# LETTERS TO THE EDITOR

## Ocular ball bullet injury: detection of gonioscopically unrecognisable cyclodialysis by ultrasound biomicroscopy

EDITOR,—Ocular ball bullet (BB) injuries are vision threatening and more than 1200 people every year are reported to sustain these injuries in the United States.[1] Fewer cases with BB injuries have been reported in Japan.[2–4] Ultrasound biomicroscopy (UBM) is useful in the morphological evaluation of the anterior segment of the eye.[5] We report a 13 year old boy who sustained cyclodialysis from a BB injury, which was not revealed by gonioscopy but was by UBM. To our knowledge, this is the first report describing detection of cyclodialysis from BB injury by UBM.

CASE REPORT

A 13 year old boy sustained an ocular BB injury in his left eye and visited us the following day. The boy was accidentally shot by his elder brother. The BB was made of plastic. His best corrected visual acuity was right eye, 1.2 and left eye, 0.06. Intraocular pressure was right eye, 17 mm Hg and left eye, 11 mm Hg. Slit lamp examination revealed corneal oedema and Descemet's folds in his left eye. The anterior chamber was of normal depth and showed moderate inflammation with aqueous cells (1+), flare (1+), and faint fibrinous exudate. Gonioscopy revealed an angle recession inferonasally with a trace hyphaema. The lens, vitreous body, and fundus of the eye were normal. Penetration of the globe was not identified. The patient was treated with atropine 1% three times daily and fluorometholone 0.1% six times daily. The next day, although corneal oedema decreased, the depth of the anterior chamber became shallower than that of the first examination and intraocular pressure decreased to 8 mm Hg. Funduscopy revealed chorioretinal folds in the posterior pole. We performed UBM, which demonstrated a small cyclodialysis in the 5 to 7 o'clock position (Fig 1), which was not apparent gonioscopically. Six days after the injury, the cornea became clearer and no inflammation in the anterior chamber was noted. Even though the depth of the anterior chamber had increased, ciliochoroidal fluid became evident (Fig 2). Intraocular pressure was 7 mm Hg. Ten days after the injury, intraocular pressure increased to 17 mm Hg. Chorioretinal folds gradually disappeared and best corrected visual acuity returned to 1.0 six weeks after the incident.

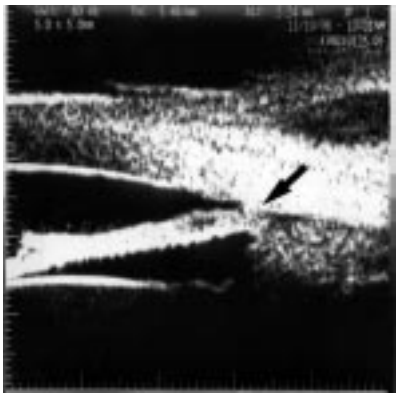

*Figure 1 Ultrasound biomicroscopy discloses that ciliary body is disinserted from the scleral spur (black arrow), which is obscured by iris. Note the shallow anterior chamber.*

*Figure 2 While the anterior chamber becomes deeper, ciliochoroidal fluid (white arrows) is discerned with persistent ciliary body detachment.*

COMMENT

Ocular BB injuries are vision threatening and more than a few of them result in eventual enucleation.[1 6–8] However, the patients without open globe injuries have better prognoses.[1 6 8] Our patient, who sustained closed globe injury, also regained visual acuity of 1.0. The usual muzzle velocity of a BB gun manufactured in the USA is 350 feet per second and its weight is 0.346 g.[1] Therefore, its kinetic energy is calculated at approximately 2.0 J. In our case, the weight and kinetic energy were 0.2 g and 0.4 J, respectively. We speculate that our patient's good visual prognosis may be associated with the relatively low kinetic energy generated by the BB gun. Airgun manufacturers' cooperation in Japan regulates their products to generate kinetic energy of 0.4 J or less. Takashima *et al* [3] reviewed 50 Japanese patients with ocular BB injury in the literature and described that none of the 50 patients sustained open globe injury and all but one patient had final visual acuity of 0.7 or better. In contrast with the good visual prognoses in Japan, Schein *et al* reported that 78 of 140 (56%) victims of ocular BB injury in the USA sustained open globe injury and only 31 of 140 (22%) achieved visual acuity of 20/40 or better.[6]

Cyclodialysis is the disinsertion of the ciliary body from the scleral spur and one of its main causes is blunt trauma. Sternberg *et al* [1] examined globes enucleated as a result of ocular BB injuries and elucidated frequent damage to the ciliary body histopathologically. The damage included tears into the ciliary body and haemorrhagic necrosis, often accompanied by choroidal haemorrhage and detachment. In the clinical setting, however, it is common that disrupted ocular tissue prevents us from assessing damage to the ciliary body. Additionally, cyclodialysis cleft is often not apparent gonioscopically, even if disruption of the ocular tissue is minimal and the anterior segment is clearly visible. This is because the iris is against the scleral spur, and the cleft is not open[9] as in this particular case. In the series of 140 ocular airgun injuries, the mean age of the victims was 13 years.[6] UBM might be well tolerated by even younger ages because of its non-invasive character. Therefore, the method seems to be useful to evaluate the anterior segment of the patients with closed globe injuries from BB guns.

