

SHORT REPORT

Painful intraorbital meningiomas

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Abstract

Three cases are reported of meningioma involving the orbital apex presenting with either progressive or episodic retro-ocular pain. In two cases pain was steroid responsive and in one of these patients this led to an initial diagnosis of orbital pseudotumour. Pain preceded visual loss and other symptoms by many months and these cases illustrate the importance of full investigation and where necessary surgical exploration in cases of apparent orbital inflammatory disease with an atypical or progressive course.

A recurrent problem in the neuro-ophthalmology clinic is the patient who presents with persistent retro-ocular pain associated with abnormal tissue in the orbit on CT scanning. Although pain alone is rarely a presenting feature of orbital meningioma,¹ we have recently seen three such cases. In two patients the pain responded to steroids and the third initially had periods of spontaneous remission.

Case reports*Case 1*

A 47 year old male presented in August 1988 with a nine month history of worsening right sided headache and retro-ocular pain, with progressive visual loss in the right eye for six weeks. The pain radiated to the right temple and occiput, was not helped by analgesics and was aggravated by pressure over the eyelid. The patient had a longstanding left amblyopia.

On examination he had no perception of light in the right eye with corrected visual acuities on the left of 6/24, N36 with 13/13 Ishihara colour plates correctly identified. The left visual field was normal and fundoscopy showed a pale swollen right optic disc with nerve fibre layer haemorrhages. He had a right relative afferent pupillary defect. Examination was otherwise normal.

Visual evoked potentials (VEPs) were unobtainable from the right eye and of normal latency but slightly reduced amplitude from the left. CT and MRI showed a fusiform swelling of the right optic nerve. A right lateral orbitotomy was performed on 30 September 1988 and biopsy of the lesion showed an optic nerve sheath meningioma with extradural spread.

Severe retro-ocular pain persisted and a left fronto temporal craniotomy was performed on

the 26 October 1988 to identify any intracranial tumour spread. The optic chiasm and optic nerves looked normal; biopsy of the dura overlying the skull base showed no evidence of tumour. Percutaneous radiofrequency rhizolysis of the ophthalmic division of the trigeminal ganglion was performed for persistent pain with some benefit.

Case 2

A 37 year old housewife presented in October 1987 with a six month history of worsening pain in and around the left eye and progressive blurring of vision for three months. She described a throbbing pain as if she had been punched in the eye which radiated to the left face and nose. Analgesics produced only slight relief. Blurred vision had been noticed on waking one morning three months before admission and at its onset was accompanied by double vision with oblique separation of objects. The diplopia disappeared after about four weeks.

Examination of the right eye was normal, visual acuity in the left eye was 3/60 with no colour plates identified. She had a left central scotoma, left relative afferent pupillary defect and optic disc swelling. Elevation and abduction of the left eye were slightly restricted.

VEPs were delayed and of small amplitude from the left eye and normal from the right eye. CT showed swelling of the left optic nerve with no widening of the optic canal. Injection of intravenous hydrocortisone 200mg produced complete pain relief within 20 minutes which lasted for 40 hours. A diagnosis of orbital pseudotumour was made and she was discharged on a reducing schedule of prednisolone starting at 60mg/day.

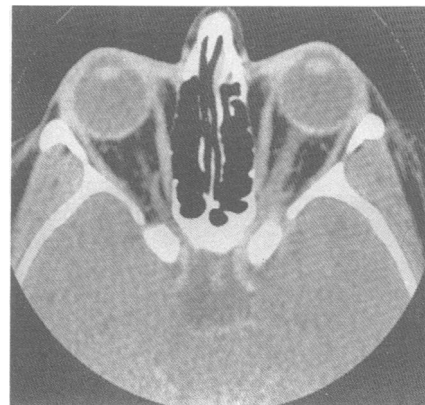


Figure Case 2. Contrast enhanced CT scan showing abnormal tissue in the left orbital apex.

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Received 6 February 1991
and in revised form
18 April 1991.
Accepted 26 April 1991.

