

Blood flow alterations in cavernous sinus dural arteriovenous fistula: Optical coherence tomography angiography findings

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ABSTRACT

Purpose: This case report details the diagnostic process for a patient with an initial diagnosis of scleritis who was unresponsive to typical treatment modalities, culminating in the identification of a cavernous sinus dural arteriovenous fistula (CS-DAVF). The case highlights the role of anterior segment optical coherence tomography angiography (OCTA) in the diagnosis of this vascular anomaly and in monitoring the response to treatment.
Observations: A 45-year-old man with persistently elevated intraocular pressure (IOP) and ocular congestion in the left eye was unresponsive to treatment for scleritis. The persistent ocular symptoms and new-onset tinnitus prompted further investigation. Anterior segment OCTA revealed vascular anomalies, and magnetic resonance imaging confirmed a CS-DAVF. The patient underwent endovascular treatment for the CS-DAVF. This intervention led to a significant reduction in IOP in the left eye and the resolution of ocular congestion.
Conclusions and importance: This case highlights the diagnostic complexities of ophthalmic symptoms that mimic those of other conditions. Furthermore, it demonstrates the essential role of anterior segment OCTA in the accurate diagnosis and effective management of CS-DAVF and highlights the need for comprehensive diagnostic approaches in ophthalmology.

1. Introduction

Dural arteriovenous fistulas (DAVFs) are anomalous vascular connections between the arterial and venous systems within the dura mater, the protective layer surrounding the brain and spinal cord.^{1–5} These fistulas can lead to a wide spectrum of clinical manifestations. Neurologically, they present with symptoms ranging from headaches to seizures due to cerebral infarction or hemorrhage. However, the manifestation of symptoms largely depends on the anatomical location of the fistula and affected vascular structures. Particularly, cavernous sinus (CS) DAVFs present with ophthalmological symptoms, such as elevated intraocular pressure (IOP), diplopia, exophthalmos, vascular engorgement, changes in visual acuity, and, in severe cases, vision loss. Although the symptoms of CS-DAVFs are often distinct, sometimes, they can mimic other ophthalmic conditions such as scleritis, an inflammatory condition characterized by redness, pain, and potential vision

impairment.^{6–8} This symptom overlap between CS-DAVFs and scleritis is challenging for clinicians, and potentially delays accurate diagnosis and appropriate treatment.

In this report, we highlight a unique case of CS-DAVF wherein optical coherence tomography angiography (OCTA) was instrumental in diagnosing and delineating the ocular pathology and monitoring symptomatology throughout the course of neurosurgical intervention.

2. Case report

A 45-year-old man presented to a community ophthalmology clinic with the primary complaint of vascular engorgement in his left eye. The patient had a significant history of retinopathy of prematurity in both eyes, but no history of trauma, hypertension, diabetes, or other cardiovascular risk factors. After clinical evaluation, the patient was diagnosed with left-eye scleritis. Treatment was initiated with both 1 % topical

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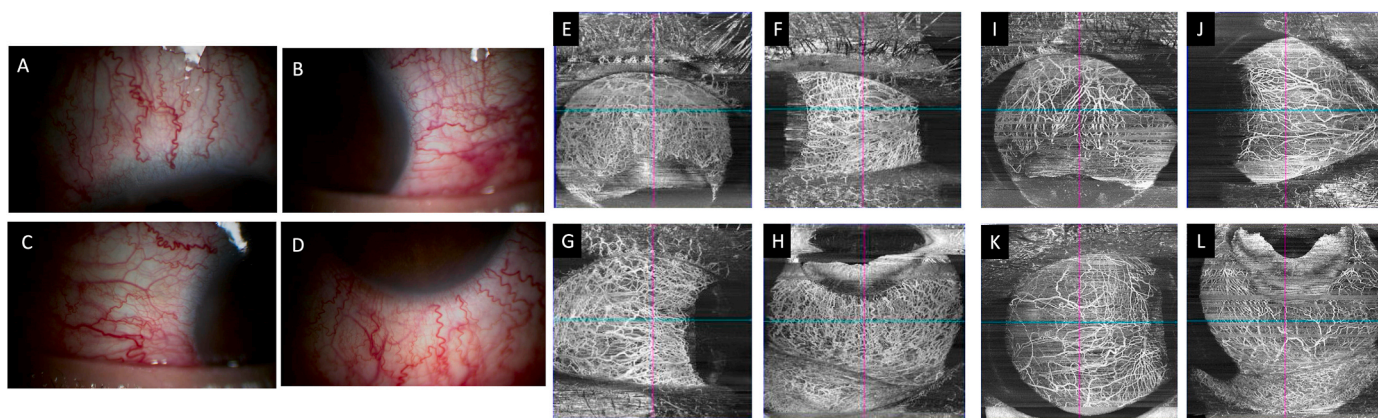


