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Optic Nerve Sheath Meningioma: Diagnostic Features and Therapeutic Alternatives

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Summary
Twenty-two cases of optic nerve sheath meningioma were reviewed. The clinical features included slowly progressive visual loss in every case. Orbital signs of limited ocular movements and mild proptosis were present in only a third of cases; gaze-evoked amaurosis occurred in 3 cases. The disc was abnormal in every case, usually swollen if vision was 6/12 or better, atrophic if the vision worse than 6/12. Optociliary shunt vessels occurred in 5 patients and were of diagnostic significance. The diagnosis was made by a high-resolution CT scan of the orbits showing tubular expansion of the optic nerve sheath. Calcification of the optic nerve was present in 12 cases. Tubular expansion of the optic nerve sheath may occur in raised intracranial pressure, optic nerve glioma, granuloma, lymphoma or metastatic disease. In the absence of calcification these alternative diagnoses must be considered. Surgery, undertaken for the intracranial component of the tumour, did not halt progressive visual loss.

Optic nerve sheath meningioma characteristically presents with slowly progressive visual loss. The findings of disc swelling or atrophy, optociliary shunt vessels, and a high resolution CT scan showing tubular thickening and calcification of the optic nerves allow a clinical diagnosis.[1–10]

The management of optic nerve sheath meningioma is debated. There have been advocates of fine needle aspiration biopsy.[2] Surgical decompression of the optic nerve has been undertaken,[3] though results of such surgery have been disappointing.[4] Excision of the nerve has been recommended because of the aggressive nature of the tumour in young patients.[5,6] However, in view of the operative and visual morbidity of biopsy, decompression or excision, a conservative approach may be warranted.

Twenty-two cases of optic nerve sheath meningioma, seen over the past 8 years at the Department of Neuro-Ophthalmology, were reviewed. The purposes of the review were to identify the diagnostic clinical and radiological features and to establish whether cases managed conservatively or surgically fared differently.

Patients and Methods

The clinical diagnosis was based on a history of progressive visual loss, changes at the optic disc, and a high resolution CT scan of the orbits demonstrating tubular optic nerve enlargement. Twenty-two patients with a clinical diagnosis of optic nerve sheath meningioma were studied: eleven involved the left optic nerve, six involved the right, and five were bilateral. Histological confirmation of the diagnosis was obtained in 10 patients. The age of onset of symptoms ranged from 4 to 64 years

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(median 29); in 18 patients (81 per cent), symptoms developed below the age of 40 (Fig. 1). There were 14 females (63 per cent), and 8 males.

Visual loss was the presenting symptom in 21 patients (95 per cent); the remaining patient was not aware of decreased vision in the affected eye. Nine patients (40 per cent) complained of headache; minimal proptosis was present in 8 patients (36 per cent); limited ocular movements, particularly upgaze, were found in 9 patients (40 per cent). Three patients (14 per cent) complained of loss of vision on ocular movement.

Visual acuity

When first seen at the National Hospital, visual acuity in the affected eyes varied from 6/5 to no perception of light. In 15 eyes (55 per cent), visual acuity was 6/12 or better; in 6 eyes (22 per cent) visual acuity was counting fingers or better; in 6 eyes (22 per cent) vision was hand movements or worse. In 6 of the 15 eyes with good vision (6/12 or better), colour vision was impaired when tested with the Ishihara colour plates.

Visual fields

Most commonly, the visual field was contracted, this being the main finding in 11 eyes (41 per cent). In 8 eyes there was a central scotoma (30 per cent).

