

Primary orbital schwannomas

JACK ROOTMAN,¹ CHAIM GOLDBERG,² AND WILLIAM ROBERTSON³

From the ¹Departments of Ophthalmology and Pathology, University of British Columbia and Vancouver General Hospital; the ²Department of Ophthalmology, University of British Columbia and Vancouver General Hospital; and the ³Department of Radiology, Vancouver General Hospital, Canada

SUMMARY Seven histologically proved cases of primary orbital schwannoma have been seen at the University of British Columbia Orbital Clinic between September 1976 and December 1980. We describe here their varied clinical presentations, preoperative investigations, operative findings, and appearances on light and electron microscopy. Although no single feature is pathognomonic, a multiplicity of clinical, radiographic, and surgical features point to this lesion. Of preoperative investigations the computed tomography scan was the most helpful, especially in localising the lesion. Of the 7, 4 were intraconal and 3 were extraconal. The surgical approach was dictated by tumour site and included anterior, lateral, and panoramic orbitotomies. At surgery the nerve of origin of 4 of the tumours was identified. All tumours were excised totally or subtotally. There has been no recurrence to date.

Schwannomas account for 1% of orbital tumours^{1,2} and may arise anywhere within or adjacent to the orbit.³ We have seen 7 histologically proved cases of schwannomas in the Orbital Clinic at the University of British Columbia between September 1976 and December 1980. We describe here their varied clinical presentations, preoperative investigations, operative findings, and appearances on light and electron microscopy.

Materials and methods

During the period from September 1976 to December 1980 484 cases were seen in the Orbital Clinic at the University of British Columbia, of which 123 represented mass lesions. Seven of these were found to be schwannomas, representing 1.4% of all orbital cases seen and 5.7% of mass lesions.

Diagnostic investigations included computed tomography (CT) scan (EMI 1010S or GE 8800), A and B ultrasonography, and skull series. Tomography and contrast arteriography were used when indicated.

Surgical approaches included lateral, anterior, and panoramic orbitotomies, depending on the specific preoperative localisation.

Postoperatively the excised tumour was submitted for light and electron microscopic analysis.

CLINICAL PRESENTATIONS

Of the 7 cases 4 were intraconal and 3 were extraconal. The latter group consisted of the following sites: ethmoid sinus, superior orbit and upper lid, and inferoanterior orbit. Five case histories will be presented.

Case reports

CASE 1

A 60-year-old Caucasian man had noted slowly progressive right proptosis from age 24. There were no other ophthalmic complaints until August 1976, when he complained of blurred vision of his right eye followed by intermittent horizontal diplopia. In addition he had occasional frontal pain and tenderness.

Best corrected right visual acuity was 6/7.5. The right eye was 8 mm proptotic and displaced laterally 2 mm (Fig. 1). The orbit was ballotable and the globe could be retropulsed. He had a right hypertropia of 2 prism dioptres in up gaze and a 4 prism dioptre exotropia in left lateral gaze. The right optic nerve head appeared somewhat pale. The remaining ophthalmic and physical examination was unremarkable.

CT scan showed a smooth, well-defined, oval, intraconal mass lesion in the posterior one-half of the right orbit. It demonstrated mild, uniform enhancement, and a focal area of calcification was seen centrally in the mass (Fig. 2). Orbital x-rays were

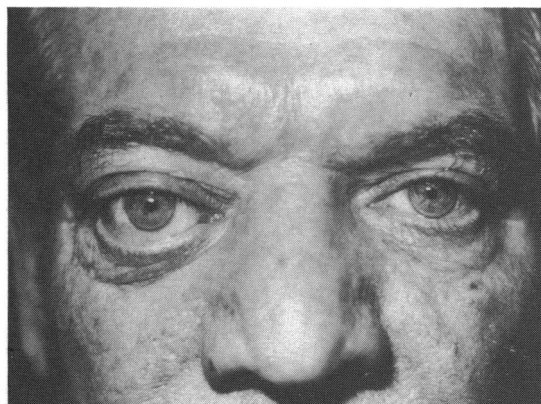


Fig. 1 Case 1: 60-year-old male with long-standing right proptosis, showing 8 mm axial and 2 mm lateral displacement of the globe.

