

Ophthalmic Manifestations of Intracranial Dural Arteriovenous Fistula — Report of Four Cases

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ABSTRACT

Objective: Intracranial dural arteriovenous fistulas (AVFs) account for 10%-15% of all intracranial arteriovenous lesions. Some dural AVFs produce ocular symptoms include proptosis, diplopia, episcleral venous engorgement, periorbital swelling, extraocular muscle limitation, visual field defect, and papillaedema. **Materials and Methods:** We reviewed four patients who had dural AVFs with ophthalmic manifestations in the past three years (2000 November to 2003 October.) The diagnosis of dural AVF was confirmed by image studies, such as magnetic resonance imaging (MRI), magnetic resonance angiography (MRA) and cerebral angiography. **Results:** There were three men and one woman. According to Cognard's classification, there were four types: type I, type II a, type II a+b, and type III. Symptoms at initial presentation included headache, diplopia, proptosis, episcleral vein engorgement, periorbital swelling, visual field defect, and papillaedema. Three patients received transarterial embolization (TAE) in our hospital. Improvement in ophthalmic symptoms and signs was noted after TAE treatment, although multiple interventions were required.

Conclusions: Dural AVFs can produce various ocular symptoms and signs at their initial manifestation. The symptoms are believed to reflect venous hypertension in the superior sagittal sinus, resulting from the shunted flow, which interferes with normal venous drainage. Transarterial embolization can be an effective treatment to close the fistula, restore sinus function and improve ocular symptoms. (*Tzu Chi Med J* 2005; 17:93-97)

Key words: dural arteriovenous fistula, transarterial embolization, episcleral vein engorgement

INTRODUCTION

Intracranial dural arteriovenous fistulas (AVFs) account for 10%-15% of all intracranial arteriovenous lesions [1]. The clinical presentation of dural AVFs is related to the pattern of venous drainage. Their symptoms and signs are highly variable. Some dural AVFs may produce tinnitus or ocular symptoms; others are associated with neurological symptoms or even intracranial hemorrhage [2,3]. Dural AVFs are located within the wall of the venous structure and most commonly involve the transverse, sigmoid, and cavernous sinuses. Other locations include the tentorial incisura, superior sagittal sinus, anterior cranial fossa, and superior petrosal sinus

[3]. Dural AVFs have long been regarded as benign lesions compared with other brain arteriovenous malformations. However, catastrophic complications such as intracranial hemorrhage can occur [1-3]. Different patterns of AVF venous drainage have been described, and intracranial dural AVFs can behave aggressively depending on this pattern [3]. Clinical ophthalmic symptoms and signs of dural AVFs are rarely reported but may include proptosis, diplopia, engorged episcleral vessels, periorbital swelling, extraocular muscle limitation, visual field defect, and papillaedema [2-9]. These symptoms and signs usually lead the patient to visit an eye care practitioner.

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MATERIALS AND METHODS

Patients who had dural AVFs with ophthalmic manifestations in the past three years (2000 November to 2003 October) were studied. The diagnosis of dural AVF was confirmed by image studies, such as MRI, MRA and cerebral angiography. The basic patient profiles and initial presentations are summarized in the Table 1. The classification of dural AVFs was according to Cognard's classification which was proposed in 1995 [2]. In the type I, the dural AVFs was located in the main sinus with antegrade flow. In the type II, the dural AVFs was located in the main sinus, with reflux into the sinus (IIa), cortical veins (IIb), or both (IIa + b). In the type III, the dural AVFs was located in the main sinus with direct cortical venous drainage without venous ectasia. In the type IV, the dural AVFs was located in the main sinus with direct cortical venous drainage with venous ectasia.