Fig. 1. Slit-lamp image of the left eye and anterior segment optical coherence tomography angiography (OCTA) images of both eyes at the initial consultation. Slit-lamp examination revealed vascular engorgement in the left eye captured from multiple anatomical viewpoints: superior (A), temporal (B), nasal (C), and inferior (D). Concurrently, anterior segment optical coherence tomography angiography (OCTA) revealed dilatation of the episcleral vessels in the left eye, as evidenced by various orientations: superior (E), temporal (F), nasal (G), and inferior (H). In contrast, the OCTA images of the right eye (I–L) do not exhibit such findings, indicating the absence of similar vessel dilatation in the right eye.

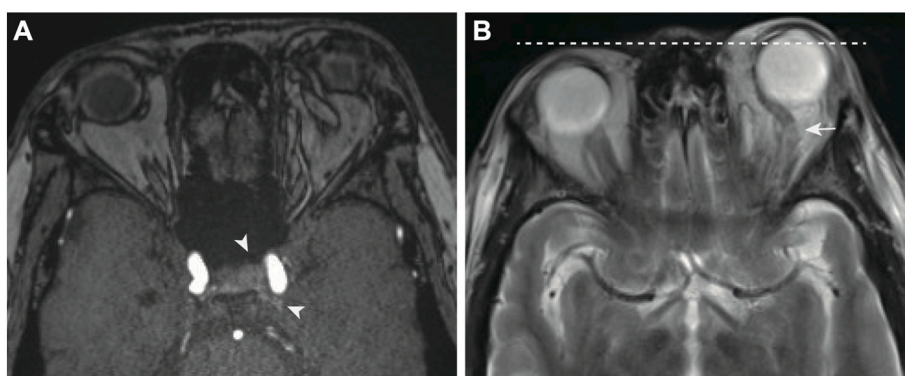


Fig. 2. Contrast-enhanced Magnetic Resonance Imaging (MRI) when the patient experienced tinnitus after consultation. A: MR angiography revealed abnormal vascular flow around the left internal carotid artery (arrowhead). B: T2 weighted imaging showed the enlarged left superior ophthalmic vein (arrow) and left exophthalmos (dotted line). These findings are consistent with a cavernous sinus dural arteriovenous fistula (CS-DAVF).

prednisolone acetate eye drops, administered every 2 h, and 30 mg oral prednisone daily. Despite adhering to treatment for two months, the symptoms showed no improvement, and the IOP in the left eye consistently ranged from 25 to 30 mmHg. Thus, two months after the initial

consultation at the previous clinic, the patient was referred to our institution for further evaluation and management.