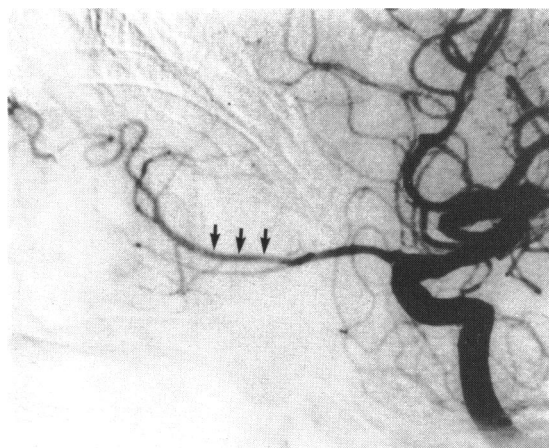


Fig. 3 Case 1: Carotid arteriogram, subtraction view, demonstrating inferior displacement of ophthalmic artery with no tumour blush.

normal. Carotid angiography demonstrated inferior displacement of the ophthalmic artery and its lateral branch, with no tumour blush (Fig. 3).

Ultrasonography revealed an intraconal mass lesion with a well-demarcated anterior border. Considerable sound attenuation within the tumour indicated a relatively solid nature. Some cystic areas were also present.

The patient underwent a right lateral orbitotomy. A brown-yellow, encapsulated, multilobular mass was found within the muscle cone beneath the superior rectus muscle and extended medially to lie adjacent to the optic nerve. The tumour had multiple dark varicosities on it, and a large artery branched into the medial surface. Postoperatively the patient recovered

uneventfully and has full ocular function without recurrence (Fig. 4).

Light microscopic examination showed a schwannoma with both Antoni types A and B patterns, Verocay bodies, and calcification. The tumour was encapsulated, and abnormal tumour vessels were present with perivascular lymphocytic infiltrates.

CASE 2

A 54-year-old Caucasian woman 1.5 years prior to admission noticed a subjective right central scotoma which she described as a 'white spot and like looking through a heat wave.' Appreciation of fine detail for the right eye diminished after that, and the visual



Fig. 2 Case 1: CT scan (EM 1010s) showing smooth, well defined, oval intraconal mass lesion with central calcification (arrow).

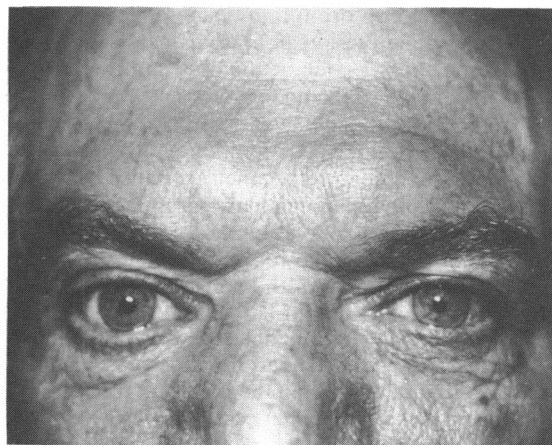


Fig. 4 Case 1: Postoperative photograph.

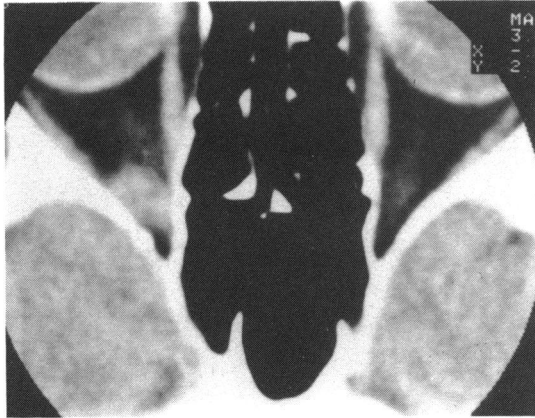


Fig. 5 Case 2: CT scan (GE 8800) demonstrating apical intraconal lesion.



