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Sir,

Atypical Cogan's syndrome presenting with bilateral acute glaucoma

Eye (2004) **18,** 328–329. doi:10.1038/sj.eye.6700655

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.

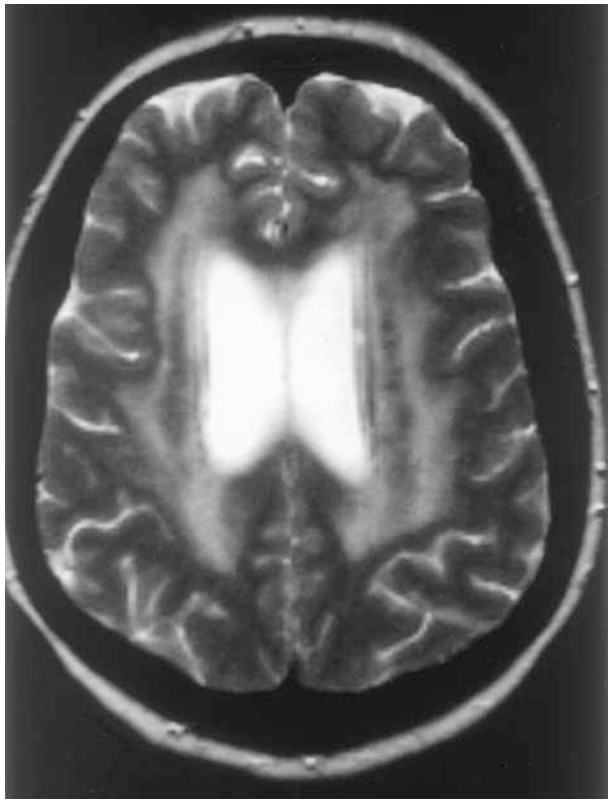

Figure 1 T2-weighted axial MRI brain scan. Large areas of increased signal reflecting widespread, diffuse ischaemic damage to both cerebral hemispheres.

The patient relapsed 3 months later with headache and a transient ischaemic attack while on prednisolone 30 mg/day. Labyrinthine failure had developed. Prednisolone was increased to 1 mg/kg/day and methotrexate was introduced at 15 mg/week. Within 3 months, she had developed left anterior optic neuritis with retinochoroidal oedema causing reduced visual acuity (6/24) and colour vision (15/17 Ishihara). On MRI brain scan, there was progression of ischaemic changes. Topical steroid was given and methotrexate was increased and continued at 20 mg/week. Acuity and colour vision improved to normal. Prednisolone was tapered slowly to 10–15 mg/day. Neither the keratitis nor the uveitis has recurred to date. The patient has been stable for 2 years.

Comment

CS presents almost equally to ophthalmology and ENT and median time to second organ involvement is 1 month.[5] At least one serious outcome (blindness, deafness, vasculitis, aortic insufficiency, or death) occurs in 63% of cases.[2] In all, 10% of patients have neurologic involvement;[2,5] although widespread CNS ischaemia has been reported in only two individuals.[5] About 30% of CS is atypical and more likely to be associated with systemic features and a poorer prognosis.[3,4] Our patient had an atypical ophthalmic mode of presentation, which probably represented acute on chronic angle closure glaucoma. Furthermore, and in keeping with her atypical CS, she had severe, widespread neurologic involvement with CNS ischaemia, optic neuritis, and peripheral neuropathy.

The most common serious complication of CS is deafness, which may be ameliorated if oral steroids are started within 2 weeks of onset.[3] Ocular inflammation frequently responds to topical steroids. Systemic vasculitis usually responds to oral steroids, although aortitis may require aortic valve replacement.

The role of other immunosuppressants is not established and, owing to the rarity of CS, may depend upon the collected experience of case reports. Three patients with diagnosed CS, who underwent methotrexate therapy experienced stabilised hearing and were able to reduce or discontinue steroids.[6] Our experience supports the use of methotrexate in steroid-resistant patients, especially those with CNS involvement.

We recommend that all patients with CS, especially atypical cases, should have regular ENT and ophthalmic assessments and that neurologic symptoms require urgent assessment. Such collaboration will help ensure appropriate and timely therapy.

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Sir,

Valsalva retinopathy associated with riding a motorcycle

Eye (2004) **18,** 329–331. doi:10.1038/sj.eye.6700658

The Valsalva manoeuvre comprises forcible exhalation against a closed glottis, thereby creating a sudden increase in the intrathoracic or intraabdominal pressure.[1] Spontaneous rupture of peri-foveal capillaries may develop, leading to a characteristic clinical picture of a retinal haemorrhage in an otherwise healthy eye.[2] The haemorrhage typically occurs at the macula and in the vast majority of cases is an isolated and self-limited event.[2] A case is presented of a transient maculopathy in a young healthy male attributable to a Valsalva