Upon initial evaluation at our institution, his left eye continued to show elevated IOP and persistent vascular engorgement, with only slight

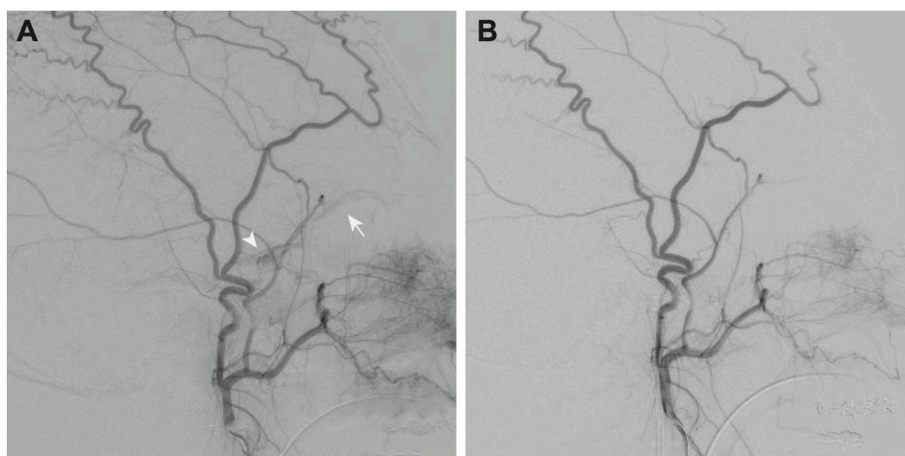


Fig. 3. Pre- and postoperative digital subtraction angiography. A: Preoperative left external carotid angiograph (lateral projection) showing an arteriovenous shunt in the left cavernous sinus (arrowhead). Flow drains into the left superior ophthalmic vein (arrow). B: Postoperative left external carotid angiography (lateral projection) reveals that the arteriovenous shunt has disappeared after shunt occlusion by the coils.

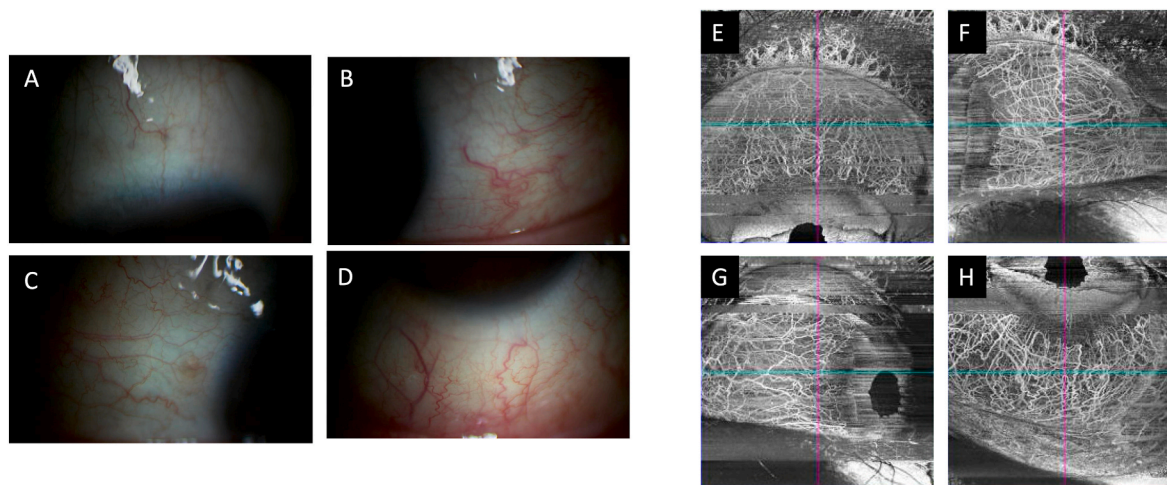


Fig. 4. Progress of eye condition at one month after treatment. Slit-lamp examination demonstrated amelioration of vascular engorgement in the left eye, as observed from various angles: superior (A), temporal (B), nasal (C), and inferior (D). Furthermore, anterior segment optical coherence tomography angiography (OCTA) of the same eye revealed a reduction in the dilation of episcleral vessels and decreased vascular density compared to the pretreatment findings, also captured from multiple perspectives: superior (E), temporal (F), nasal (G), and inferior (H).