Fig. 6 Case 3: 32-year-old woman with 4 mm downward displacement of right globe and facial asymmetry.

acuity was noted to have fluctuated between finger counting and 6/7·5. She denied ocular pain, photophobia, or changes in her facial appearance.

On examination the best corrected visual acuity in her right eye was 6/7·5. The visual field to confrontation was diminished in the right eye. There was a right relative afferent pupillary defect. Ocular and physical examinations were otherwise unremarkable.

CT scan showed a well-defined intraconal lesion in the posterior aspect of the orbit displacing the optic nerve medially (Fig. 5). The posterior margin was well defined and spared the apex. The lesion was approximately 1×2 cm in diameter, and showed a moderate uniform enhancement. No bone destruction was evident. Carotid arteriography did not demonstrate a blush.

Ultrasonography failed to reveal any abnormality.

The patient underwent a combined frontal craniotomy and lateral orbitotomy panoramic approach. An encapsulated lesion 1 cm in diameter was found within the muscle cone at the apex attached to the superior branch of the oculomotor nerve adjacent to the optic nerve. It had fine varicosities on its surface. Because of extreme space limitations it was incised under the microscope, a specimen examined by frozen section, and the remaining mass carefully evacuated leaving the adjacent nerves intact.

Light microscopy showed predominantly Antoni type A schwannoma with many Verocay bodies. Vessels within the tumour had thick walls.

Postoperatively the patient had a partial right ptosis and paresis of the right superior rectus muscle, which remitted completely within the next 6 months.

The remaining intraconal cases had long-standing histories, and the tumours were well encapsulated, did not extend into the apex of the orbit, enhanced

mildly on CT scan, and failed to take up contrast on angiography. At surgery they were encapsulated, had varicosities on their surfaces, and were extirpated intact with no postoperative problems. In one a distinct origin from the sixth nerve was noted.

The following 3 cases were extraconal in location.

CASE 3

A 32-year-old Caucasian woman complained of a recent onset of diplopia in up gaze. She had noted abnormality of her right upper lid 8 years prior to admission and was aware of downward displacement of her right globe. Other than flushing of the right side of her face she denied any other orofacial symptoms or evidence of masses elsewhere. On examination she had a number of smooth, mobile, soft masses in her right upper lid which were tender to touch. On the right she showed 4 mm downward displacement of the globe, hemifacial asymmetry, and a hypotropia of 4 prism dioptres, without proptosis (Fig. 6). Visual acuity, pupillary function, visual field, intraocular pressure, and fundus examinations were normal bilaterally. A CT scan done on the EML 1010S scanner did not demonstrate a mass.

An anterior orbitotomy was carried out through a brow incision. A multilobular, yellow, solid tumour mass was identified occupying the entire roof of the orbit (Fig. 7). It extended posteriorly to the apex and appeared to come out of the superior orbital fissure. Numerous nerve-like structures were noted extending from the mass. The bulk of this tumour was removed piecemeal.

Histologically it was a typical schwannoma: encapsulated, Antoni type A, with numerous Verocay bodies with thickened blood vessels cuffed by lymphocytes and plasma cells. Electron microscopy

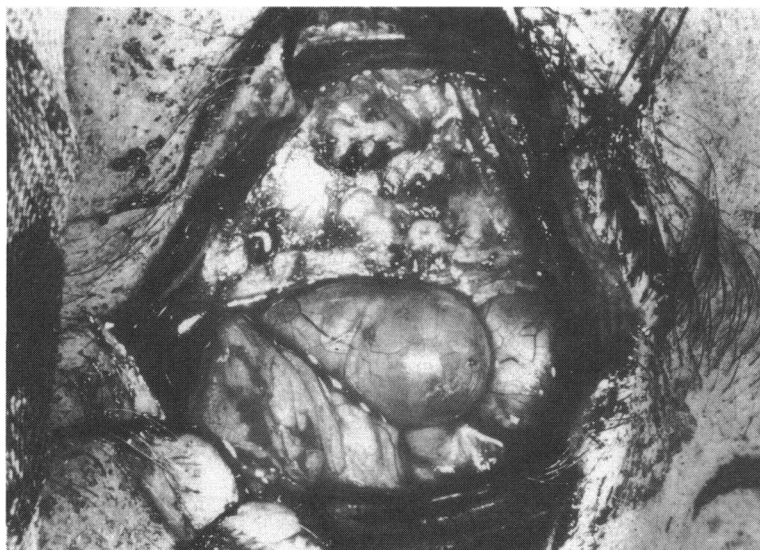


Fig. 7 Case 3: Anterior orbitotomy showing multilobular, solid tumour mass. Note surface vessels.

showed many Schwann cells with stacked pale processes surrounded by a well defined basement membrane.