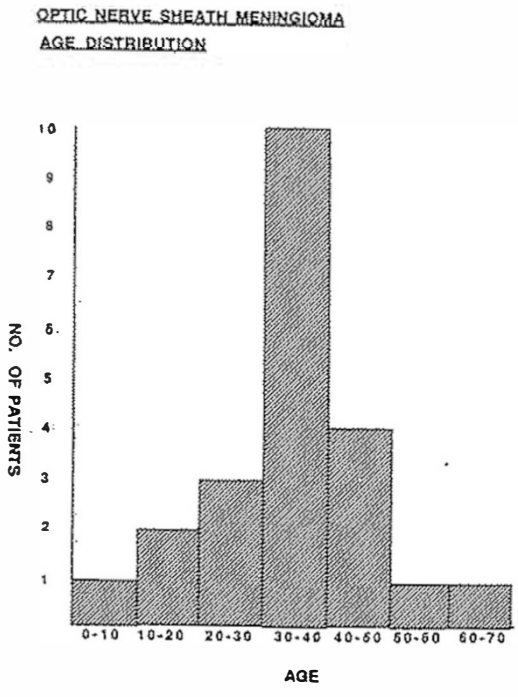

Fig. 1. *Histogram showing the age at presentation of signs of optic nerve sheath meningioma.*

In 4 eyes there was an altitudinal or arcuate field defect (15 per cent). In 3 eyes there was no perception of light at presentation (11 per cent). In 1 eye there was enlargement of the blindspot (3 per cent).

Optic disc changes

On presentation the optic discs were atrophic in 17 eyes (63 per cent); the discs wereswollen in 10 eyes (37 per cent). In 15 eyes with 6/12 vision or better, the discs were swollen in 8 and atrophic in 7. In 12 eyes with less than 6/12 vision, 11 discs were atrophic and 1 was swollen. Optociliary shunt vessels were present in 7 eyes (26 per cent) of which 4 had swollen and 3 had atrophic discs (Fig. 2). Refractile bodies on the disc were present in 3 eyes (11 per cent), 2 being atrophic, one swollen.

CT scan appearances

CT orbital scans in the most favourable axial and coronal views were performed in all patients. The optic nerve was diffusely enlarged in a tubular pattern in all patients and was calcified in 12 (54 per cent), including all bilateral cases (Fig. 3).

In this series, there were no patients with neurofibromatosis. Eleven patients (50 per cent) had intracranial involvement at presentation diagnosed either radiologically or at surgery. (Fig. 4). Of the 10 patients who had orbitocranial surgery, seven had evidence of intracranial involvement. At surgery, 3 cases had no evidence of intracranial involvement.

Progress

Seventeen patients have been followed, over periods with a range of 1–11 years (median 4.5). In this group of patients, there were 19 eyes with some vision remaining at presentation. Six of these patients had orbito-cranial surgery while they still retained some vision. The deterioration in vision is illustrated in Figure 5; the vision declined in those patients managed conservatively and those who had surgery. Of the patients who had surgery, three were troubled by epilepsy post-operatively. Recurrence of tumour, either intra-orbital or intra-cranial, occurred in 5 of the 10 patients who had surgery.

Two patients with bilateral optic nerve meningioma were treated by radiotherapy to the orbits when visual acuity began to decline in their only seeing eye. One patient had 1,325 cGy to both orbits and the chiasm in 4 treatments, but radiotherapy was terminated because his vision declined during treatment. The other patient received 4000 cGy to both orbits and chiasm in 12 treatments. There was no improvement in acuity, but two months later, the field had improved slightly.

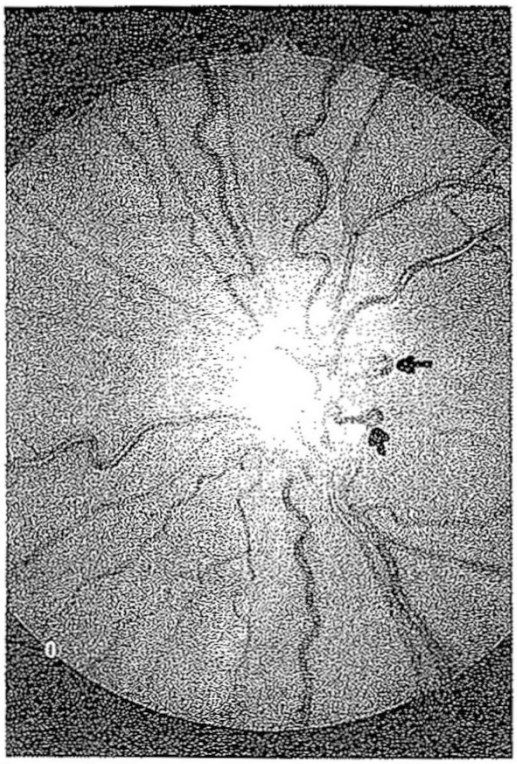

Fig. 2. *Photograph of left optic disc showing pallor of the disc and opto-ciliary shunt vessels (arrowed).*

Unfortunately, the field declined further within the next two months.