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## Optic canal mucocoele from anterior clinoid pneumatisation

EDITOR,—We describe a 68 year old man who suffered from progressive visual loss in his left eye to no light perception within 10 days and subtle involvement of ipsilateral cranial nerves V(1) and VI. Both computed tomography (CT) and magnetic resonance imaging (MRI) showed a left optic canal lesion with expansion to the superior lateral (anterior clinoid) and inferior walls of the optic canal. Left supraorbital craniotomy was performed by a neurosurgeon. A mucocoele containing turbid fluid in the left strut with compression to the optic nerve was found during the operation. After removal of the lesion, the patient's best corrective visual acuity was improved to hand movements.

Though visual loss related to mucocoeles of the paranasal sinuses is not rare the frontal, ethmoid, and sphenoid sinuses are most often implicated. Only rarely has a pneumatised anterior clinoid been reported as a primary location for a mucocoele associated with visual loss.[1 2] We report a case of anterior clinoid mucocoele producing optic neuropathy and other subtle cranial neuropathies. We emphasise the relation of the optic strut to the optic canal and the superior orbital fissure in producing a symptom complex distinct from optic neuritis and orbital apex syndrome.

CASE REPORT

A 68 year old healthy man noticed progressive loss of vision in the left eye 2 weeks before

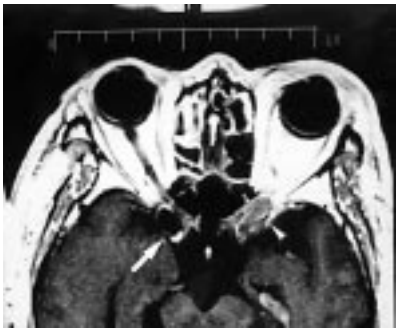

*Figure 1 T1 weighted MRI showed a bright lesion over the lateral wall of the left side optic canal with intracanalicular optic nerve compression (arrowhead). The right side anterior clinoid process is hyperpneumotised (arrow).*

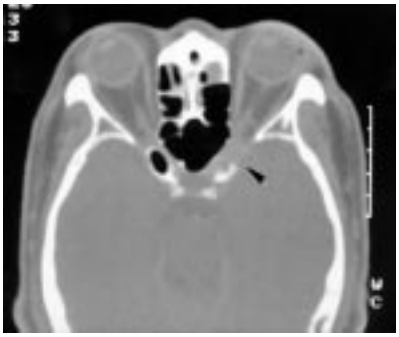

*Figure 2 The CT scan with bone window demonstrated the complete removal of the roof and lateral wall of the left side optic canal (arrowhead).*

admission. He had had frequent attacks of headache in the temporoparietal region for 2 months, but denied orbital pain or double vision. He had a history of hypertension for about 10 years with good medical control. Non-insulin dependent diabetes mellitus was noted recently. On admission his blood pressure was 140/70 mm Hg. General neurological examination was normal. Visual acuity of the left eye was no light perception (NLP). There was an amaurotic pupil on the left. There was mild limitation of abduction of the left eye. Corneal sensation was mildly impaired in the left eye, and there was no epiphora. No proptosis was present. Ophthalmoscopy revealed normal optic disc, retina, and vessels in both eyes. CT and MRI showed a minimally enhancing lesion of the left anterior clinoid, which encroached upon the superior, lateral, and inferior walls of the optic canal (Fig 1). The optic nerve appeared to be compressed medially. The right anterior clinoid process was pneumatised, suggesting a possible mucocoele of left anterior clinoid process.

Under general anaesthesia, the left optic canal was exposed extradurally via a supraorbital craniotomy. Mucoid fluid leaked out as soon as the roof of the optic canal was opened. The postoperative CT scan showed complete removal of the lesion and decompression of the lateral wall of the canal (Fig 2). The patient was discharged 2 weeks after surgery when the visual acuity improved to light perception. In spite of successful decompression, optic atrophy developed eventually. Two years later, after surgery for cataract, the visual acuity in the left eye improved to hand movements.

COMMENT

Mucocoeles involving the optic canal are extremely rare. Optic canal mucocoele is an ophthalmological emergency, since, without effective management, complete visual loss may develop within a few days. Two reported cases of a mucocoele originating in the anterior clinoid process can be found from the literature.[1] Both cases demonstrated bilateral pneumatisation of the anterior clinoid. The first patient developed severe visual loss with minimal recovery after surgery. The second patient, who declined surgery, had a recurrence of symptoms, resulting in optic atrophy. Another case report revealed significant visual improvement from 20/400 to 20/20 after surgery.[2] Again, this patient had bilateral pneumatisation of the anterior clinoid, very similar to our presented case. The cause of the mucocoele formation is uncertain, since there is no known ostium to become obstructed. Cystic degeneration or secondary inflammation is the proposed mechanism.[1] Pneumatisation of the sphenoid sinus can extend into the anterior clinoid. We believe that pre-existing anterior clinoid pneumatisation with secondary inflammation or degeneration rather than osteum obstruction is the cause of anterior clinoid mucocoele.

