trauma, should be considered in the differential diagnosis and should be well managed in terms of complications.

Ethics

Informed Consent: Obtained.
Peer-review: Externally peer-reviewed.

Authorship Contributions

Concept: R.E.Ş., Ş.B., Design: R.E.Ş., K.A.K., Ş.B., Data Collection or Processing: R.E.Ş., K.A.K., H.B.O., Ş.B., Analysis or Interpretation: R.E.Ş., K.A.K., H.B.O., Ş.B., Literature Search: R.E.Ş., K.A.K., H.B.O., Ş.B., Writing: R.E.Ş.

Conflict of Interest: No conflict of interest was declared by the authors.

Financial Disclosure: The authors declared that this study received no financial support

References

1. Almouhawis HA, Leao JC, Fedele S, Porter SR. Wegener's granulomatosis: a review of clinical features and an update in diagnosis and treatment. J Oral Pathol Med 2013;42:507-516.
2. Docherty G, Eslami M, Jiang K, Barton JS. Bilateral carotid cavernous sinus fistula: a case report and review of the literature. J Neurol 2018;265:453-459.
3. Uysal F, Ceylan OM, Üstünsöz B, et al. Detachable balloon embolization in a patient with caroticocavernous fistula. Gulhane Med J 2006;48:56-58.
4. Kohli GS, Patel BC. Carotid Cavernous Fistula. In: StatPearls [Internet]. Treasure Island(FL):StatPearls Publishing; 2022 Jan-. Available from: https://www.ncbi.nlm.nih.gov/books/NBK535409/
5. Halbach VV, Hieshima GB, Higashida RT, Reicher M. Carotid cavernous fistulae: indications for urgent treatment. AJR Am J Roentgenol 1987;149:587-593.
6. Tran T, Maingard J, Amukotuwa S, Seneviratne U. Bilateral spontaneous carotid-cavernous fistulae. Pract Neurol 2021;21:261-263.
7. Lang M, Habboub G, Mullin JP, Rasmussen PA. A brief history of carotid-cavernous fistula. J Neurosurg 2017;126:1995-2001.