The patient is asymptomatic, has no evidence of recurrence, and no sensory loss in the supraorbital dermatome.

CASE 4

A 24-year-old man noted sudden onset of a constant dull ache in the region of the inner bony margin of his

left orbit, associated with a sensation of left retrobulbar pressure. He had a history of chronic nasal discharge and had recently noted left ocular mucopurulent discharge and crusting of the lids on waking.

On examination he had a 4 mm left proptosis and lateral displacement of his globe. Examinations of his pupils, visual acuity, intraocular pressure, and fundi were normal bilaterally. He had diminished nasal air movement and hearing changes compatible with past ear disease.

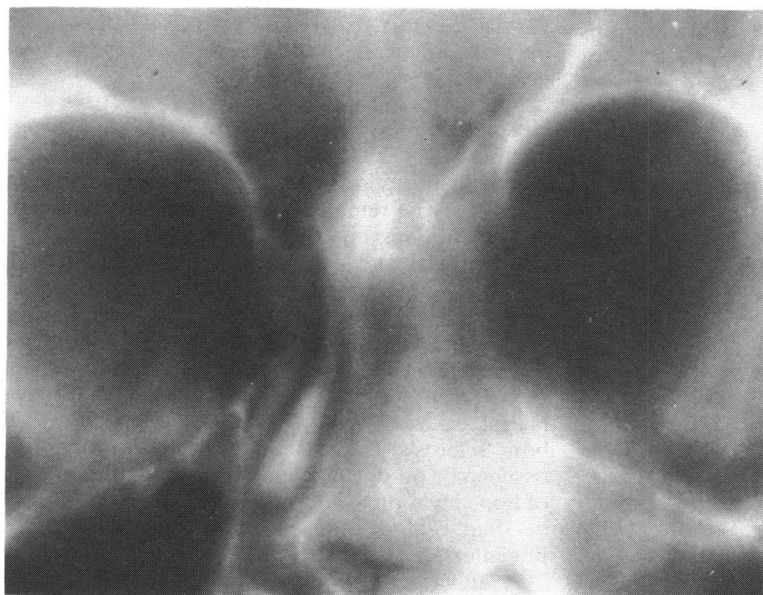


Fig. 8 Case 4: Polytomogram demonstrates opacification of left frontal, ethmoid, and maxillary sinuses and middle turbinate.

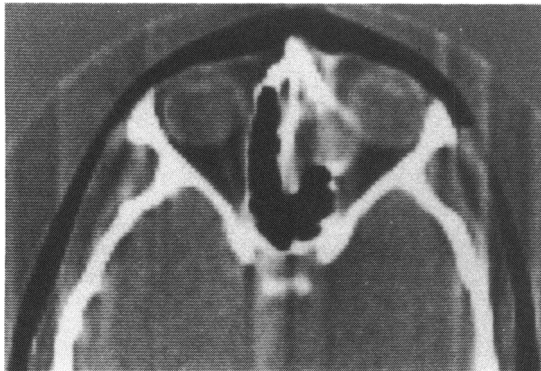


Fig. 9 Case 4: CT scan (EMI 1010s) showing mass extending from left ethmoid sinus into the medial aspect of the orbit.

Physical examination was otherwise unremarkable.

Plain films and polytomography showed opacification of the left frontal, ethmoid, and maxillary sinuses, with bone destruction of the medial wall of the orbit (Fig. 8).

A CT scan confirmed opacification of the left ethmoid sinus and adjacent left middle turbinate. A soft tissue mass bulged into the medial aspect of the left orbit with slight indentation of the globe (Fig. 9). The lesion did not enhance.