The optic strut is a segment of bone that joins the lesser wing of the sphenoid to the body of the sphenoid bone and forms the inferior and lateral walls of the optic canal, thus separating the canal from the superior orbital fissure.[3] Thus, symptoms and signs from a lesion within the strut, such as mucocoele, may affect structures of the optic canal and the superior orbital fissure, simulating an orbital apex syndrome. Our case showed severe dysfunction of the left optic nerve and signs consistent with mild compression of the left cranial nerves V(1) and VI.

The differential diagnosis includes mucocoeles of the sphenoid sinus, retrobulbar neuritis, and space occupying tumours such as craniopharyngioma, Rathke cleft cyst, pituitary adenoma, epidermoid cyst and carcinoma, cholesteatoma, meningioma, lymphoid tumour, optic glioma, and arachnoid cyst.[2–4] Reports of similar lesions are initially diagnosed as acute retrobulbar neuritis.[2 3] However the prevalence of retrobulbar neuritis in patients older than 60 is low. Tumour is considered unlikely in these cases because radiological studies reveal a lesion originating from within the optic canal and not from the brain parenchyma or meninges.

Surgical decompression is the treatment of choice for optic canal mucocoeles. For medial compression of the canal, a variety of otolaryngological external and internal approaches may be utilised. In this case, however, with mucocoele involving the lateral wall of the optic canal principally, the transcranial approach is preferred. A delay in surgery of more than 7–10 days after the onset of visual dysfunction is often associated with poor visual prognosis.[2]

We emphasise the importance of imaging in the evaluation of patients with atypical "optic neuritis." Inappropriate age for onset of demyelinating disease is a general concern that should lead to additional examination. However, the importance of other subtle cranial neuropathies may help identify a lesion not only of the orbital apex but also of the optic structure which straddles the superior orbital fissure and the optic canal. Prompt diagnosis and surgical intervention may improve the visual outcome in these patients.

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## Optic nerve compression by the internal carotid artery in patients with normal tension and high tension glaucoma

EDITOR,—It is generally accepted that ectatic or even normal intracranial blood vessels can cause dysfunction of cranial nerves when situated in an aberrant location.[1–3] Although the occurrence of such compression damage to the optic nerve is poorly recognised, several studies have shown the optic nerves can in fact be damaged by vascular compression.[4–8] Recently, optic nerve compression by normal internal carotid arteries (ICA) has been suggested as a possible cause of visual field defects in patients with optic neuropathy[4–6] and normal tension glaucoma (NTG).[7 8] Various mechanisms have been proposed to explain the aetiology of NTG; however, the exact cause of NTG remains to be elucidated.[9]

The present study was designed to determine whether optic nerve compression by the ICA can play a role in the visual field defects in patients with NTG.

Sixteen Japanese patients with NTG (average 65.3 (SD 11.9) years) and 16 age matched patients with high tension glaucoma (HTG) (average 65.6 (12.7) years) were included in this study. Magnetic resonance imaging was performed on a 1.5 T system (Signa Advantage, General Electric, Milwaukee, WI, USA). To determine the spatial relations between the optic nerves and the adjacent ICA, coronal and sagittal $T_1$ weighted images of these structures were taken with the spoiled gradient recalled acquisition in the steady state, one of the magnetic resonance angiography methods. Radiological diagnosis was made by two or three experienced radiologists who were informed of the age and sex but masked to the type of glaucoma of the patients. The relation between the optic nerve and the adjacent ICA was designated as either "with compression" or "without compression" (Fig 1).

In this series, none of the patients had intracranial abnormalities such as tumours, aneurysms, or significant atherosclerotic changes of the ICA. In the NTG group, compression by ICA was found in 24 (75%) of 32 optic nerves. Bilateral compression was observed in 12 NTG patients. In the HTG patients, compression by ICA was found in 12

