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Primary optic nerve meningioma

J. E. WRIGHT, N. B. CALL,* AND S. LIARICOS†
From the Orbital Clinic, Moorfields Eye Hospital, City Road, London EC1V 2PD

SUMMARY Twenty-seven patients with a meningioma arising from within the optic nerve sheath have been seen during the past 12 years. Twenty-one were women, the majority aged 39–64 years. The men were younger, all except 1 being between 20 and 41 years when first seen. There were no patients younger than 20 years. Twenty-two patients had noticed a deterioration in the vision as their initial symptom. The optic nerve head was abnormal in all patients; 14 were swollen and 13 atrophic and flat. Neurofibromatosis was not associated with this condition. Treatment was essentially surgical.

Primary orbital optic nerve meningiomas arise from the cap cells of the arachnoid round the intra-orbital portion of the optic nerve. More rarely meningiomatous tissue may extend from adjacent structures into the orbit in the subdural space of the optic nerve (Figs. 1 and 2), a secondary optic nerve meningioma. The true origin of any meningiomatous tissue found in relation to the optic nerve must therefore be defined accurately.

Failure to do this has caused some confusion about the symptomatology and incidence of the various types of meningioma affecting the orbit. This communication analyses the mode of presentation of a series of patients with a proved primary optic nerve meningioma, correlating the signs and symptoms with the findings at surgery.

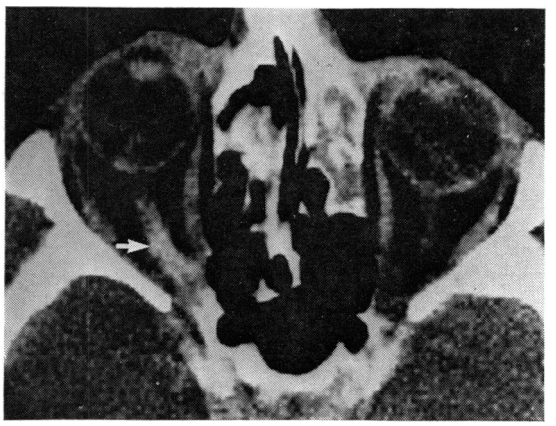

Fig. 1 *Axial CT scan showing enlarged optic nerve (arrowed) caused by secondary meningioma.*

Patients and methods

Twenty-seven patients with a primary optic nerve meningioma were seen in the Orbital Clinic at Moorfields Eye Hospital since 1968. A detailed clinical history was obtained in all cases and a thorough ophthalmic and neurological examination carried out, including a search for signs of neurofibromatosis. Oblique radiographs and axial hypocycloidal tomograms of the optic canals were obtained in all patients. Since 1964 patients have been CT scanned in the axial plane; recently coronal scans have been taken. These radiographic investigations were supplemented in some cases by b and c mode ultrasonic examination of the optic nerve and orbital contents. Fluorescein angiograms were performed on selected patients to define the state of the optic disc.

All patients included in this series were microsurgically explored by lateral orbitotomy.[1] In most patients the meningioma was totally removed, leaving the optic nerve intact. Latterly the optic nerve has also been excised from the globe to the optic foramen. Patients with useful vision had biopsies taken and were decompressed by incising the dural sheath. Later, when vision failed, the lesion was excised totally.

Patients have been reviewed at 6-monthly or yearly intervals. Axial hypocycloidal tomography and CT scans have been performed on selected patients.

Correspondence to Mr John E. Wright, Moorfields Eye Hospital, City Road, London EC1V 2PD.

*Present address: 324 Tenth Avenue, Suite 185, Salt Lake City, UT 84103, USA.

†Present address: Department of Ophthalmology, Greek Anticancer Institute, St Savvas Hospital, Athens, Greece.