CASE REPORTS

Patient 1

A healthy 31-year-old man was referred with a 1 month history of diplopia and blurred vision. The diplopia was obvious with right gaze. In addition, the patient reported a dull headache. His best corrected visual acuity was 20/20 in both eyes. Intraocular pressure was 24 mmHg in both eyes. The pupil diameter was 3 mm in both eyes and no relative afferent pupillary defect (RAPD) sign was noted. The ocular mobility was limited at right lateral gaze. A slit-lamp examination appeared normal in the anterior segment. Fundus examination revealed disc swelling without spontaneous venous pulsation in both eyes (Fig. 1). A mildly enlarged blind spot was noted on visual field examination. MRI study showed a prominent venous system at the bilateral basal ganglia and thalamus. Further cerebral angiography demonstrated intracranial bilateral dural AVFs supplied from the superior temporal arteries and occipital arteries. The venous drainage was from the superior sagittal sinus. No retrograde venous flow or sinus thrombosis was found. The dural AVF of this patient was classified as type I according to Cognard's classification because it was located in the main sinus with antegrade flow only. His ocular symptoms subsided gradually after angiography was performed.

Patient 2

A 45-year-old man suffered from left eye redness for one month. He was treated for conjunctivitis but topi-

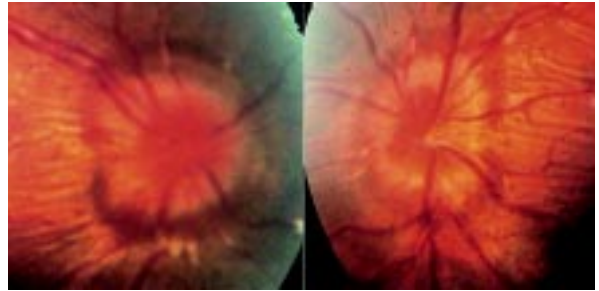


Fig. 1. Disc photography of patient 1. The discs in both eyes are swollen without spontaneous pulsation.

cal treatment had no effect. In addition, he also complained of headache for one month and consulted a neurologist. Visual acuity, visual fields, pupillary responses, and ophthalmoscopic examinations were all normal. The ocular tension was 10 mmHg in the right eye and 15 mmHg in the left eye. Mild proptosis was noted. The Hertel exophthalmometer showed 4 mm more proptosis in the left eye than in the right eye. Episcleral vein engorgement in the left eye was also noted. Head MRI demonstrated venous thrombosis of the superior sagittal sinus and a recent infarct at the right periventricular area. A dural AVF at the occipital region with retrograde venous drainage into the superior sagittal sinus was found on cerebral angiography. The dural AVF of this patient was classified as Cognard type IIa, because it was located in the main sinus with reflux into the sinus. Transarterial embolization (TAE) for dural AVF was performed twice. Episcleral vein engorgement and proptosis of the left eye significantly improved after TAE treatment.

Patient 3

An otherwise healthy 44-year-old woman suffered from progressive left eye proptosis, periorbital swelling and redness for 2 months. She was treated for episcleritis. She also had a headache over the bilateral temporal area and an annoying audible bruit. Visual acuity, visual fields, pupillary response, and ophthalmoscopic examinations were normal. Ocular tension was 10 mmHg in the right eye and 12 mmHg in the left eye. The Hertel exophthalmometer showed 2 mm more proptosis in the left eye than in the right eye. Two prominent left upper eyelid subcutaneous vessels and episcleral vein engorgement were found (Fig. 2). Head MRI demonstrated abnormal vasculature and shunting at the bilateral lateral sinus. A prominent right middle meningeal artery and bilateral occipital arteries were noted. A major dural AVF located at the left transverse sinus was noted on cerebral angiography (Fig. 3). The feeders include the left



Fig. 2. Proptosis, two prominent subcutaneous vessels in the left upper eyelid, and episcleral vein engorgement are noted in patient 3.

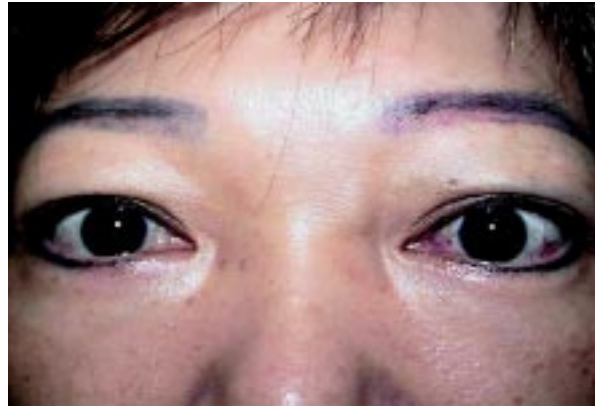


Fig. 4. Appearance of patient 3 after treatment. Episcleral vein engorgement and proptosis of the left eye have improved.