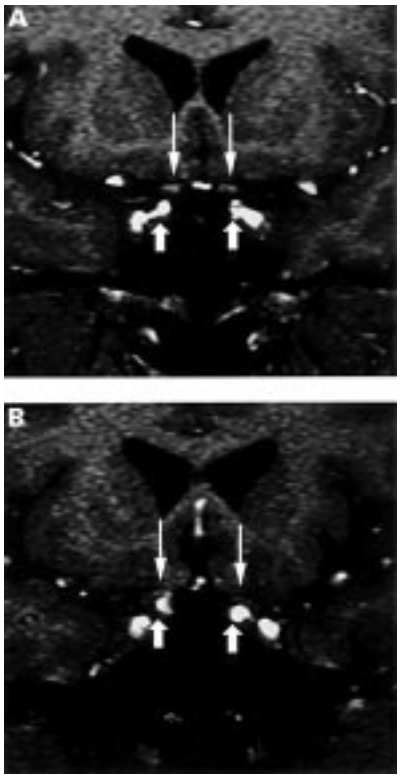


*Figure 1 Coronal magnetic resonance imaging of the optic nerve and surrounding area using spoiled gradient recalled acquisition in the steady state. The anatomical relations between the optic nerves (thin arrows) and the internal carotid arteries (thick arrows) are summarised. (A) Without compression. There is no contact between the optic nerve and the internal carotid artery. (B) With compression. The optic nerve is in contact with the internal carotid artery and there is distortion of the optic nerve contour.*

(37.5%) of 32 optic nerves. Bilateral compression occurred in only three patients. Statistical analysis showed that the occurrence of optic nerve compression was significantly higher in NTG group than in the HTG group ($p = 0.002$, $\chi^2$ test).

Clinical characteristics of the NTG and HTG patients with compression were compared with those without compression. No statistically significant differences were found in the patients' age, visual acuity, cup to disc ratio, and visual field variables of the Humphrey 30-2 program (Table 1).

*Table 1 Relation between vascular compression and visual function in normal tension glaucoma and high tension glaucoma*

| | *With compression* | *Without compression* | *p Value** |
|---|---|---|---|
| Normal tension glaucoma | | | |
| Number of eyes | 24 | 8 | |
| Age (years) | 65.6 (11.97) | 64.3 (11.7) | 0.786 |
| Visual acuity (logMAR) | 0.16 (0.44) | 0.06 (0.14) | 0.548 |
| Cup to disc ratio | 0.80 (0.17) | 0.66 (0.23) | 0.083 |
| MD (dB) | −15.02 (8.77) | −15.95 (5.94) | 0.781 |
| PSD (dB) | 9.49 (3.70) | 11.96 (1.84) | 0.082 |
| CPSD (dB) | 8.36 (4.03) | 11.39 (3.01) | 0.061 |
| High tension glaucoma | | | |
| Number of eyes | 12 | 20 | |
| Age (years) | 68.56 (10.4) | 63.9 (13.5) | 0.321 |
| Visual acuity (logMAR) | 0.20 (0.27) | 0.53 (0.99) | 0.277 |
| Cup to disc ratio | 0.62 (0.22) | 0.78 (0.23) | 0.063 |
| MD (dB) | −11.23 (10.60) | −15.26 (12.63) | 0.361 |
| PSD (dB) | 6.20 (4.27) | 5.76 (4.51) | 0.787 |
| CPSD (dB) | 5.48 (4.66) | 5.01 (4.98) | 0.795 |

*Student's *t* test; MD = mean deviation; PSD = pattern standard deviation; CPSD = corrected pattern standard deviation.

## COMMENT

Compressive optic neuropathy is usually caused by intracranial lesions, such as brain tumours and aneurysms, and not by normal vessels.[10] However, neuropathy of the trigeminal, facial, and abducens nerves caused from compression by normal blood vessels has been described.[1–3] Furthermore, Nishioka *et al* reported cases in which impaired visual function was improved by surgical release of compression from normal brain vessels.[4] These finding may support the idea that even the optic nerves can be damaged by the compression of normal appearing ICAs that do not show atherosclerotic or aneurysmal changes.

In spite of the high correlation between the presence of vascular compression in patients with NTG, our data do not prove that vascular compression is a major cause of the field defects in NTG. In fact, 25% of our NTG patients did not show vascular compression, and almost half of the control HTG patients also had vascular compression. Moreover, there was no statistically significant difference in the clinical characteristics of the eyes with optic nerve compression compared with those without. However, we did find a significantly higher percentage of patients who showed compression of the optic nerves by the ICA in the NTG than in the HTG patients. This difference suggests the possibility that vascular compression by normal ICA may play a role in the visual field defects in some cases of NTG.

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## Intermittent ptosis due to large exophoria

EDITOR,—Involuntary closure of the eyelid can be due to either a ptosis with dysfunction of the levator muscle, or a form of blepharospasm with exaggerated contraction of the orbicularis muscle. In true ptosis, no lid crease can be observed in the upper eyelid, whereas in (essential) blepharospasm or (secondary) pseudoptosis a lid crease is present.[1] We report a case of involuntary intermittent eyelid closure secundary to an exophoria.

### CASE REPORT

A 67 year old man had complained for 3 years that his left eyelid seemed to fall down and close spontaneously several times a day, and with increasing frequency. This happened especially when he was at home quietly watching television or when he was talking to someone, which made him feel embarrassed. He was able to open the eye again voluntarily, if he paid attention. The eyelid closure never occurred when he was driving a car. Also, when he closed his right eye, he could keep the left one open without any problem. When spontaneous eyelid closure did occur, he sometimes had double vision for a very short while. It had also been noted by his wife that his left eye deviated outward sometimes.