The preoperative diagnosis was mucocoele of the left ethmoid sinus with pansinusitis.

The patient underwent an ethmoidectomy through a Lynch incision. At surgery the medial wall of the orbit was bowed laterally and showed bony dehiscence. Beneath the bone and within the sinus there was a yellow-pink tumour with a fibrous capsule. It was easily peeled off and removed. Histo-



Fig. 10 Case 5: 31-year-old man with mass involving left lower lid.

logically it was typical of schwannoma. The tissue was loosely organised in some areas showing prominent nuclear palisading and numerous Verocay bodies. There was considerable nuclear pleomorphism with some large atypical nuclei. Blood vessels were thickened and hyalinised.

CASE 5

A 31-year-old East Indian man first noted in November 1979 a palpable mass involving the left orbital rim and lower lid. It was associated with a local deep, boring, knife-like pain, occurring once a week, lasting a few minutes, and resolving spontaneously. The lesion grew slowly and was biopsied by a general surgeon in January 1980. The histology was equivocal, with a possible diagnosis of fibrous histiocytoma. Four months later on admission to the Vancouver General

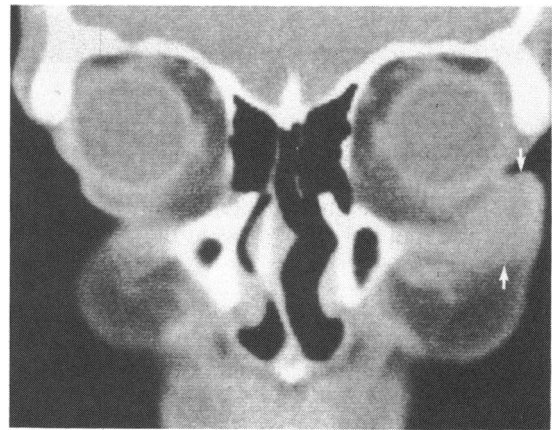


Fig. 11 Case 5: CT scan (GE 8800) demonstrating thickening of soft tissues anterior to the left infraorbital margin.

Hospital showed a firm, smooth, nontender, immobile mass 23×25 mm involving the left lower lid and extending posteriorly over the inferior orbital margin (Fig. 10). The remainder of his ocular and physical examination was otherwise unremarkable.

Both axial and coronal CT scans demonstrated thickening of the soft tissues anterior to the left infraorbital margin (Fig. 11). There was no enhancement of this area, and the lesion was not well defined but appeared to be infiltrative in nature. Tomograms of the left orbit showed a slight depression of the inferolateral aspect of the orbital floor with mild localised bony sclerosis, in keeping with a slow-growing soft-tissue mass producing pressure erosion with evidence of bone invasion.

A and B mode ultrasonography showed numerous fine interfaces within a fairly solid mass lesion.

A left anterior orbitotomy revealed a well encapsu-



Fig. 12A CT scan (GE 8800) demonstrating intraconal, well defined mass lesion.



Fig. 12B Same lesion as Fig. 12A showing mild enhancement following contrast injection.

lated tumour with abnormal varicose vessels overlying the surface. There were adhesions to the orbital septum and the periosteum of the maxillary bone. The mass appeared to extend from the infraorbital foramen laterally and was stripped from the nerve removing it in toto. Postoperative recovery was complete and uneventful.

Light microscopic examination showed a relatively encapsulated tumour mass with numerous small vascular channels and perivascular lymphocytic cuffing. The cells were both spindle and stellate shaped, arranged in a variety of patterns. Some were noted to be loosely entwined within a myxoid-appearing stroma, while others were more compactly arranged in a streaming formation. In some sections a dense, homogeneous, eosinophilic, collagenous stroma was noted. Electron microscopic findings

supported the diagnosis of a fibrous schwannoma.

Results

All 7 patients had histologically confirmed schwannomas. None had systemic manifestations of neurofibromatosis. Their ages at the time of presentation ranged from the third to seventh decades. The tumour sites were 4 intraconal and 3 extraconal. Of the latter, one was in the ethmoid sinus, another anterior to the infraorbital margin, and one involved the superior orbit and upper lid. Displacement of the affected globe was related to the site of the mass and ranged from marked anteroinferolateral to none at all. Visual acuity on presentation was uniformly 6/7.5 or better except for one case having acuity of 6/12 (due to macular degeneration).