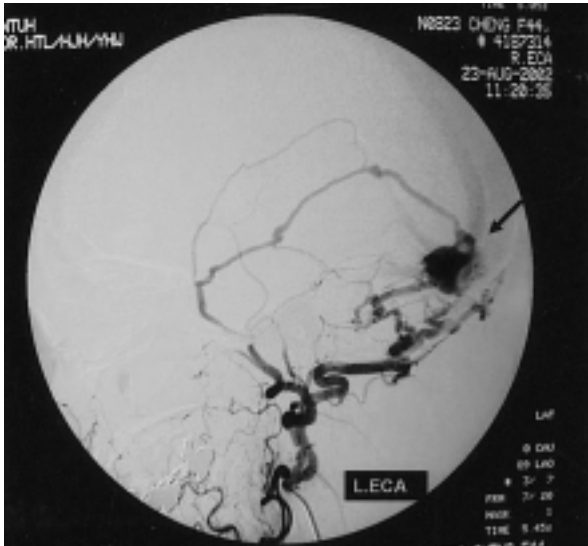


Fig. 3. Major dural AVF was located at the left transverse sinus supplied from the meningeal branches of external carotid artery in patient 3.

middle meningeal artery, dural branches of the bilateral occipital arteries, and dural branches of the left vertebral artery. The dural AVF of this patient was classified as Cognard type IIa+b, because it was located in the main sinus with reflux into the sinus and the cortical veins. Five TAE procedures were performed to close her dural AVF. Her proptosis, periorbital swelling and left eye episcleral vein engorgement markedly subsided after TAE treatment (Fig. 4).

Patient 4

This 43-year-old man suffered from a severe headache with visual field defect for two days. He was sent to the emergency department for help. Visual acuity, pu-

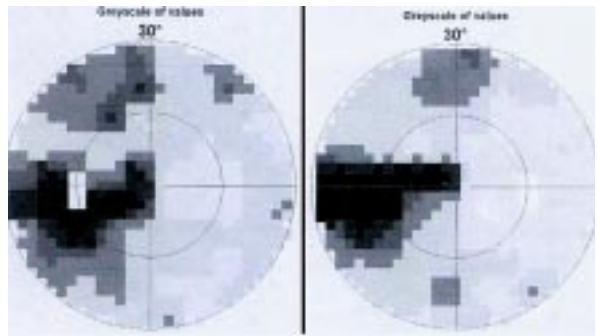


Fig. 5. The Octopus visual field of patient 4 three months after TAE. It demonstrated left incomplete homonymous hemianopia.

pillary responses, and ophthalmoscopic examinations were all normal. A confrontation visual field test at the bedside revealed left complete homonymous hemianopia. CT scan showed right occipital intracranial hemorrhage. Cerebral angiography revealed a dural AVF in the right occipital region supplied from the meningeal branch of the right external carotid artery with direct cortical venous drainage into the superior sagittal sinus without ectasia. The dural AVF of this patient was classified as Cognard type III. Transarterial embolization (TAE) for dural AVF was performed to prevent further intracranial hemorrhage. His visual field defect had resolved partially 3 months after TAE treatment (Fig. 5).

DISCUSSION

The dural AVF is composed of a nidus of abnormal arteries and veins with arteriovenous shunting contained

Table 1. Summary of Clinical Manifestations of Four Cases

	Age	Sex	Initial presentation	Cognard type
Case 1	31	M	diplopia, headache, papilledema, elevated intraocular pressure	I
Case 2	45	M	Episcleral vein engorgement, proptosis, headache	IIa
Case 3	44	F	Proptosis, periorbital swelling, episcleral vein engorgement, headache	IIa+b
Case 4	43	M	Visual field defect, headache	III

entirely within the sinus of the dura [10]. Controversy still persists concerning the pathogenesis of dural AVFs. One theory says that thrombophlebitis of a dural sinus is the initiating factor in acquired AVFs. However, in the younger population, large direct shunts are observed and well tolerated, and are thought to be of congenital origin [3]. Dural AVFs can exist in any sinus in the dura of the intracranial cavity.