The patient had been known to have hypertension for 20 years and had been treated with medication by his family physician; 6 months before his first visit to our department a light form of diabetes mellitus type 2 had been diagnosed.

When the complaints started 3 years earlier, the patient was seen by a neurologist in a peripheral medical centre, who found no other neurological abnormalities than the intermittent ptosis. Blood tests and EMG for myasthenia were negative. Other blood tests were also normal. At the second visit an active contraction of the left eyelid was noticed and a blepharospasm was suspected. The patient was referred to the neurology department of an academic medical centre for treatment of the blepharospasm with botulin injections.[2]

At his first visit there, a mild blepharospasm of his left eyelid was diagnosed. It was also noted that while the left eyelid closed, the left

eye deviated outward and stayed there until the patient blinked several times. Treatment with botulin toxin was started but had no success. A computed tomography scan of the head was normal. Magnetic resonance imaging of the mesencephalon and the brain showed no abnormalities. A ptosis hook on his glasses was subsequently tried but brought no relief.

Then the patient was referred to the blepharospasm group of the neurology department of our hospital. There it was noticed that when the lid opening narrowed, the eye always deviated outward. With the alternate cover test a latent divergent squint was found. Therefore the patient was referred to us for neuro-ophthalmological evaluation.

We found a visual acuity of 20/20 in the right eye and 16/20 in the left, with a hypermetropic correction of about +2.50. Anterior segments, lens, and funduscopy were unremarkable. Pupillary light reactions were normal, and confrontational field testing was full. With close observation in the examination room we saw that the involuntary closure of the left eyelid was always preceded by an exodeviation of the left eye. This was later confirmed with the use of a video camera. After closure, a lid crease could still be observed in the upper lid.

Subsequent orthoptic examination showed a large exophoria (30 prism dioptres) which easily decompensated in a manifest divergent angle (30 prism dioptres). The eye movements were unrestricted and concomitant. The voluntary convergence was excellent. When a manifest deviation occurred, there was mostly suppression of the left eye, although the patient sometimes experienced double vision, especially when asked about it. At reading distance (30 cm), there was some binocular and stereoscopic vision (TNO stereotest 240"). With the Bagolini striated glasses there was a good fusion area between 20 prism dioptres base temporal and 14 prism dioptres base nasal.

On the basis of these orthoptic results, a presumptive diagnosis was made of a large exophoria of the left eye with a secondary blepharospastic eyelid closure to prevent diplopia. Eye muscle surgery was proposed, and a recession (5 mm) of the lateral rectus and a resection (5 mm) of the medial rectus muscle of the left eye was performed. Postoperatively, the eyes were straight with normal binocular single vision, and no more involuntary eyelid closure or double vision has occurred after a postoperative follow up of 14 months.

COMMENT

In this patient, the easily decompensating exophoria caused diplopia, and this provoked involuntary eyelid closure. The patient was not aware of the diplopia occurring just before his eyelid fell down; he had experienced diplopia occasionally, but could not indicate when. Monocular eye closure in intermittent exotropia has been described and has been thought to be due to avoidance of diplopia.[3] However, Wiggins and von Noorden[4] point out that bright light also may cause monocular eye closure, especially in intermittent exotropes, even when they do not experience diplopia. The authors demonstrated with video recordings that eyelid closure occurred before exodeviation in most patients with intermittent exotropia. We did not test our patient under bright light. The patient's history indicated, however, that the eyelid fell down especially in quiet indoor situations. We also observed it under (dim) examination room lighting. Moreover the patient had good convergence and fusion. Therefore it does not seem to us that bright light played a role in the eyelid closure in this case.

Although in our patient the exophoria had in fact been noticed at several neurological examinations, it had not been recognised as such, or as the possible trigger for secondary ptosis. Neuro-ophthalmological examination at an earlier stage could have prevented unnecessary neuroimaging and botulin injections.

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## Permanent extraocular muscle damage following botulinum toxin injection

EDITOR,—The use of botulinum toxin in the management of ocular motility disorders is well established.[1] While transient side effects like ptosis[2] and diplopia[3 4] due to local spread of the toxin do occur, to our knowledge permanent extraocular muscle damage has not been reported. We report a patient with congenital right superior oblique weakness who underwent botulinum toxin injection to the left inferior rectus muscle. This resulted in permanent and profound loss of inferior rectus muscle function, with atrophy of the muscle, confirmed by magnetic resonance imaging scan.