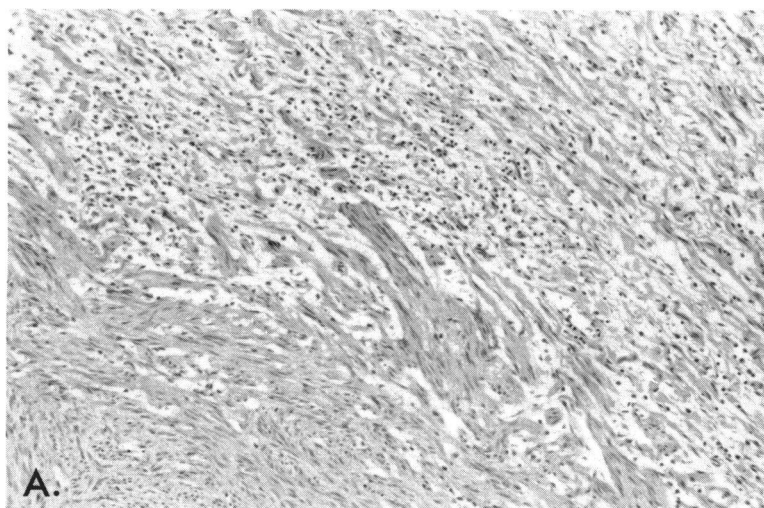
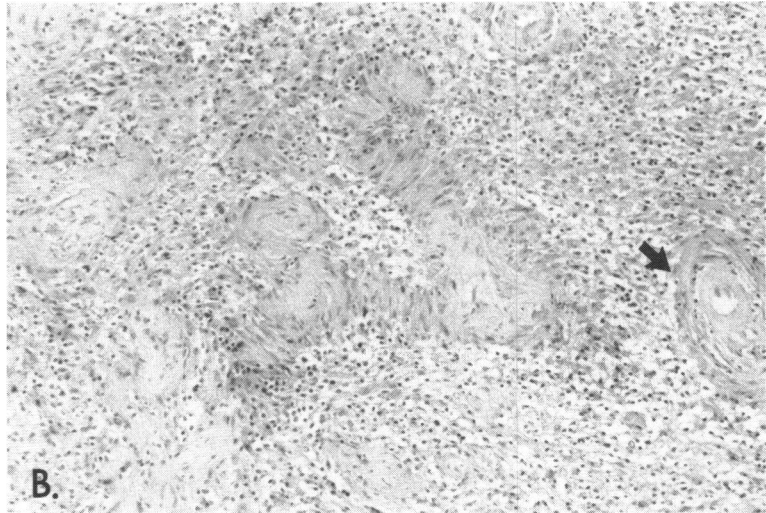


Fig. 13A Histopathology of an intraconal schwannoma demonstrating an admixture of compactly arranged interlacing fascicles (Antoni A pattern—lower right) and more loosely arranged cells separated by a clear matrix (Antoni B pattern) with a background of lymphocytoid cells. (Haematoxylin and eosin, $\times 44$).

Fig. 13B *Histopathology of another area of the same tumour showing a hyalinised vessel (arrow) with an organoid cellular pattern centrally (Verocay body). Background pattern is basically Antoni B with lymphocytoid and lipid laden foam cells. (H. and E. $\times 44$).*



Diplopia and strabismus on a mechanical basis were noted in 2 patients. Only one patient had an abnormal funduscopic examination with choroidal striae and peripapillary haemorrhages.