Results

Some of the clinical details of the 27 patients studied are presented in Table 1. Twenty-one patients were women. Twenty of these were aged

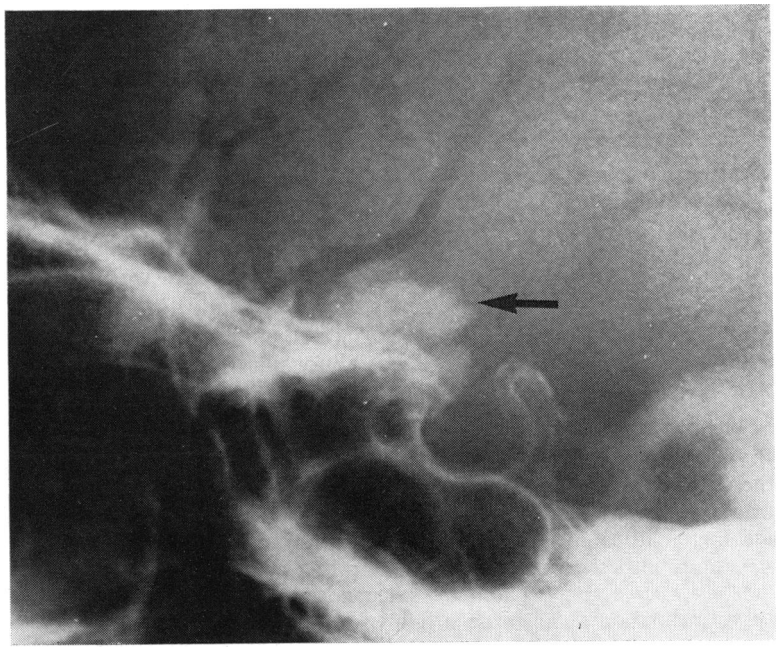

Fig. 2 *Radiograph from the same patient shown in Fig. 1, with sclerotic changes on the planum sphenoidale (arrowed).*

Table 1 *Clinical details of 27 patients*

Patient no.	*Age (yr)*	*Sex*	*Side*	*Initial symptoms*	*Proptosis*		*Optic disc*	*Visual acuity*
					Degree	*Duration*		
1	50	F	L	Visual loss, 18 mo	None	None	Swollen	Counts fingers
2	31	M	L	Visual loss, 7 yr	11 mm	6 yr	Swollen	Light perception
3	49	F	L	Visual loss, 5 yr	7 mm	8 mo	Atrophy	Counts fingers
4	64	F	R	Lid swelling, 1 yr	7 mm	1 yr	Atrophy	Counts fingers
5	43	F	R	Visual loss, 1 yr	6 mm	6 mo	Swollen	6/36
6	61	F	R	Visual loss, 3 yr	4 mm	2 mo	Atrophy	Counts fingers
7	49	F	R	Visual loss, 18 mo	2 mm	Not noticed	Swollen	6/18
8	50	F	L	Visual loss, 3 yr	2 mm	Not noticed	Atrophy	Hand movements
9	47	F	R	Diplopia, 1 yr	3 mm	9 mo	Swollen	6/9
10	28	F	R	Visual loss, 6 mo	4 mm	3 mo	Swollen	Light perception
11	41	M	L	Visual loss, 1 yr	2 mm	Not noticed	Swollen	6/60
12	39	F	L	Visual loss, 4 yr	5 mm	1 yr	Atrophy	Light perception
13	63	F	L	Visual loss, 1 yr	2 mm	Not noticed	Swollen	Light perception
14	37	M	R	Intermittent visual loss, 6 mo	None	—	Swollen	6/6
15	46	F	L	Proptosis, 6 mo	3 mm	6 mo	Swollen	Hand movements
16	48	F	L	Visual loss, 2 yr	3 mm	Not noticed	Atrophy	Counts fingers
17	48	F	L	Visual loss, 4 mo	2 mm	Not noticed	Atrophy	6/60
18	40	F	L	Visual loss, 6 yr	5 mm	3 yr	Atrophy	Hand movements
19	52	F	R	Visual loss, 4 yr	3 mm	Not noticed	Atrophy	No perception of light
20	40	F	L	Proptosis, 18 mo	4 mm	18 mo	Atrophy	6/9
21	62	M	L	Diplopia, 3 yr	2 mm	Not noticed	Atrophy	6/9
22	45	F	L	Intermittent visual loss, 5 yr	5 mm	2 yr	Swollen	6/12
23	63	F	R	Visual loss, 12 yr	3 mm	8 yr	Swollen	Counts fingers
24	54	F	L	Visual loss, 3 mo	2 mm	Not noticed	Swollen	6/60
25	40	F	L	Visual loss, 3 mo	3 mm	Not noticed	Atrophy	Hand movements
26	20	M	R	Visual loss, 6 mo	2 mm	Not noticed	Swollen	6/18
27	20	M	R	Visual loss, 3 yr	6 mm	2 yr	Atrophy	6/18

between 39 and 63 years. The exception was a 28-year-old woman who developed a rapidly progressive proptosis during pregnancy (case 10). The 6 men were younger; 5 were between 20 and 42 years; the oldest patient was 62 years old. No systemic abnormalities were found in any of these patients. Specifically no patient had signs of neurofibromatosis.