CASE REPORT

A 70 year old white man was seen for increasing angle of deviation of a long standing right hyperphoria which had previously been controlled with a small prism correction. His general health was excellent and his only medication was phenelzine 15 mg daily. His visual acuity was 6/5 in each eye. There was a small right hyperphoria (8 prism dioptres) for near and distance with right superior oblique underaction, right inferior oblique overaction, left superior rectus underaction, and left inferior rectus overaction. Symptoms of difficulty maintaining binocular vision while reading were initially alleviated by increasing his prism correction, but 9 months later the deviation had increased, measuring 13 dioptres in the reading position and 4 dioptres for distance. A decision was made to proceed with left inferior rectus botulinum toxin injection. The injection was performed under electromyography control using a 27 gauge monopolar needle. The injection was performed through the lower eyelid, angled upwards, advancing the needle posteriorly, superiorly, and nasally by a surgeon (BWF) experienced in the technique. 2.5 U "Botox" botulinum toxin A were injected. The EMG response from the muscle was low to moderate, but there was no apparent complication associated with the procedure.

At review 1 month later the patient complained of diplopia in all directions of gaze.

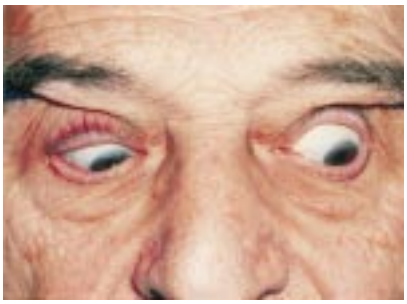

*Figure 1 Limited function of left inferior rectus muscle 10 months after botulinum toxin injection.*

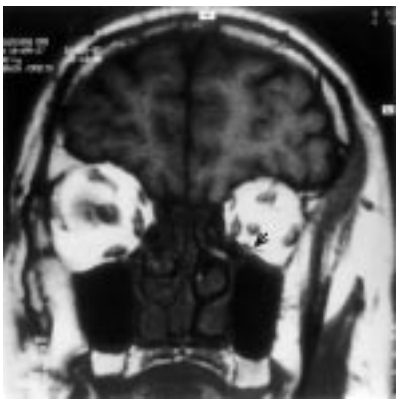

*Figure 2 Coronal scan of orbits 10 months after botulinum toxin injection. Note atrophy of left inferior rectus muscle (arrow).*

There was a left hypertropia of 20 dioptres in primary position, which increased on laevodepression, in keeping with left inferior rectus muscle paresis. Over the following 10 months, there was persisting diplopia with no change in Hess chart measurements, and no recovery of left inferior rectus muscle function (Fig 1). Forced duction test did not reveal any significant left superior rectus contracture. Investigations for thyroid dysfunction and myasthenia were negative. Magnetic resonance imaging of the patient's orbits showed atrophy of the left inferior rectus muscle (Fig 2).

Inferior transposition of the medial and lateral recti muscles was performed (by JPL). The inferior rectus muscle insertion appeared normal at the time of surgery. No attempt was made to explore the muscle more posteriorly. The procedure produced satisfactory alignment in primary position, with a small overcorrection in laevoversion.

COMMENT

Injection of botulinum toxin into a clinically overacting muscle produces a temporary reversible paralysis of that muscle. The result of this paralysis is a change in the force dynamics of the paired antagonistic muscles, which allows the weaker opposing muscle to gain force advantage.

The paralytic action of botulinum toxin is attributed to blockade of neuromuscular transmission by interfering with the release of the neurotransmitter acetylcholine at the motor end plate.[5] The paralysis following the use of botulinum is generally associated with complete recovery of neuromuscular function over 3–4 months.[6] Permanent histological changes have been reported in animal studies of the orbital, singly innervated muscle fibre of adult monkey extraocular muscles. Structural changes in muscle fibres and decrease in the density and lumenal area of vasculature of the

muscle fibres was seen.[7] Histological evidence of atrophy in the leg muscles has also been reported as a distant effect of botulinum treatment of cervical dystonia.[8]

Permanent superior rectus muscle weakness in association with botulinum induced ptosis has also been reported, and was presumed to be due either to breakdown of fusion or contracture of the ipsilateral antagonist.[9] The possible causes of left inferior rectus muscle atrophy following botulinum toxin injection in our case include intramuscular haematoma or direct damage to the nerve to the muscle within the muscle cone. Inferior rectus muscle paresis has been reported following retrobulbar anaesthesia for cataract surgery,[10] and the mechanism may have been similar.

Permanent damage to an extraocular muscle following botulinum toxin injection is a rare complication of the procedure. With increasing numbers of patients undergoing the procedure, both the patient and the surgeon should be aware of this rare complication of botulinum toxin injection.

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## Delayed diagnosis of homocystinuria as a cause of vascular retinal occlusion in young adults

EDITOR,—Retinal vascular occlusions in young adults are seen very infrequently and are generally associated with systemic disorders.[1] Diagnosis of the underlying disease is very important because of treatment and prevention of recurrence.[2] We report two cases initially presenting with systemic neurological disease. Both developed retinal vascular occlusions and the diagnosis of the underlying cause was only made afterwards.