Discussion

This paper will consider the diagnostic criteria of optic nerve sheath meningioma in the light of the sophisticated neuro-radiology now available, and evaluate the role of surgery.

Visual loss was the only symptom in almost two thirds of this series and the absence of other orbital signs probably led to the delay in diagnosis which characterises the condition. In all cases, the disc was swollen or atrophic in the affected eye. Optociliary shunt vessels occurred in five of our patients (22 per cent) (7 eyes), similar to the proportion of patients in the series reported recently by Sibony *et al.*[7] The association of optociliary shunt vessels with orbital meningioma has been known since 1898 when Elschnig traced the shunt vessels from the retina through the choroid into the vortex veins.[8] When the retrobular central retinal vein becomes congested, bypass channels develop between the central retinal vein and the peripapillary choroidal capillaries. Optociliary shunt vessels may occur in other causes of chronic disc swelling so, alone, are not pathognomonic of optic nerve sheath meningioma.[9]

Transient visual loss on ocular movement or gaze-evoked amaurosis has been recorded in several cases of optic nerve sheath meningioma.[4,7] Constriction of the central retinal artery and splinting of the ocular muscles producing a rise in intraocular pressure are the probable mechanisms for this symptom. Gaze-evoked amaurosis was recorded in 3 of these patients. Gaze-evoked amaurosis is not pathognomonic of optic nerve sheath meningioma since it has also been reported in optic nerve glioma.[10]

With the advent of high-resolution CT scanning of the orbits, reliable diagnosis of optic nerve tumours became possible without resorting to biopsy. Optic nerve sheath meningioma may be distinguished from optic nerve glioma on CT scan appearances. In a series reported by Jakobiec *et al.*,[10] biopsies

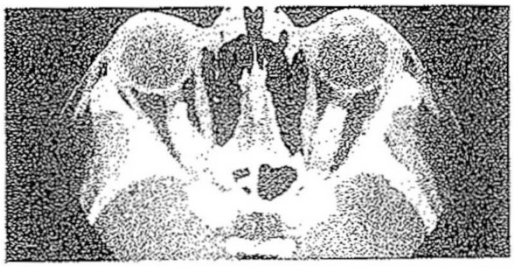

Fig. 3. *CT scan of orbits showing enlargement of both optic nerve sheaths with calcification, characteristic of optic nerve sheath meningioma.*

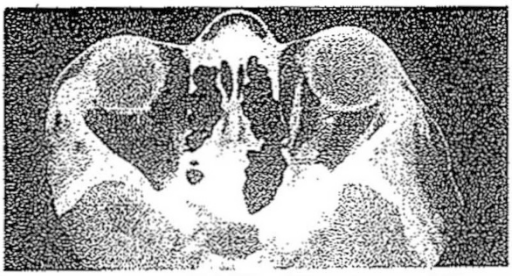

Fig. 4. *CT scan of orbits showing sphenoid ridge hyperostosis and thickening of the left optic nerve, indicating sphenoid ridge meningioma with extension along the ipsilateral optic nerve.*