Twenty-two patients experienced a deterioration in vision of the affected eye as their initial symptom. In 20 deterioration was progressive and inexorable. Two patients experienced intermittent visual loss. One, a van driver (patient 14), noticed that his vision failed when the eye was abducted; vision returned to normal within a few minutes of his eyes assuming the straight ahead position. Five patients initially experienced no visual deterioration. Three noticed proptosis followed after an interval by visual loss. Two patients experienced double vision; one developed proptosis within 3 months, and the other had double vision for 2 years before her vision deteriorated.

Most of the patients had very poor vision in the affected eye. However 8 patients had 6/24 vision or better when they were first seen. Within 18 months their vision had deteriorated to worse than 6/60.

Twenty-five patients had a relative proptosis of 2 mm or more , with a range of 2 mm–11 mm (mean 4 mm). There was some degree of restriction of movement of the eye in 19 patients, with upward movement particularly affected in 12 patients.

The optic disc was abnormal in all patients. Fourteen were swollen, 13 were atrophic and flat. Opticociliary shunts were present in 9 patients.

Radiographs of the bones surrounding the orbit were normal in all except 2 patients, who had slight enlargement of the anterior end of the optic canal shown by axial hypocycloidal tomograms. CT scans accurately delineated the extent of the optic nerve enlargement and defined the abnormal optic nerve in the coronal view (Fig. 3).

The lateral orbitotomy approach gave an excellent exposure of the optic nerve from the globe to the optic foramen. Six patients with retained central vision were decompressed by incising and removing a portion of dura overlying the tumour. The visual deterioration progressed in all patients, and useful vision was lost within 18 months. One patient with poor vision was explored and a large quantity of cerebrospinal fluid (CSF) was released once the dura was incised. No orbital tumour was found, and the lesion was thought to be an arachnoid cyst. Subsequently the proptosis increased. CT scans showed a mass in the apex of the orbit, which was a meningioma. All these patients were re-explored and the meningioma excised. Case 20 was

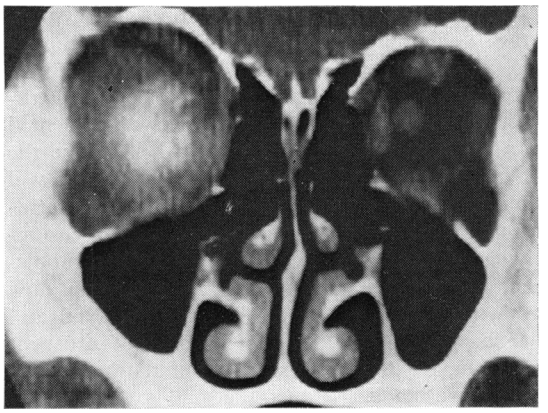

Fig. 3 *Coronal CT scan showing a large optic nerve meningioma with central calcification.*

an exception. Fifteen months after decompressive surgery the tumour had infiltrated the structures of the posterior third of the orbit, so that local excision with preservation of the globe was impossible. The patient awaits exenteration.

The meningioma with or without the optic nerve was excised as the primary procedure in 18 patients with poor vision when first seen.

There has been one recurrence (patient 10). A large angioblastic meningioma was removed, leaving the optic nerve intact. Six years later the proptosis recurred. A diffuse meningioma filled the inner surgical space; it was removed by partial exenteration.

Discussion

Orbital meningiomas are either primary or secondary depending on their site of origin. Primary meningiomas arise either from the cap cells of the arachnoid surrounding the nerve, that is, a primary optic nerve meningioma, or, more rarely, from cells external to the dura of the nerve but within the orbital periosteum, the 'extradural orbital meningioma' described by Craig and Gogela.[2]

Secondary meningiomas extend into the orbit from adjacent structures by expansion of the sphenoid or less commonly the frontal bone. Occasionally a meningioma may originate in the middle fossa and enter the orbit through the superior orbital fissure or extend down the optic canal and enter the orbit in the subdural space. This latter type of presentation accurately mimicks a primary optic nerve meningioma.

