

PAINFUL FOURTH CRANIAL NERVE PALSY CAUSED BY POSTERIORLY-DRAINING DURAL CAROTID-CAVERNOUS SINUS FISTULA

Rong-Kung Tsai, Hsin-Yi Chen, and Hwei-Zu Wang

Abstract: A 65-year-old man with a dural carotid-cavernous fistula (DCCF) presented with sudden onset of painful trochlear nerve paresis. Typical signs of DCCF including conjunctival arterialization, chemosis, and proptosis did not become manifest until 4 months later. This unusual presentation of DCCF was caused by drainage of the fistula posteriorly into the inferior petrosal sinus with low flow. With this condition, patients may present with trochlear nerve palsy without a red eye. Although rare, DCCF must be considered in patients presenting with isolated painful trochlear palsy.

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Key words:
dural carotid-cavernous fistula
trochlear nerve palsy
white-eyed shunt

Dural carotid-cavernous fistula (DCCF) usually presents with conjunctival injection, chemosis, proptosis, pulsatile tinnitus, diplopia, and headache [1-3]. Sanders and Hoyt found the following presenting symptoms: bruit (75%), proptosis (69%), redness and swelling of conjunctiva (36%), diplopia (24%), ipsilateral blurred vision (16%), and orbital pain (16%) [4]. Because dilatation of the conjunctival vein is absent or mild in one-third of cases [3], the condition is sometimes initially misdiagnosed as mononeuropathy in patients who present with isolated oculomotor or trochlear nerve palsy [2, 4-6]. The abducens nerve is involved in about half of patients who have extraocular motor dysfunction secondary to carotid-cavernous sinus fistula [7]; oculomotor and trochlear nerve palsies occur less often. The diagnosis of DCCF would not ordinarily be considered in the absence of external ocular signs.

Here, we describe a patient with DCCF, who presented with isolated trochlear nerve palsy without classical external ocular findings initially. The angiographic characteristics of this disorder are presented, along with a discussion of the apparent mechanism based on a review of the literature.

Case Report

A 65-year-old man had sudden onset of binocular vertical diplopia and right orbitofrontal pain. He had no history of other neurologic complaints or any significant head trauma. He suffered from adult diabetes mellitus without retinopathy, and had no hypertension history. Best corrected visual acuity was 20/30 in each eye. The other pertinent findings were absence of ocular congestion, proptosis, and ptosis. Bielschowsky/Parks' Three-Step Test showed 6 prism diopters (D) of right hypertropia in the primary position, 8 D right hypertropia on left gaze, orthophoria on right gaze, 10 D right hypertropia on right head tilt, and orthophoria on left head tilt. These results were compatible with the diagnosis of right superior oblique paresis. Corneal and facial sensation were normal and no bruit was heard subjectively or objectively. On examination, moderate lens opacity was found in both eyes and ocular funduscopy findings appeared normal in each eye.

Department of Ophthalmology, Kaohsiung Medical University, Kaohsiung.

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Reprint requests and correspondence to: Dr. Hwei-Zu Wang, Department of Ophthalmology, Kaohsiung Medical University, 100 Shih-Chuan 1st Road, Kaohsiung, Taiwan.

Right fourth-nerve palsy caused by diabetes-related ischemia was suspected, and a daily dose of 30 mg prednisolone was given orally for 1 week with gradual tapering. The headache improved but the fourth-nerve palsy persisted. Therefore, supportive treatment with regular follow-up was suggested.

Four months later, he suddenly developed conjunctival congestion with mild proptosis in the right eye, and the vertical diplopia disappeared at the same time. On ophthalmologic examination, he had mild limitation of extraocular motility in the upward direction and abduction of the right eye, but orthophoria in the primary position. These limitations of extraocular motility were probably manifestations of restriction myopathies caused by DCCF-induced orbital congestion. A bruit could not be auscultated over the eyes or the head.

Contrast-enhanced high-resolution computed tomography (CT) of the orbits showed marked dilatation of the right superior ophthalmic vein (Fig. 1). An intra-arterial digital subtraction angiogram revealed a right DCCF fed by meningeal branches of the right internal carotid artery with flow mainly directed posteriorly into the inferior petrosal sinus, with partial reflux anteriorly into the right superior ophthalmic vein (Fig. 2). The patient received angiographic embolization treatment and the signs of red eye and diplopia improved 1 month later.

