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Sir,

A case of trigeminal schwannoma presenting as Raeder's syndrome in a child

Raeder's paratrigeminal syndrome consists of Horner's syndrome in association with headache and trigeminal dysfunction. Raeder's syndrome is perhaps now more often associated with unilateral cluster headache and atypical migraine. The original description was of five cases of painful post-ganglionic Horner's syndrome, considered to be due to lesions arising from the middle cranial fossa. There are a multiplicity of other causes for this combination of signs, including tumours arising from the trigeminal nerve. We describe such a patient.

Case report

A 10-year-old girl presented with an 18 month history of painless blurring of vision in her right eye. She had been diagnosed as suffering from allergic keratoconjunctivitis and treated with topical steroid and sodium cromoglycate. Her eye had failed to respond to this treatment and the diagnosis had been revised to one of floppy eyelid syndrome. All topical therapy was stopped for a period but there was no improvement in her condition.

She was referred to the oculoplastic service at Manchester Royal Eye Hospital. Her examination findings were as follows: Her corrected visual acuity was 6/24 right and 6/5 left. The right cornea showed punctate epithelial erosions, anterior stromal scarring and peripheral neovascularisation. She had a right 3 mm ptosis. The right pupil was miosed, but reacted normally to light and near. Her ocular motility was normal; neurological examination showed reduced soft touch sensation on the right side of her face and anaesthesia of the right cornea. There was wasting of the temporalis and masseter muscles, and weakness of the pterygoids. Her examination findings were otherwise normal. In particular, there were no clinical features of neurofibromatosis and she had no other abnormal neurological findings.

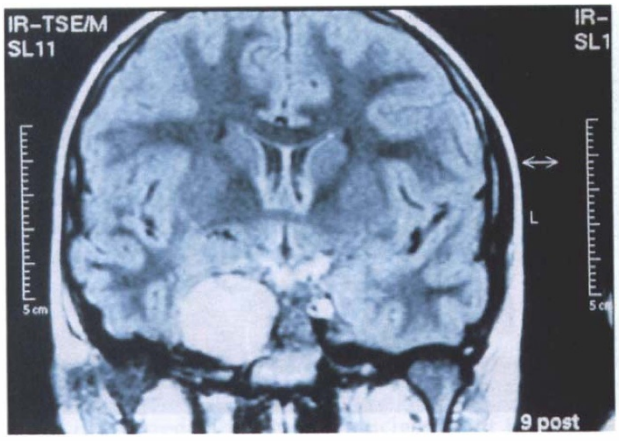

Fig. 1. *Coronal T2-weighted MRI scan showing the 5 cm trigeminal schwannoma.*

The findings of a right Horner's syndrome combined with signs of ipsilateral trigeminal dysfunction suggested a lesion of the floor of the middle cranial fossa. An MRI scan showed a large (5 cm diameter) tumour arising from the right trigeminal nerve (Fig. 1). She underwent neurosurgical resection of the lesion. Histological examination showed a tumour composed of spindle cells staining strongly for vimentin and S100, and negative for neurone-specific enolase and smooth muscle actin. The diagnosis was therefore a schwannoma of the trigeminal nerve.

Her post-operative course was complicated by the development of a large central neurotrophic ulcer in the right cornea (Fig. 2). This was treated initially with topical antibiotics and lubricants followed by botulinum-toxin-induced ptosis. As a definitive protective procedure, she later required a lateral tarsorrhaphy, which allowed the cornea to heal completely. Her corrected visual acuity was reduced to counting fingers secondary to corneal scarring. A permanent central tarsorrhaphy was required for corneal protection. This had obvious cosmetic implications and therefore following careful discussion with the parents she underwent an enucleation with primary hydroxyapatite implant wrapped in autologous fascia lata.

Comment

Raeder first described his paratrigeminal syndrome in 1924. He presented four cases of incomplete, post-ganglionic Horner's syndrome with trigeminal and other cranial nerve involvement and one case with isolated trigeminal neuralgia.[1] In each of the original descriptions there was either facial or periorbital pain. His first case was found to have an endothelioma of the trigeminal nerve arising from the Gasserian ganglion at post-mortem. Ford and Walsh[2] discussed a group of patients with what is now recognised as cluster headaches, all of which were benign. Grimson and Thompson,[3] in their excellent review, classified Raeder's syndrome into three main groups: those with multiple parasellar cranial nerve involvement and, of those without this, the classic cluster headache group and the atypical headaches group (in which there may be associated systemic or local disease,

