Comment

It is important to recognise the association of blepharospasm and eyelid apraxia because the implications for successful treatment of both conditions may vary when they coexist in the same patient.[1,2] Although they are both preceded by an increased frequency of blinking, patients with blepharospasm have forceful orbicularis spasms following which the lids spontaneously open. However, in patients with apraxia, the lids will not open even in the absence of any orbicularis spasm; instead, the brows are elevated secondary to frontalis overaction.[2] Although patients with apraxia of eye opening often complain of involuntary lid closure, the problem is actually in overcoming the levator inhibition. It is a transient phenomenon, which can cause backward thrusting of the head to interrupt the inhibition of levator muscles, following which lids open suddenly.[4]

Anderson *et al*[1] have proposed a defective circuit theory to explain the pathogenesis of blepharospasm: an afferent arm with impulses travelling via the trigeminal nerve, a control centre in or near the basal ganglia, and an efferent arm via the facial nerve to the eyelid protractors (orbicularis, corrugator superciliaris, and procerus). Treatment can be aimed at all arms of the circuit, but is predominantly directed at the efferent arm.

The two most successful treatment options for blepharospasm are BTA and myectomy of the eyelid protractor muscles.

BTA is effective in up to 86% of patients with blepharospasm, but the effect lasts less than 4 weeks in 13% of responders.[1] It does not treat the eyelid apraxia component when this is present. Approximately 50% of patients with blepharospasm and eyelid apraxia show no therapeutic response to BTA.[1] The reason for this is unknown, but may be related to immune-mediated phenomena.[5,6] Myectomy has been reported to be successful in relieving symptoms in 88% of these cases, but more than one operation is sometimes necessary.[1,2]

Hirayama *et al*[7] have reported the beneficial effects of goggles in patients with apraxia of the lids. Eyelid opening was improved in two patients with apraxia from Parkinson's disease. They suggested that the improvement resulted from the additional proprioceptive input leading to a modulation of dystonic impulses from the basal ganglia.

We have been able to treat the apraxia component of this condition using a scleral contact lens. In theory, this is possible because of the increased proprioceptive input as a result of the eyelids resting on the lens. The lens has also been effective in preventing complete closure of the lids during episodes of blepharospasm. This may be because of the mechanical effect of the lens on lid excursion and also possibly because of reducing stimuli from the afferent arm of the 'defective circuit' as reported by Anderson *et al*.

This treatment has been very successful in providing temporary benefit and restoration of binocular function in this patient with blepharospasm and eyelid apraxia. To the best of our knowledge, this has not been reported before.

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If there are any queries about the procedures used for designing or fitting the scleral lenses, please write to brian.melia@hey.nhs.uk

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Sir,

Acute angle-closure glaucoma and pupil-involving complete third nerve palsy as presenting signs of thrombosed cavernous sinus aneurysm

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Complete third nerve palsy (CTNP) with a fixed and dilated pupil is in most cases the presenting sign of intracranial aneurysm, usually at or near the junction of

the internal carotid artery and the posterior communicating artery.[1] Intracavernous sinus aneurysms (ICSA) are much less frequent, and typically produce combined oculomotor cranial neuropathies, trigeminal neuropathy, and Horner's syndrome in addition to TNP (nonisolated palsy).[1] Acute CTNP because of ICSA is extremely rare, and to the best of our knowledge there are no such reports in the literature.

We present a patient with atypical features of this disorder, who presented with acute angle-closure (AAC) glaucoma and CTNP as presenting signs of thrombosed ICSA.

Case report

A 67-year-old woman presented to the emergency room with a history of headache, left eye pain, and complete ptosis of 2 days duration. Medical history revealed mild hypertension, and left lumpectomy for breast carcinoma 10 years ago. The patient was not previously examined by an ophthalmologist, and no earlier ocular problems were reported. Left eye presented with visual acuity of 6/18, complete ptosis, exotropia, and mild hypotropia (Figure 1). Complete absence of supraduction, infraduction, and adduction was observed. Abduction was normal, and a minor incyclotorsion was noted. There was mild conjunctival congestion, mild corneal epithelial oedema, shallow anterior chamber, 360° closed angle seen by gonioscopy with appositional closure, and a fixed 6 mm dilated pupil nonreactive to light. Intraocular pressure (IOP) was 61 mmHg. Lens was clear, optic nerve head was normal, and retina was flat. In the right eye, visual acuity was of 6/6, with normal anterior and posterior segments, except for a mild shallow anterior chamber; gonioscopy revealed a grade I angle. The patient was treated by p.o. 120 cm^3 glycerol, i.v. acetazolamide 500 mg, and pilocarpine 2% drops every 10 min in the left eye for the first 2 h, and then q.i.d., and dexamethasone $\times 4$/day.