Discussion

Anatomically, carotid–cavernous fistulas can be classified by anatomy into direct and dural types. The direct type is characterized by a direct connection between the cavernous sinus segment of the internal carotid artery and the cavernous sinus. These fistulas usually have high flow, most often caused by a single tear in the arterial wall, and are, thus, called direct

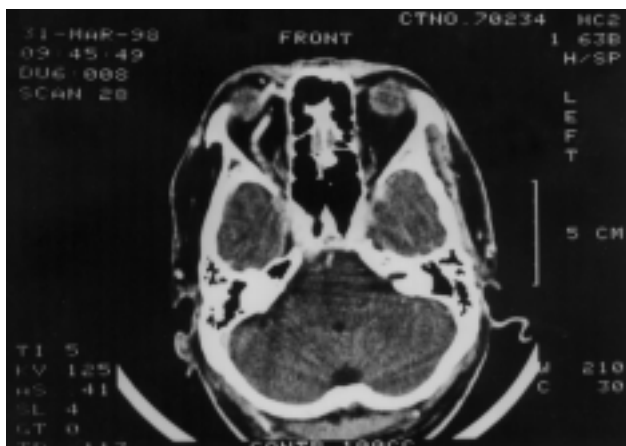


Fig. 1. Computed tomography scan at the time of red eye reveals an engorged right superior ophthalmic vein.

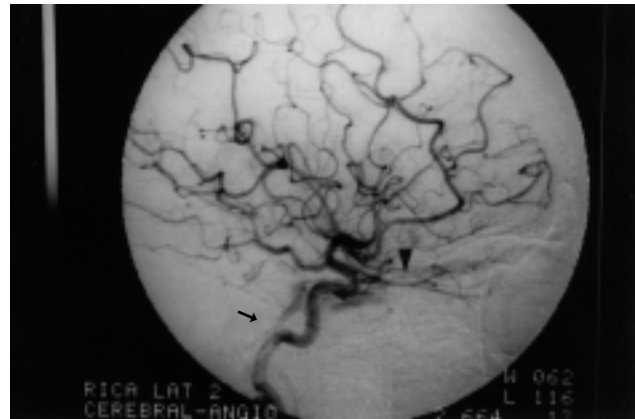


Fig. 2. Right lateral intra-arterial digital subtraction angiogram reveals a prominent cavernous sinus fed by dural branches of the right internal carotid artery with flow mainly directed into the inferior petrosal sinus (arrowhead), and with partial reflux to the right superior ophthalmic vein (arrow).

carotid–cavernous sinus fistulas. DCCF consists of a communication between the cavernous sinus and one or more meningeal branches of the internal carotid artery or external carotid artery. These fistulas usually have low arterial flow and develop spontaneously [1].

Although DCCF should be included in the differential diagnosis of painful ophthalmoplegia, diagnoses are usually made based on clinical presentations of external ocular signs [1–5, 7–9]. In most instances, older patients present with isolated oculomotor, trochlear, or abducens nerve palsies without proptosis or conjunctival congestion, and the condition is, therefore, easily misdiagnosed initially as mononeuropathy of ischemic origin.

DCCF is formed when thin-walled dural vessels rupture into the cavernous sinus. The venous drainage of the cavernous sinus can be either anterior or posterior. Anteriorly and superiorly, the cavernous sinus drains via the superior ophthalmic vein into the angular vein, then into the facial vein. Anteriorly and inferiorly, drainage is from the inferior ophthalmic vein into the pterygoid plexus and then into the facial vein [2]. Posteriorly, the cavernous sinus drains into the transverse sinus via the superior petrosal sinus; into the jugular vein via the inferior petrosal sinus; and into the pterygoid plexuses via several small emissary veins [2].

If the shunt drains anteriorly into the superior ophthalmic vein, red eye, chemosis, proptosis, and ophthalmoplegia may occur. Phelps et al used the term “red-eyed shunt syndrome” to describe this condition [6]. These signs are secondary to elevation of venous pressure [1–9]. Posteriorly-draining dural shunts are less frequent, and in such situations, the congestive features will be absent and the diagnosis of “white-eyed shunt” will be missed unless angiography is performed [10].