CASE REPORTS

A 30 year old, obese woman presented to the neurologist with acquired perceptive deafness, a tetrapyramidal syndrome, with gait problems and urinary incontinence. Multiple sclerosis was considered, but cerebral magnetic resonance imaging scan showed only atypical lesions. Symptoms gradually decreased without disappearing completely. Six months later an occlusion of the retinal nasal inferior artery occurred in the right eye, visual acuity was 20/20 in both eyes (Fig 1). Haematological evaluation revealed a disturbed determination of peak levels of homocysteine after oral loading with methionine, suggesting homocystinuria causing thromboembolic processes in the cerebrum and in the eye. Repeated determination after oral loading with methionine after vitamin B12 treatment was still disturbed, confirming the diagnosis of homocystinuria. The patient was treated with pyridoxine and acetylsalicylic acid. Two out of four of the patient's sisters were also found to suffer from homocystinuria.

A 23 year old obese female, who smoked and had used contraceptives, underwent caesarean section because of negative discongruency and breech presentation. Postoperatively, thrombophlebitis of the arm and erythema nodosum of the legs developed. Four weeks later visual acuity in the left eye deteriorated, caused by massive occlusive retinal vasculitis, retinal haemorrhages, papillitis, and massive retinal oedema with Roth spots (Fig 2). A biopsy of the skin lesions of the leg showed leucocclastic vasculitis. Further investigation could not reveal the underlying cause. Treatment with clindamycin, acetylsalicylic acid, and methylprednisolone was started and panretinal laser therapy was performed. Four months after treatment was completed, the other eye developed retinal haemorrhages and occlusion of the upper, temporal branch of the main retinal vein.

Analysis showed elevated peak levels of homocysteine after loading with methionine, suggesting homocystinuria. Analysis of DNA and cystathionine β synthetase in the cultured fibroblasts confirmed the diagnosis of homocystinuria. Treatment with acetylsalicylic acid, folic acid, and pyridoxine resulted in the normalisation of the homocysteine level. The visual acuity 1 year after the first event was 20/15 and 20/50 in the right eye and left eye, respectively.

COMMENT

Homocystinuria consists of a group of different metabolic disorders, all resulting in elevated levels of homocysteine in blood and urine. The most important is the cystathionine β synthetase reduction. Homocysteine interferes with crosslinking of collagen resulting in disturbances in the endothelial synthesis. Also there is an increased platelet adhesion causing thrombotic occlusive disease. The vascular complications led to occlusion of coronary, renal, and cerebral arteries and veins.[3]

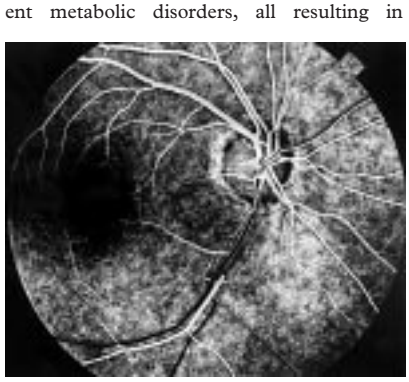

*Figure 1 Early phase fluorescein angiography of right eye of patient 1, showing occlusion of the retinal nasal inferior artery.*

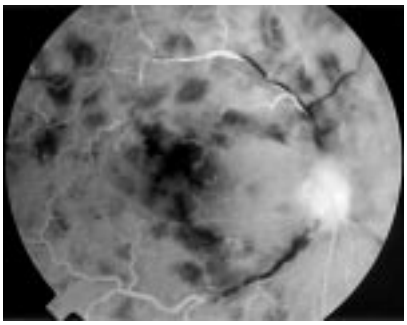

*Figure 2 Fluorescein angiography of right eye of patient 2, showing massive occlusive retinal vasculitis, papillitis, retinal haemorrhages, and massive retinal oedema and Roth spots.*

The most frequent manifestations in homozygous patients are disorders of the lens (ectopia 85%, microspherophakia), mental retardation, cardiovascular defects, and skeletal changes.[4 5] Retinal vascular occlusion is an infrequent manifestation of homocystinuria.[5–7]

The homozygotic form leads to very early vascular disease with early death and the above mentioned manifestations. Heterozygotic carriers (1:70 in general population) are at risk for occlusive vascular disease at a young age. This can often be prevented by treatment with low doses of acetylsalicylic acid, pyridoxine, and sometimes folic acid.[3] Factors suggesting other causes of thromboembolism (pregnancy, obesity, oral contraceptives) can delay the diagnosis of homocystinuria and successful treatment.[2]

In conclusion, homocystinuria should be considered in cases of young adults with retinal vascular occlusions, even if there are no other ocular abnormalities—for example, ectopia lentis. The possible sequelae for general health should be of concern when evaluating and treating these patients.