In the meanwhile, she was referred for immediate neurologic consultation, which was normal, except for the CTNP. The patient underwent computed tomography (CT) without contrast, showing a 2×2.5 cm mass in the left cavernous sinus scalloping the sphenoid bone. CT angiography revealed a left cavernous sinus aneurysm mostly thrombosed with small patent lumen extending into the thrombus. Magnetic resonance imaging T1-weighted showed an isointense mass within the cavernous sinus with a small hyperintense lesion corresponding to recent thrombus (Figure 2a). Postgadolinium T1-weighted study demonstrated peripheral and small central enhancement within the cavernous sinus (Figure 2b). Normal flow void was seen in the carotid artery but was absent within the lesion.

At 3 h after treatment initiation, visual acuity in the left eye improved to 6/9, IOP dropped to 14 mmHg, the pupil constricted, and the angle was opened. Eye pain and headache were partially relieved. Neodominium : YAG laser peripheral iridotomy was subsequently performed in both eyes in the following days. After neurosurgical and neuroradiological evaluation, observation only was recommended because of the thrombosis and the low flow within the aneurysm. During the follow-up of 2 months, clinical features remained unchanged.

Comment

ICSA occur predominantly in women in their sixth decade. They represent 2–3% of all intracranial aneurysms, more than 15% of symptomatic aneurysms without rupture, and 20–25% of cavernous sinus lesions. Acute presentation of ICSA is unusual, and their rupture is infrequent (0.5–2%)[2] causing carotid–cavernous fistula.

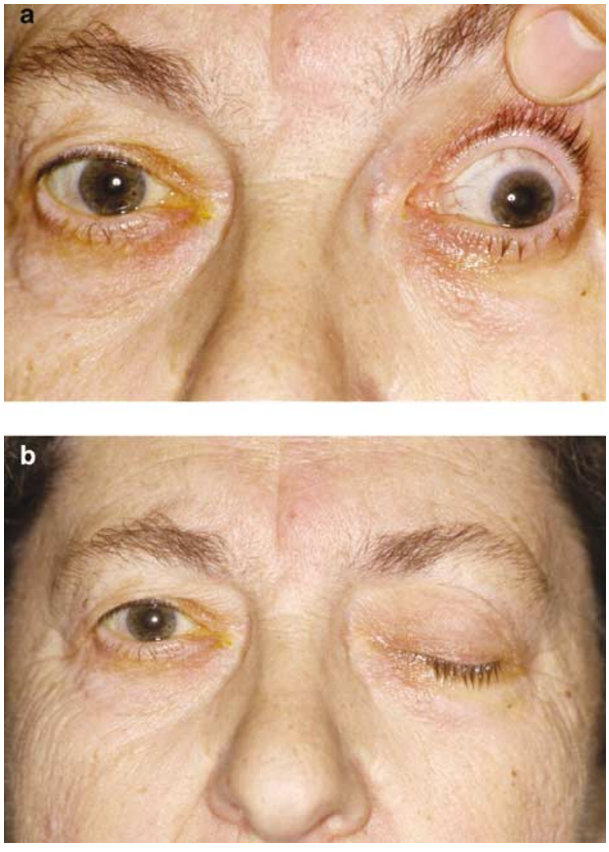

Figure 1 (Top) complete left ptosis; (Bottom) after manual elevation of upper left eyelid, we can observe left exotropia and hypotropia, and mid-dilated pupil.