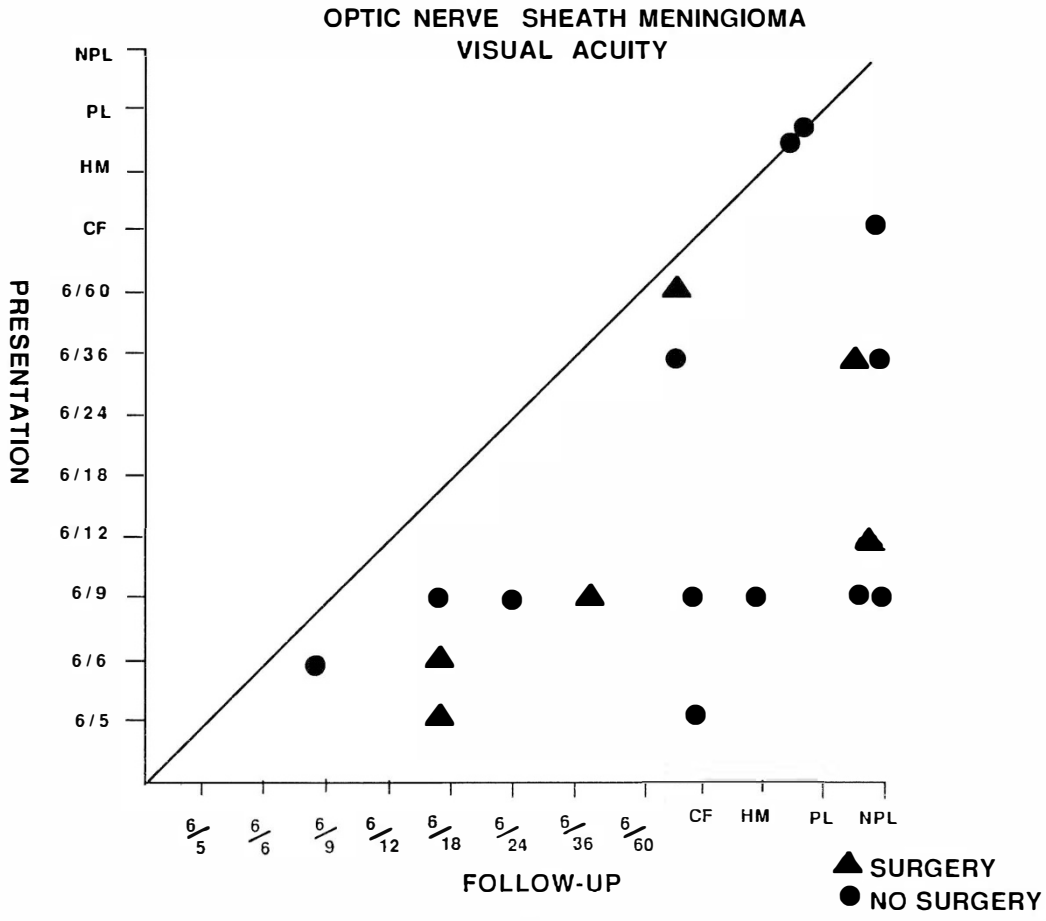

Fig. 5. *Graph of visual acuity at presentation and latest follow-up to show deterioration in acuity in group treated surgically and group managed conservatively.*

were performed on 18 of 22 patients with optic nerve glioma and 30 of 47 patients with optic nerve sheath meningioma; CT scan patterns of enlargement of the nerve were characterised for each diagnosis and no pattern was included without biopsy confirmation. Nineteen of 22 optic nerve gliomas showed fusiform expansion of the nerve or cystic degeneration. Four patterns typical of optic nerve sheath meningioma were defined:

(1.) Diffusely enlarged nerve wtih posterior expansion at the apex of the orbit (16 cases).

(2.) Diffusely enlarged optic nerve often with a central negative shadow outlining the optic nerve (11 cases).

(3.) Calcification within the optic nerve (10 cases).

(4.) Diffuse, irregular expansion (7 cases).

However, diffusely enlarged optic nerves are not pathognomonic of optic nerve sheath meningioma. Diffuse, narrow expansion was found in two gliomas in Jakobiec's series and we have seen it in one case. Diffuse expansion of the optic nerves may also be seen in papilloedema due to raised intracranial pressure (Fig. 6). Unilateral disc swelling may be the only sign of raised intracranial pressure; in Jakobiec's series, it is not stated whether cases of optic nerve sheath meningioma which were not biopsied had lumbar punctures to measure the intracranial pressure. Diffuse