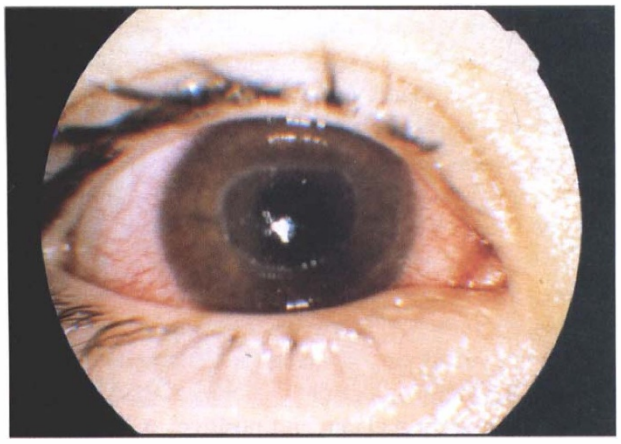

Fig. 2. *Central neurotrophic corneal ulcer of the right eye*

such as paranasal sinusitis, hypertension and diabetes mellitus). These latter two groups were associated with a benign outcome and did not need further investigation and had only the ophthalmic division of the trigeminal involved.

Painless presentation of parasellar lesions has been reported previously and indeed the presence or absence of pain and its character have been shown to be unhelpful in predicting underlying diagnosis of parasellar syndromes.[4] Six cases of isolated painless post-ganglionic Horner's syndrome were identified by Grimshaw and Thompson[3] in which there was no cranial nerve involvement, and these were considered to represent a benign syndrome.

In the case of our patient, the diagnosis was delayed for a number of reasons. Isolated trigeminal schwannoma is rare. A survey of the literature shows an incidence of 0.2-0.4% of brain tumours with presentation usually in the third or fourth decade.[5-7] Raeder's syndrome is associated with conjunctival injection and may lead to a misdiagnosis of conjunctivitis.[8] Unilateral allergic eye disease, however, is atypical and the poor response to steroids did not support this diagnosis. It is also unusual for allergic disease in childhood to cause scarring and neovascularisation of the cornea.[9] Reduction in corneal sensation is associated with a number of other diseases, in particular herpetic disease, but this was not clinically evident. On retrospective questioning by the neurologist it was stated by the mother that her daughter had complained of altered sensation around the mouth for some time and that crumbs seemed to gather there without her noticing when she ate, suggesting involvement of the maxillary branch of the trigeminal. Later examination confirmed all three divisions to be involved. It is an important clinical finding remarked upon by Grimson and Thompson in their review of Raeder's syndrome[3] that a dense reduction in corneal sensation demands thorough, careful cranial nerve examination for involvement of the lower two divisions of the trigeminal nerve and subsequent neuroradiological investigation. The displacement of the internal carotid artery by this girl's tumour gave rise to her post-ganglionic Horner's syndrome. The presence of an acquired Horner's syndrome in a child is often, though not invariably, associated with neoplasia.[10] A series of 10 cases of childhood Horner's syndrome from Canada found neuroblastoma in 2 cases, but identified no lesion in 4; 2 had past cardiothoracic surgery and 2 had severe congenital abnormalities.[11]

In conclusion, a careful clinical examination and a high index of suspicion are required to diagnose trigeminal schwannoma because of its rarity and subtle presenting neurological signs early in its course. Despite improved imaging techniques tumours may be large when first scanned.[12] In the series of 27 patients with trigeminal schwannoma seen by Samii *et al.*[5] between 1982 and 1992 the duration of symptoms ranged from 2 months to 6 years. The most frequent symptom was of unilateral pain or numbness of the face. Follow-up at 60 months revealed only 2 cases of recurrence following surgery in this group. MRI scanning is accepted as giving an accurate pre-operative diagnosis of trigeminal schwannoma on the basis of its characteristic dumbbell appearance and also the extent of involvement.[6] MRI may also show wasting of the masticatory muscles due to the trigeminal motor division involvement. Treatment is by surgical excision after careful delineation of the tumour by MRI, CT and angiography, as proximity to the internal carotid artery, spread into the middle ear and towards the cerebellum are possible.

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Sir,

Sinonasal melanoma: an unusual cause of proptosis

Malignant melanomas of the nasal cavity and paranasal sinuses are rare, accounting for less than 1% of all melanomas in the Western population. We report a case of sinonasal melanoma presenting with proptosis and a visible medial canthal mass.