It is clear that dural–cavernous sinus shunts produce symptoms that are dependent on the direction of drainage from the shunts; the direction of drainage can change [11], as in the present case. McKinna reported that all of his 63 patients with carotid–cavernous sinus fistulas had proptosis

and vascular congestion of the globes and orbits [12]. The exception to this is the dural shunt, in which a low-flow shunt develops between dural arteries and the cavernous sinus, when external signs may be less conspicuous [12].

Acierno et al reviewed the white-eyed shunt cases in the literature; including their two, there were a total of 30 reported cases [10]. Headache and diplopia were the principal symptoms. All three oculomotor nerves were affected with various incidences, including 14 third-nerve palsies, 13 sixth-nerve palsies, and three fourth-nerve palsies [2, 5, 6]. In 10 of these 30 cases, the eye eventually turned red after weeks to months (delayed red-eyed shunts), as in the present case at 4 months, suggesting that fistular drainage had shifted anteriorly. Interestingly, a bruit was described in only four cases [10].

The three previously reported cases and the present case of dural fistulas with painful fourth-nerve palsy included three men and one woman, aged between 40 and 65 years [2, 5, 6]. All patients reported pain around the involved eye, or headache. A bruit was found in only one case [5]. Three patients had delayed red-eyed shunt. The CT scans and angiographs, available for three patients, showed engorged superior ophthalmic veins in two patients. One had exclusively posterior drainage in the angiogram, while the other two had a combination of anterior and posterior drainage. Similar to the case reported by Phelps et al [6], the fourth-nerve palsy in our patient disappeared when delayed red-eyed shunt occurred. A possible explanation is that the change of the drainage flow to the anterior direction may have relieved a certain degree of venous pressure in the cavernous sinus and inferior petrosal sinus, which compressed the trochlear nerve when the fistula drained exclusively posteriorly.

The pathogenesis of oculomotor nerve palsy in DCCF may be due to nerve compression by an expanding sinus, or ischemic neuropathy secondary to venous congestion or arterial steal [8]. It has been proposed that distension of the inferior petrosal sinus compresses the sixth cranial nerve in Dorello's canal [1]. It remains unclear why particular oculomotor nerves are affected, and angiographic findings from this case and previous reports provide no reasonable answers.

The size and shape of the cavernous sinus was different in each patient. Inoue et al classified the most common shapes of the cavernous sinus on lateral projection as anterior-inferior, intermediate, and posterior-superior dominant types [13]. Oculomotor nerve palsy in DCCF is most likely caused by a combination of several factors, including an individual's anatomic variance [14].

Among patients who have fourth-nerve palsy and headache, the diagnosis of white-eyed shunt is likely to be delayed because cerebral angiography is not part of the initial evaluation of this condition [10]. The reasonable differential diagnosis includes diabetic ophthalmoplegia, Tolosa-Hunt syndrome, head injury, and intracavernous aneurysm. The initial fourth-nerve palsy in our patient might not have been of diabetic origin, since there was no improvement after 4 months of follow-up. As for the later-developing limitations of extraocular motility at the time of red-eye shunt, since there

was orthophoria in the primary position, the defects were probably caused by mechanical restrictions of orbital congestion rather than by oculomotor nerve palsy.

To minimize the chance of missing the diagnosis of DCCF, auscultation and palpation of the orbit and cranium should be done in cases of painful ophthalmoplegia [2], though the incidence of bruit among patients with white-eyed shunt is low. As others have emphasized [10], a posteriorly-draining fistula should be considered prominently as a cause of painful oculomotor palsies that do not remit after 3 to 6 months, and magnetic resonance angiography or cerebral angiography should be used to aid diagnosis. Careful follow-up is essential to make an early diagnosis.

In conclusion, we have described a case of unilateral fourth-nerve palsy caused by DCCF. The final cause was not found until delayed red-eye shunt occurred. Although rare, DCCF should be considered in the differential diagnosis of painful ophthalmoplegia.

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