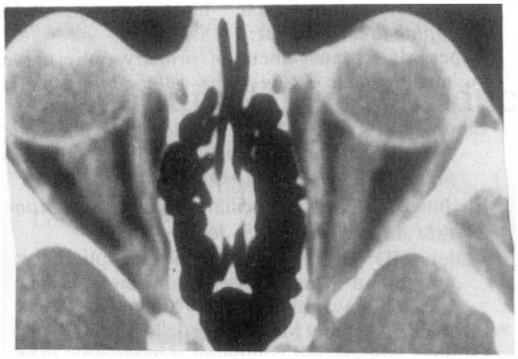

Fig. 6. *Ct scan of orbits showing enlargement of the optic nerve sheaths in a case of benign intracranial hypertension.*

enlargement of the optic nerve may also occur in orbital pseudotumour, lymphoma, sarcoidosis, or metastatic carcinoma. In all these diagnoses, the history of visual loss is usually acute not chronic. In pseudotumour, pain is often present and thickening of the posterior aspect of the globe may be a clue to the presence of an associated posterior scleritis. A full general examination and routine haematology, biochemistry, a chest *x* ray and a lumbar puncture should help exclude lymphoma, sarcoid or carcinoma.

In Jakobiec's series, no calcification was seen in glioma—despite evidence that calcospherites have been identified histopathologically in glioma.[11] Calcification of the optic nerve on CT scanning has been said to be diagnostic of optic nerve sheath meningioma.[12] The presence of calcification is not in itself a guide to the vision in the eye; one of our cases had no perception of light in one eye and 6/5 vision with a full field in the other, yet the scan appearances of the two nerves were similar.

A review of the literature emphasises the uniformly poor visual prognosis of optic nerve sheath meningioma with a slow painless inexorable decline. Surgery undertaken for the intracranial component of the tumour in our cases did not halt or prevent this decline. Slitting the optic nerve sheath to preserve vision has not usually been successful and has provoked orbital recurrence.[4,7]

Prognosis for survival of the adult patient should be good; the tumour is peripheral and slow growing. Even if intracranial invasion occurs, the risks of radical surgical removal may out-weigh the benefit. Surgery itself may alter the natural history of the tumour so that it becomes more aggressively invasive locally.

In children, Walsh,[13] and more recently, Karp,[5] and Alper[6] have argued that the tumour behaves more aggressively. Karp reviewed cases stretching from 1927 to 1970 from the files of the Armed Forces Institute of Pathology; of 8 cases presenting under the age of 20 years, only one was known to be alive without recurrent tumour. Four cases had orbital recurrence; two cases died from surgical complications of removal of posterior fossa tumours; one case died from complications of intracranial extension. The high mortality may reflect earlier surgical techniques and may be associated with the existence of second tumours. Alper, in a review of 55 cases culled from the same source, found that 3 patients out of 29 presenting under the age of 35 died from intracranial extension, three died from associated causes and twelve had orbital recurrence. These figures were compared with 26 cases presenting after the age of 35 years, of whom none died of intracranial extension but five died of associated causes and two had orbital recurrence. Since no indication of the length of follow-up was given for either group, these figures may not be strictly comparable.

In this series, follow-up information is too short to indicate the prognosis for survival of patients with optic nerve sheath meningioma whether managed conservatively or surgically. There is no indication that patients who have not had surgery have survived for shorter periods. In the group of patients who had surgery for intracranial extension, postoperative epilepsy and orbital recurrence were not uncommon. Further follow-up information on all these patients will be of great interest.

The value of radiotherapy in optic nerve sheath meningioma is questionable. One report documented improvement in visual function in 4 out of 5 cases of optic nerve sheath meningioma treated by radiotherapy.[14] These results have not been repeated. One of our cases showed an initial increase in the size of the visual field after radiotherapy; the initial effect of radiotherapy would be

increased vascularity of the tumour and it is tempting to speculate that this produced the encouraging response. However the field subsequently deteriorated further so the long-term results of treatment were disappointing.

Meningiomas are hormonally responsive.[15] Two of our cases reported that vision varied during the menstrual cycle or declined during pregnancy. Recently cell culture has established that the growth of certain meningiomas can be influenced by certain steroids.[16] To date, there are no reports of benefit from endocrine manipulation in these patients though Medroxyprogesterone has been tried.[17] We may hope that drugs will be found which can be used to treat these tumours which are both safe and effective.

I am most grateful to the physicians and surgeons of the National Hospital for Nervous Diseases for allowing me to study their patients, and especially to Mr. M. D. Sanders for his help in the preparation of this paper. I thank Mr. Richard Dewhirst at the National Hospital and the Department of Medical Illustration at St Thomas' Hospital for the illustrations.

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