Case report

An 85-year-old man presented to our department with a 3-month history of a pigmented lesion and mild swelling in his left medial canthus. His first symptom had been epiphora 9 months prior to presentation with some swelling of his medial canthus 3 months later associated with some scant epistaxis. Six weeks prior to presentation the epiphora had worsened and he had noticed some proptosis and diplopia.

The patient had worked as a French polisher for many years and was on 5 mg prednisolone daily for polymyalgia rheumatica. Although frail, he had no other specific medical problems.

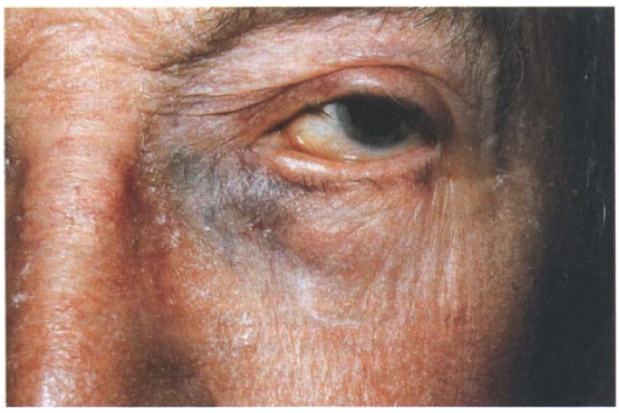
Examination revealed an obvious pigmented mass in the left medial canthus measuring approximately 15 mm × 15 mm. The mass was firm and non-tender, with no associated lymphadenopathy. There was 4 mm of non-axial proptosis with 6 mm of lateral globe displacement and markedly restricted extraocular movements (Fig. 1a). Fundoscopy revealed a raised lesion on the left inferonasal retina, but B-scan ultrasound indicated indentation of the left globe rather than a choroidal mass. MRI scan showed an extensive mass in the left ethmoid sinus eroding into the orbit and indenting the globe. No intraocular mass was evident, nor was there evidence of midline spread (Fig. 1b).

The patient was referred to an ENT surgeon who performed a transnasal biopsy, as the mass was accessible via this route. Histological examination confirmed a malignant melanoma of mixed cell type with positive staining for melanin and positive immunochemistry for S100 and HMB-45 (Fig. 2). Due to the extent of the tumour and the patient's frailty, it was decided that exenteration and block clearance was not indicated. He was therefore referred for palliative radiotherapy.

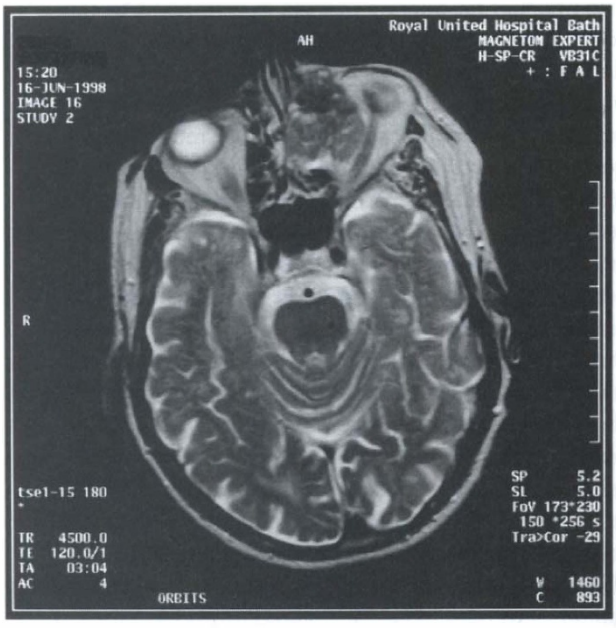
Discussion

Primary malignant melanoma arising from the sinuses and nasal mucosa is rare and carries a poor prognosis. It accounts for less than 1% of all melanomas in Western populations with an apparent higher incidence of 7–11% in Japanese populations.[1] It tends to occur more commonly in elderly people with a peak incidence in the sixth to eighth decades with no difference in sex prevalence.[2] Causal relations are not well established.

Presentation is often varied and occurs late in the natural history of the disease, contributing to a poor prognosis. The most common site of occurrence is the nasal cavity and patients may present with unilateral nasal obstruction and epistaxis.[3] Swelling of the nose and pain are less common presenting symptoms. Various studies have indicated a prevalence of 78% in the nasal cavity and only 5% in the ethmoid sinuses.[4,5]



(a)



(b)

Fig. 1. *(a) Slate-grey medial canthal mass causing proptosis and lateral displacement of the globe. (b) MRI scan showing a left ethmoid mass eroding into the orbit and indenting the left globe.*