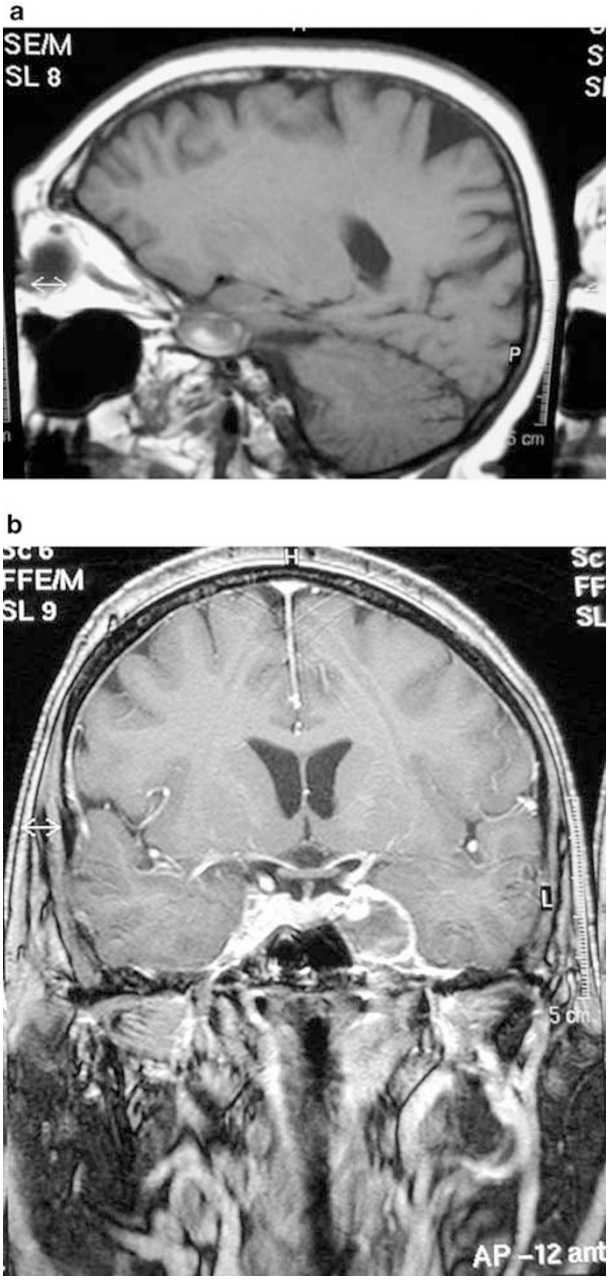

Figure 2 (a) Sagittal T1-weighted MRI scan without gadolinium, demonstrating a small hyperintense area within the thrombus because of recent haemorrhage. (b) Coronal T1-weighted MRI scan with gadolinium, showing peripheral enhancement of the aneurysm and small patent lumen within the thrombus.

Clinical findings are usually because of compression, unlike posterior communicating aneurysms, which can cause a life-threatening subarachnoid haemorrhage. The onset of signs and symptoms of ICSA is usually insidious. The pattern of serial involvement of the cranial nerves within the cavernous sinus is usually as follows: sixth, third, fifth, and fourth. However, acute presentation of ICSA is very rare. The pathogenic mechanism for the acute cranial neuropathies associated with ICSA has not been proven, and is probably because of acute cranial nerve ischaemia from thrombosis or compression of the branches of the intracavernous arteries that supply the cranial nerves during their intracavernous course.[3]

When an aneurysm compresses the oculomotor nerve, the pupil is usually dilated and reacts poorly to light, because of the injury to the superomedial pupillary fibres along the subarachnoid oculomotor nerve.[4] Our patient was predisposed to angle closure because of her anatomically narrow angles. This is a very rare sequence of events. We postulate that acute thrombosis in the aneurysm probably caused acute compression and a rapid mydriasis, precipitating pupillary block. A pupil involving TNP can theoretically produce AAC glaucoma in a predisposed eye, but to the best of our knowledge, there are only two reports in the literature of AAC glaucoma secondary to aneurysm-induced pupil-involving TNP;[5,6] none of them described a case of acute CTNP with AAC glaucoma caused by a thrombosed ICSA.

In our patient, scalloping of the adjacent bone was likely produced by a long-standing ICSA. The gold standard to rule out ICSA is cerebral angiography. The diagnosis of ICSA was confirmed by a less invasive and an available method of CT angiography, which showed a partially thrombosed ICSA. This diagnosis was also supported by MRI demonstrating a hyperintense area within the aneurysme because of recent thrombus, which was probably responsible for the presentation of acute CTNP with rapid dilatation of the pupil causing AAC glaucoma.