Over the next 18 months several attempts to reduce her steroid dose to less than 25 mg/day of prednisolone resulted in a return of the pain. Visual acuities remained unchanged and eye movements returned to normal. A CT scan in January 1989 showed abnormal tissue at the left orbital apex with expansion of the optic canal (figure). In June 1989 pain returned more severely and did not respond to increasing doses of steroids. Visual acuity on the left had reduced to counting fingers at 1m with a left central scotoma and optic atrophy. Neurological signs were otherwise unchanged.

Surgical exploration of the middle cranial fossa was carried out on 15 November 1989 and histological examination of biopsied material showed a meningioma. Her pain improved post-operatively and steroids were stopped over four months.

Case 3

A 40 year old woman was admitted as an emergency in June 1990 with a two week history of progressive visual loss in the left eye and supraorbital pain which was aggravated by eye movement or bright lights. Further questioning revealed that over the preceding two years she had experienced several episodes of left retro-ocular pain accompanied by swelling of the eyelid and more recently by blurring of vision in that eye. These symptoms had all previously resolved within a few weeks.

Visual acuity on the left was limited to finger counting at one metre. Six millimetres of proptosis were evident on the left, the left optic disc was pale, there was a relative afferent pupillary defect and left central scotoma with sensory impairment in the first and second divisions of the left fifth cranial nerve. Examination was otherwise normal.

VEP examination showed no response from the left eye and a normal response from the right eye. Orbital CT showed hyperostosis involving the lateral wall of the left orbit and sphenoid wing with left sided proptosis. A diagnosis of sphenoid wing meningioma was made but as the patient's pain improved after steroids she decided against surgical exploration.

Discussion

These three patients presented with retro-ocular pain, which was progressive in two and episodic in the third and which preceded progressive rapid visual loss by three to 24 months. The pain responded to steroids in two of the patients. Dramatic and immediate resolution of pain in one patient suggested a diagnosis of orbital pseudotumour but progressive visual loss and loss of steroid responsiveness prompted further investigation and biopsy.

Optic nerve sheath meningioma commonly presents with insidiously progressive visual loss or proptosis.¹ Mild and inconsistent headache is described in some patients but significant pain at presentation is rare and occurred in only two of 50 patients with optic nerve sheath meningioma seen at Moorfields Eye Hospital.¹

In contrast other causes of visual loss or ophthalmoplegia often present with pain as an early feature. In an earlier era Holt and Roth recognised syphilitic periostosis, tuberculosis, and nonspecific periostosis as causes of a sphenoid fissure or orbital apex syndrome. Pain was a prominent early feature and remission sometimes occurred.² Hunt *et al* considered recurrent painful ophthalmoplegia with spontaneous remission a distinct clinical entity due to a granulomatous reaction in the cavernous sinus of the type described by Tolosa.^{3,4} Two of the cases described by Hunt *et al* responded to steroids and Smith considered this a diagnostic test for what he termed the Tolosa Hunt syndrome.⁵ Orbital pseudotumour, a term used to describe inflammatory lesions of the orbit of unknown cause, usually presents with rapidly progressive proptosis and eyelid or conjunctival oedema but 23% of patients have pain when first seen.⁶ Painful ophthalmoplegia occurs in a number of other conditions including diabetes, infection, lymphoma, carcinoma, malignant epithelial tumours of the lacrimal gland,⁷ aneurysm and systemic vasculitis.⁸⁻¹⁰ Pain is present in most patients with acute optic neuritis, but is rarely severe.

Although a rapid response to steroids is characteristic of the Tolosa Hunt syndrome this may occur in other conditions including lymphoma,¹⁰ orbital pseudotumour⁶ intracavernous aneurysm,¹¹ sarcoid, chordoma and giant cell tumour.⁹ Our case demonstrates that with meningioma pain relief may be rapid and initially complete. While CT and MRI readily depict abnormal tissue at the orbital apex the diagnosis of orbital pseudotumour is one of exclusion. The cases described demonstrate that orbital meningiomas may present with pain, preceding other symptoms by many months, and that pain relief after steroids may be rapid and complete. When clinical features are atypical or progressive surgical exploration and biopsy are required, either by orbitotomy or via a transcranial approach.¹²

We are grateful to Dr A M Halliday for performing the VEP studies on these patients.

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