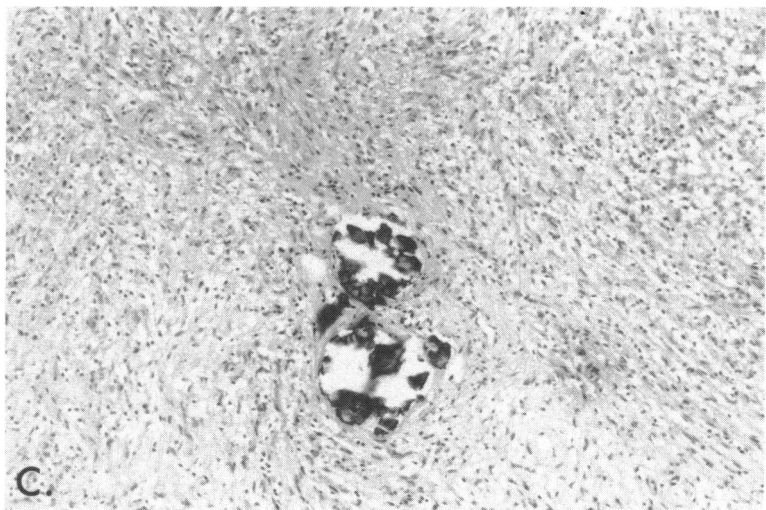
All patients were investigated by CT scanning, which detected 6 of the 7 lesions. Of these, 4 were intraconal, one was in the infraorbital soft tissues, and one was in the ethmoid sinus and middle turbinate with extension into the medial aspect of the orbit. Two tumours caused medial displacement of the optic nerve, 3 caused axial proptosis, and one caused indentation of the globe. Four of 5 contrast CT studies demonstrated enhancement, which was of a mild degree (Figs. 12A, B). In all cases except one the lesions were well defined with discrete edges. The

shape varied from round to oval. One case had associated bony indentation and one had calcification in the central part of the tumour. The intraconal lesions did not follow a muscle or mimic muscle enlargement, nor did they extend into and obscure the orbital apex.

Ultrasonography was carried out in 4 of 7 patients, all of whom had intraconal lesions. Of these, 3 were detected. All were described as encapsulated, solid lesions with a well demarcated anterior border. Within the mass were a number of tissue interfaces which attenuated dramatically. A posterior border could not be detected.

At surgery the lesions were characteristically yellowish-tan in colour, solid, encapsulated, and had

Fig. 13C *Case 1: Light microscopy of tumour from case 1 showing a basically Antoni B pattern with a central area of calcification. (H. and E., $\times 44$).*



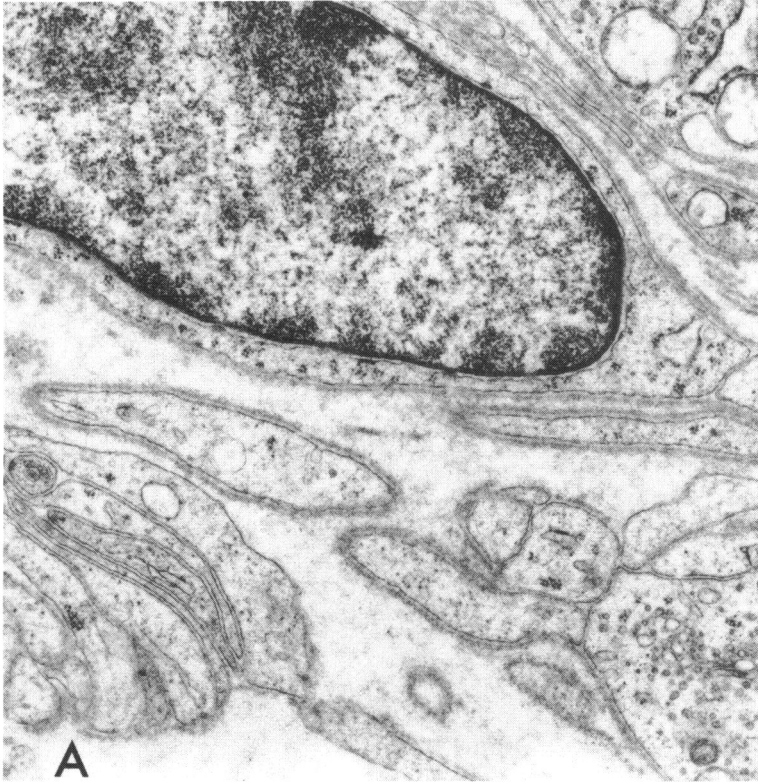


Fig. 14A Case 3: Electron micrograph of tissue from case 3 showing Schwann cells with interdigitating and convoluted cell membranes with few subcellular organelles and a prominent extracellular basement membrane. ($\times 18\,110$).