Primary optic nerve meningiomas can therefore be diagnosed only by accurately defining the origin of meningiomatous tissue found within the dural sheath of the optic nerve. The diagnosis is one

of exclusion, and it can be made only if the structures surrounding the orbit are normal. The clinical and radiological assessment of these patients must therefore be extremely thorough. Routine postero-anterior, lateral, and optic canal views augmented by axial hypocycloidal tomography will define the sphenoid bone, and high resolution CT scans will confirm these findings and often show the intracranial extent of a sphenoid meningioma. Occasionally carotid angiography may be needed to supplement these investigations. CT scanners can now show the orbital structures in both the axial and coronal planes and provide pictures of excellent definition, so that ultrasonic examination of the optic nerve has been superseded.

The predominant feature of optic nerve sheath meningiomas was early visual loss. Proptosis, when it occurred, was noticed later and was often of small degree. This type of presentation is in direct contrast to tumours which arise outside the dural sheath, for they usually cause considerable proptosis before compressing the optic nerve; the exception is an extradural tumour in the apex of the orbit. The reason for the difference is the site of origin and mode of growth of an optic nerve meningioma. The tumour arises between the dural sheath and the pia covering the optic nerve. As it grows it is confined by the tough dura and compresses the nerve, causing a dense central scotoma which usually spreads to the more peripheral parts of the visual field. This pattern of change is not universal, for 5 patients retained a visual acuity of 6/12 or better although 4 had peripheral field loss. Meningiomatous tissue probably produces visual field changes by a combination of pressure and interference with the blood supply to the optic nerve, for decompression of the nerve did not prevent the visual deterioration progressing in any patient.

The spread of the meningiomatous tissue within the dural sheath has secondary effects on the optic nerve head. All patients had a swollen or atrophic optic disc when first seen, and in many cases opticociliary shunt vessels were present. Meningioma involved the optic nerve immediately behind the globe in those patients with opticociliary shunts.[3 4] Occasionally forward extension of meningiomatous tissue is so extensive that it invades the optic nerve head.[5]

The restriction of movement of the eye seen in most patients is probably caused by splinting of the optic nerve by tumour spread within the dural sheath. It is difficult to explain why upward movement is especially affected. When the patient attempts to move the eye, there is often an accompanying rise in the intraocular pressure. This is probably caused by the vigorous contraction of the extraocular muscles attempting to overcome the mechanical resistance provided by the thickened and abnormal optic nerve. The slight proptosis observed in many of the patients can be attributed to the straightening of the normal S shape of the optic nerve within the orbit. More marked proptosis occurs if the tumour ruptures through the dura and forms a space-occupying lesion within the muscle cone.

Two patients experienced intermittent loss of vision initially. In 1 (patient 14) vision was lost when the eye was abducted. Fluorescein angiography showed that the visual loss was caused by interruption of retinal arterial flow. Two factors were probably responsible for this. The meningioma was fibrous and surrounded the point of entry of the central retinal artery so that any movement of the eye constricted the artery. The splinting effect of the tumour also produced a rise in the intraocular pressure. Thus, the perfusion pressure within the artery was reduced and the external pressure on the central retinal artery increased. Despite a decompression of the optic nerve, together with a Scheie thermal sclerostomy to lower the intraocular pressure, the obscurations gradually became more prolonged and more easily provoked. After 12 months the eye became blind and the optic disc atrophic with opticociliary shunt vessels.

A peculiar effect of the spread of tumour within the dural sheath was seen in patient 18. At the initial operation the distention of dura by a large quantity of CSF was thought to be an arachnoid cyst.[6] The patient's proptosis increased and the vision deteriorated further. During this time high resolution CT scanning had been developed, and this showed a mass in the apex of the orbit. A primary optic nerve meningioma was found surrounding the posterior third of the optic nerve. This had presumably caused CSF to accumulate in the anterior part of the subarachnoid space.