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## Familial amyloidosis of the Finnish type

EDITOR,—Familial amyloidosis of the Finnish type (FAF), also known as Meretoja syndrome, is a rare autosomal dominant disorder first described by Meretoja in 1969.[1] It is thought to develop as a result of a single point mutation involving the gelsolin gene located on chromosome 9. The estimated total number of patients in Finland is 400. Approximately 15 cases have been described outside Finland.[2] We present the first case to be recognised in the UK demonstrating the classic signs of corneal lattice dystrophy, cranial neuropathy, and skin changes with an autosomal dominant pedigree.

### CASE REPORT

A 73 year old woman presented with gradual reduction in visual acuity in her left eye. She had suffered recurrent corneal erosions affecting her left eye and was diagnosed as having corneal lattice dystrophy 18 years previously. At the time of presentation she was under investigation by a neurologist for progressive weakness of her facial muscles. There was no medical or drug history of note. Family members affected with corneal lattice dystrophy included her daughter and three cousins.

Examination revealed bilateral blepharochalasis, thickened facial skin, and bilateral lower motor neuron facial nerve palsies (Fig 1). Her visual acuity was 6/9 in the right eye and 6/60 improving to 6/36 with a pinhole in the left eye. She had bilateral corneal lattice dystrophy and an area of epithelial loss and sloughing associated with a mild left sided anterior uveitis (Fig 2). Lens opacities were also noted.

She was managed with topical mydriatics, antibiotics, and intensive lubricants. Despite an initial improvement, the cornea failed to re-epithelialise and a combined left penetrating keratoplasty and extracapsular cataract extraction was performed 5 months later. One year postoperatively, her acuity was 6/18 in the right eye, 6/9 in the left eye, and relatively symptom free on intensive lubricants.

The possibility of FAF was considered. Histology of the left corneal button removed at keratoplasty confirmed the characteristic amyloid deposition of lattice dystrophy. Nerve conduction studies demonstrated bilateral facial nerve conduction deficits as well as a subclinical right carpal tunnel syndrome. Scintigraphy using $^{123}$iodine labelled serum amyloid P component confirmed the expected systemic nature of the condition with amyloid deposits noted in the patient's spleen and kidneys. DNA testing of the patient and her daughter revealed the presence of a point mutation in the gelsolin gene located on chromosome 9 confirming the diagnosis of Meretoja syndrome.

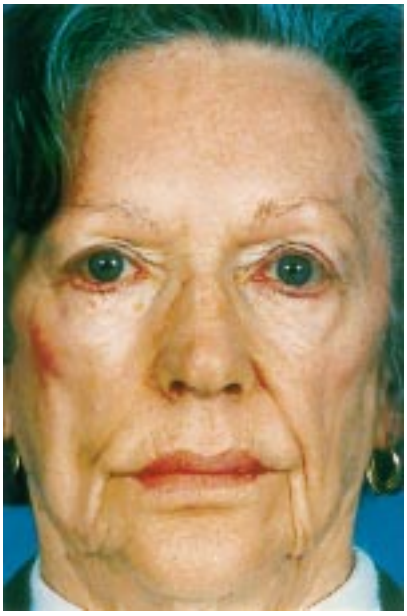

*Figure 1 Photograph showing bilateral blepharochalasis, thickened facial skin, and bilateral lower motor neuron facial nerve palsies.*

### COMMENT

The Finnish type of familial amyloidosis is a systemic disease inherited in an autosomal dominant manner characterised by progressive cranial neuropathy (particularly involving the facial nerve), corneal lattice dystrophy, distal sensorimotor neuropathy, and varying degrees of skin change. The onset of symptoms is typically in the third and fourth decades with slow progress so that the majority are still in good health in their seventh decade.[2 3]

The condition is common in Finland but rare elsewhere. This patient and her daughter are the first two cases to be reported in the UK. The patient has the classic features of the disease and demonstrates the point mutation on the gelsolin gene responsible for it. However, although the corneal histology demonstrated the presence of amyloid deposits, immunocytochemistry showed no labelling of the deposits with antibodies to pre-albumin, amyloid A, and amyloid P. This is in contrast with other cases reported where amyloid stained with antisera to serum amyloid P.[4] Whether this represents a subtype of the condition is uncertain.

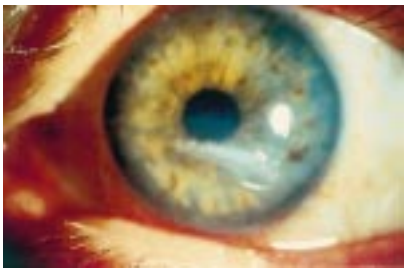

*Figure 2 Area of epithelial loss and sloughing associated with a mild left sided anterior uveitis.*

Various treatments are available targeted at each step in the pathogenesis of all types of amyloidosis with variable success and much research, including genetic manipulation, is being done in this regard.[5 6] However, at present the treatment of this disorder is mainly based on alleviating symptoms.

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