pain and discomfort in his left eye. Additionally, he presented with proptosis and ptosis of the left eye. The best-corrected visual acuity (BCVA) in both eyes was 20/20 and the IOP measurements were 13 and 28 mmHg in the right and left eye, respectively. Anterior segment examination with a slit lamp revealed vascular engorgement in the left eye. Moreover, OCTA identified dilated scleral vessels in the same eye (Fig. 1). However, fundoscopic examination of both eyes revealed no retinal vascular dilation. The observation of dilated scleral vessels exhibiting a distinct short-pitched serpiginous pattern suggested the possibility of intracranial shunt diseases rather than simple scleritis. In light of the unilateral presentation, and to rule out a viral etiology of scleritis, a polymerase chain reaction test was performed on the aqueous humor from the anterior chamber of the left eye. This test specifically targeted herpes simplex virus, varicella zoster virus, and cytomegalovirus and returned negative results. While continuing the original treatment, oral non-steroidal anti-inflammatory drugs were added to his treatment regimen. However, similar to the prior treatments, there was no improvement in the condition of his left eye.

During this period, the patient experienced tinnitus. Although initially perceived as unrelated to the ocular symptoms, the continuing auditory issue prompted a consultation at an internal medicine clinic a month after referral to our institution. Subsequent contrast-enhanced head magnetic resonance imaging (MRI) and angiography (MRA) confirmed the diagnosis of CS-DAVF (Fig. 2).

Following the diagnosis, the patient was promptly referred to our neurosurgery department. After endovascular treatment using coils that successfully occluded the fistula (Fig. 3), there was a notable reduction in IOP and resolution of vascular engorgement in the left eye (Fig. 4). One month after the intervention, the IOP of the left eye normalized to 13 mmHg, and the patient reported cessation of tinnitus episodes. Additionally, proptosis and ptosis in the left eye improved, and no ocular motility disorders were observed.

3. Discussion

This case suggests the potential utility of anterior segment OCTA in diagnosing and assessing therapeutic outcomes in patients with symptoms associated with CS-DAVF. Although the initial symptoms of our patient were suggestive of scleritis, the unchanged nature of the ocular symptoms, even after standard corticosteroid treatment, raised questions regarding the underlying pathology.

Anterior segment OCTA of the affected eye provided substantial

observations. The presence of dilated scleral vessels with a distinct short-pitched serpiginous pattern suggested a vascular anomaly that diverges from a typical inflammatory reaction. While traditional slit lamp examinations offer limited insights, anterior segment OCTA provides details of the extent of congestion and stasis, potentially explaining the elevated IOP observed. Furthermore, our observations align with existing literature that highlights the utility of anterior segment OCTA in evaluating patients with glaucoma; our application in this context is distinct.⁹

MRI and MRA are instrumental in refining the diagnosis of intracranial shunt diseases, such as CS-DAVF. Thus, given that CS-DAVFs can mirror various ophthalmological symptoms, the inclusion of anterior-segment OCTA in the diagnostic toolkit may offer additional clarity. Moreover, the role of persistent tinnitus in our diagnostic journey is noteworthy. Initially overlooked, it became increasingly relevant given its chronic nature with concurrently elevated IOP, emphasizing the need for a holistic approach for such diagnostic challenges.

Previously, the use of an anterior segment OCT B-scan has demonstrated value in monitoring scleral thickness during scleritis treatment evaluations.¹⁰ Similarly, the utility of anterior segment OCTA has been highlighted in the diagnosis of pupillary membrane remnants.¹¹ However, to the best of our knowledge, our report is among the first to use anterior segment OCTA to assess the pathology and treatment efficacy of CS-DAVF, marking a potential advancement in this domain.

In conclusion, although further research is warranted, our findings hint at the potential of anterior segment OCTA for evaluating conditions akin to intracranial shunt disease and scleritis. We believe that such tools, when employed judiciously, can equip clinicians with nuanced insights, enrich diagnostic processes, and potentially guide tailored therapeutic decisions.

4. Patient consent

Written informed consent was obtained from the parent of the patient for publication of the details of their medical case and any accompanying images.

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Declaration of competing interest

The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

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