In a previous report from this hospital[7] attention was drawn to the infrequency of optic nerve meningiomas in children. In that communication the only child was a boy of 11 who was originally diagnosed as having a primary optic nerve meningioma by an ophthalmologist, who removed the boy's left eye because it was blind and grossly proptosed. Meningiomatous tissue surrounded the excised optic nerve. Three years later, when he was first seen in this clinic, meningioma was present in the middle cranial fossa and was affecting his right optic nerve. During the past 4 years the tumour has extended into the right orbit subdurally and has also involved the chiasm, so that he is now blind. The origin of this tumour is therefore uncertain, for it could have originated on the planum sphenoidale and spread down both optic nerves rather than beginning in

the left optic nerve sheath.[3 4] The patient has not been included in this present series. A number of similar 'bilateral optic nerve meningiomas' have been seen during the period of this survey. In several there was radiological evidence that the tumour originated on the planum sphenoidale with subsequent subdural spread into each orbit. A report on the clinical and radiological features of this group of meningiomas is in preparation.

The age range and sex ratio of our patients corresponds very closely with those of other clinical series.[2 8 9] These have shown that primary optic nerve meningioma occurs predominantly in middle aged women and is rarely encountered in children. Karp *et al.*[10] reporting 25 patients considered to have a primary optic nerve meningioma found that 40% of their cases occurred in children and young adults below the age of 20. They concluded that primary optic nerve meningioma was less rare in children than has been generally assumed. It is difficult to reconcile their statistics and conclusions with our experience and that of others. It is possible that during the 40-year period of their study specimens from 10 children with definite primary optic nerve meningiomas were sent to the laboratory, whereas only a small proportion of tumours occurring in adults were referred. The selection of only 21 cases from a total of 81 on file at the registry could also have influenced the statistics. Another possibility is that the meningiomatous tissue encountered in the orbit may not have originated in the optic nerve. The pathologist is dependent on the accuracy of the clinical details and diagnosis supplied by surgeons, who may be unfamiliar with orbital disease. Some of the patients detailed by Karp and his associates were first seen at a time when investigative methods were comparatively primitive, and the clinician could not have been sure of the site of origin of the original tumour or the recurrences which occurred in 6 of the children.

That optic nerve meningiomas do occur in children is beyond doubt. A number of well documented cases have been recorded by several authors. Walsh[11] reported 7 children with this lesion with local recurrence and extension into the optic canal in several patients. He thought that these tumours were aggressive and often heralded the development of central neurofibromatosis. He suggested that diagnosis of the condition was often delayed because a child, particularly one with neurofibromatosis, would be assumed to have a glioma affecting the nerve. He therefore advocated doing a biopsy to differentiate between the two conditions.

Patients with a primary optic nerve meningioma should have a good prognosis, for the tumours are peripheral, slow growing, and isolated from the central nervous system. Henderson[8] has reported 9 adults with survival up to 19 years after the initial diagnosis was made. Two patients not included in our series have been treated conservatively without biopsy because the patients were elderly and there was little proptosis. One has shown no change over a period of 8 years.

Surgery in the patients reported in this paper has been directed towards total removal of the meningioma once useful vision had gone; biopsy and decompression of the optic nerve where vision remained; or, if there was a small anteriorly located tumour, its total removal with preservation of vision. A dilemma arises in judging whether to explore the affected optic nerve once the provisional diagnosis has been made. If the eye is blind, removal of all the meningiomatous tissue prevents growth of the tumour within the orbit or along the optic canal to the chiasm and should improve an already favourable prognosis. If useful vision remains, incising the dura should relieve the pressure of the tumour on the optic nerve. The results of this procedure have, however, been disappointing. In all patients the visual deterioration continued and useful vision was destroyed within 18 months, so that another lateral orbitotomy was required to excise the meningioma. In 1 patient (case 20) the attempt at decompression probably allowed the tumour to spread widely into the posterior half of the orbit, so that an exenteration will be needed in the near future to remove a tumour which was previously locally resectable.

We now believe that the most appropriate management for patients with relatively good vision is to wait for vision to deteriorate and then excise the optic nerve together with the meningioma. Biopsy or any surgery which transgresses the dura should be avoided. There are 2 exceptions. First, if the rate of growth of the optic nerve tumour suggests a malignant type of meningioma, the tumour should be biopsied. Secondly, if high resolution CT scans show a small, very anterior tumour and useful vision remains, the nerve should be explored, for this type of lesion can be removed without destruction of vision as in patient 2. The results of the type of treatment we advocate can be judged only with a follow-up of very many years.

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