The clinical presentations are related to the location of the fistula and the pattern of venous drainage [11]. Reported manifestations include audible bruit, headache, nonhemorrhagic neurological deficit, medically intractable seizures, progressive neurological deficit, and cerebellar symptoms [4-7]. Dural AVFs with retrograde cortical venous drainage carry a high risk of intra-cerebral hemorrhage [2,6,10,11]. Drainage through the ophthalmic vein may cause ocular symptoms such as engorged episcleral vessels, chemosis, proptosis, secondary glaucoma, diplopia, periorbital swelling, extraocular muscle limitation, and papillaedema [2-9, 11]. Ancillary diagnostic exams include orbital ultrasonography, computed tomography (CT), MRI, MRA, and cerebral angiography.

A carotid cavernous sinus fistula (CCSF) may be a direct or indirect arteriovenous shunt between the mostly internal carotid artery or its meningeal branches and the cavernous sinus. The direct type CCSF is often associated with a history of trauma and rapid onset of symptoms due to the high blood flow rate. The ocular symptoms of dural AVF outside the cavernous sinus may be similar to those of the indirect type CCSF [9]. A dural AVF outside the cavernous sinus consists of multiple arteriovenous shunts between meningeal arteries mostly from the external carotid artery and a dural venous sinus or a meningeal vein. The severity of the ophthalmic symptoms might be related to the location and duration of intracranial dural AVF [6,9,10]. In general, the more posterior the fistula, the fewer the ocular symptoms noted [9-11]. However, Liu et al reported that posterior cranial fossa arteriovenous fistulas (e.g. vertebral AVFs) presenting with exophthalmus, chemosis, and eyelid swelling resulting from retrograde venous drainage into the cavernous sinus and superior ophthalmic vein might

mimic the indirect type CCSF [12].

In one study, all patients with dural AVFs were classified according to two grading scales: the more descriptive schema of Cognard et al [2] and that recently proposed by Borden et al [6]. A correlation of clinical outcome was found between the Cognard and Borden classifications. A higher grade was correlated with a poor prognosis due to existing reflux into the sinus and venous ectasia [5].

Although Luciani et al [13] reported 3 patients with spontaneous closure of dural AVFs, intervention is needed if the symptoms persist or endanger the patient's life. The treatment modalities include transarterial or transvenous endovascular techniques and surgery [14, 15]. Stereotactic gamma knife radiosurgery has been reported as an alternative to microneurosurgical removal of selected intracranial vascular malformations [16].

In our four patients, we believed the symptoms were related to the venous hypertension in the dural sinus, resulting from the shunted flow which interfered with normal venous drainage. In patient 1, right abducens palsy may have been due to increased intracranial pressure related to venous hypertension. His symptoms improved after cerebral angiography. The disappearance of symptoms might be explained by spontaneous closure after cerebral angiography [13]. We assumed that cerebral angiography might relieve the symptoms of patients with minor type fistulas (mostly type I). In patients 2 and 3, proptosis, episcleral vein engorgement, and headache resulted from venous hypertension in the dural sinus. They received therapeutic transarterial embolization for relief of symptoms and signs and prevention of intracranial hemorrhage. In patient 4, left homonymous hemianopia resulted from right occipital hemorrhage of a dural AVF. Therapeutic TAE was performed and his visual field defect resolved after treatment.

In conclusion, early diagnosis of dural AVF is difficult but careful analysis of ocular symptoms and image studies can lead to the correct diagnosis. MRA/MRI and cerebral angiography are necessary for clinical diagnosis and even treatment. Transarterial embolization can be an effective treatment to close the fistula, restore

sinus function and improve ocular symptoms.

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