Acknowledgement

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Sir,

Atypical Cogan's syndrome presenting with bilateral acute glaucoma

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Cogan's syndrome (CS) is a rare, probably autoimmune, vasculitis. The defining features are eye problems, usually interstitial keratitis, and audiovestibular dysfunction—especially neurosensory deafness,[1] although systemic involvement occurs in 50–75% of cases.[2] CS can be classed as typical or atypical.[2–4] Typical CS manifests as interstitial keratitis with or without conjunctivitis and iritis, whereas atypical CS includes other forms of ocular inflammation with or without interstitial keratitis.[2] We report a patient presenting with bilateral acute glaucoma, that is, atypical CS and, severe neurological involvement controlled by prednisolone and methotrexate.

Case report

A previously well 40-year-old woman presented to her local ENT department with vertigo followed by bilateral neurosensory deafness. The MRI brain scan returned normal and a diagnosis of Meniere's disease was made at this time. Within 8 months, she presented to ophthalmology with a 3-week history of coloured haloes. Raised intraocular pressure (R 66, L 60 mm Hg), corneal oedema, and 360° peripheral anterior synechiae were noted and bilateral acute angle closure glaucoma was diagnosed and was treated medically. Corneal oedema and intraocular pressure resolved to reveal interstitial keratitis and uveitis, which responded to topical steroids. The cup-to-disc ratio was 0.5 and 0.2 (R and L, respectively). The patient eventually required right, then left unaugmented trabeculectomies and topical beta-blockers to optimise control.

At 36 months after initial presentation, the patient reported poor balance. Neurology assessment revealed sensory ataxia caused by axonal sensory peripheral neuropathy. There was severe, diffuse white matter ischaemia on the MRI brain scan (Figure 1) and, raised cerebrospinal fluid (CSF) total protein, and lymphocytes. All other relevant investigations were unremarkable. CS was diagnosed. Intravenous methylprednisolone (1 g/day for 3 days) was administered, followed by oral prednisolone (1 mg/kg/day) and aspirin. After 2 months, repeat CSF examination was normal. Azathioprine was added but subsequently stopped because of liver dysfunction. She declined further immunosuppression, but remained stable and prednisolone was gradually tapered.

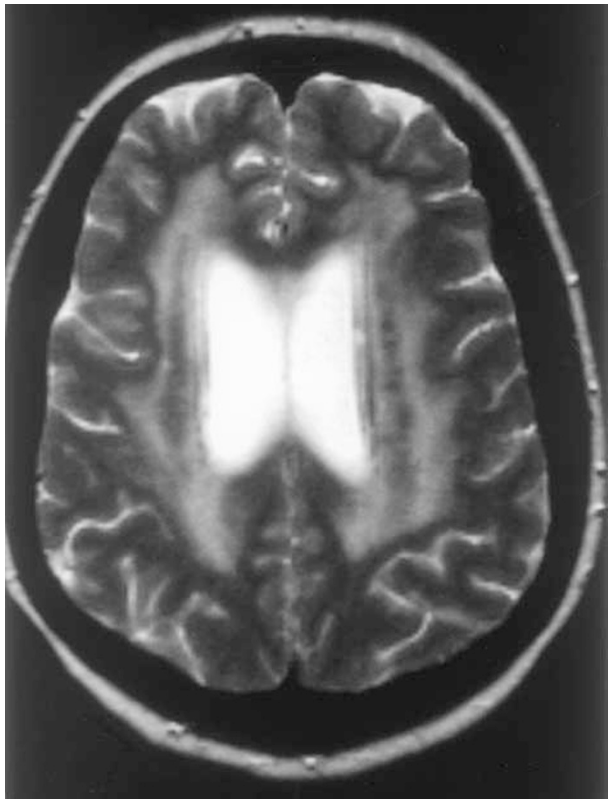

Figure 1 T2-weighted axial MRI brain scan. Large areas of increased signal reflecting widespread, diffuse ischaemic damage to both cerebral hemispheres.