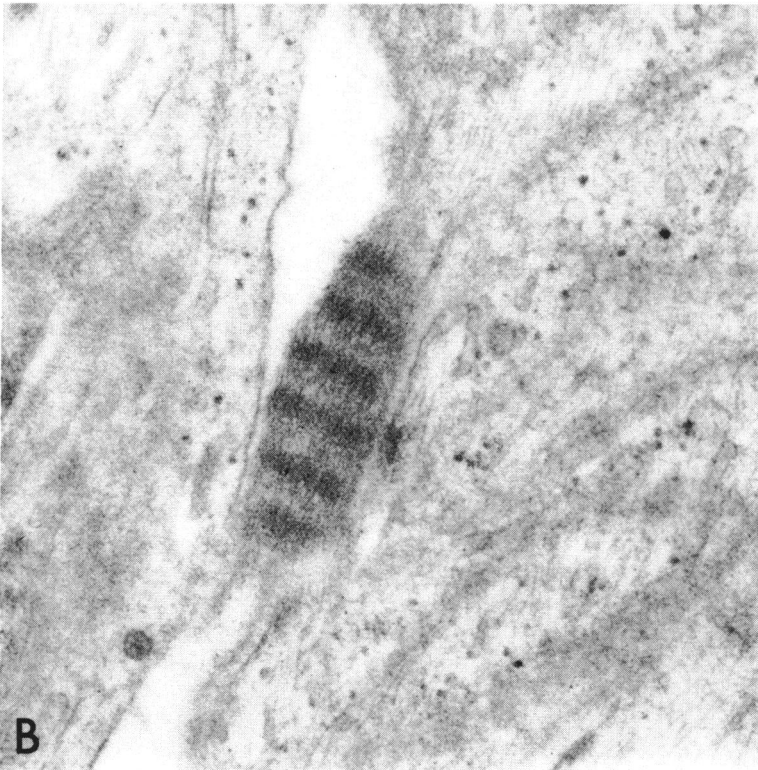
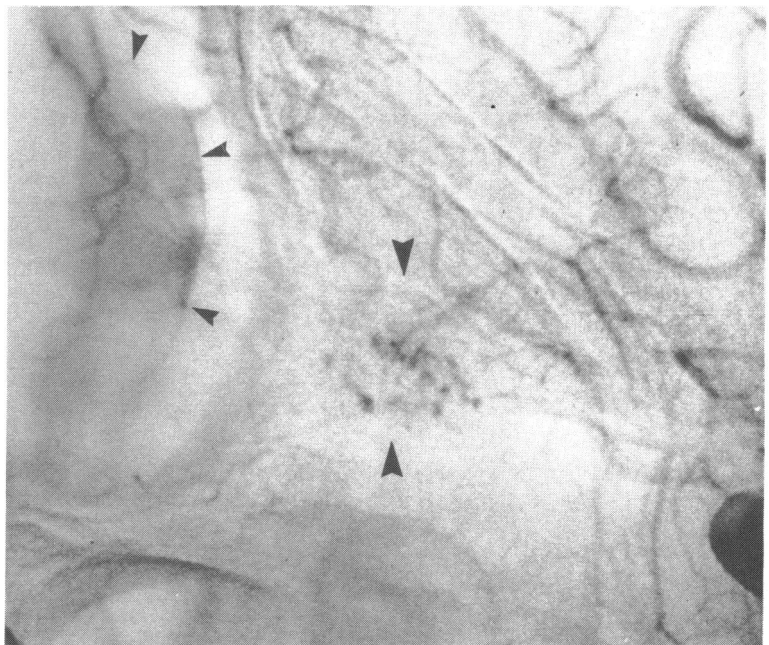


Fig. 14B Electron micrograph from an intracanal schwannoma shown in Fig. 14A. This demonstrates 'long spacing' collagen adjacent to a cell membrane ($\times 50\,700$).

Fig. 14C Case 2: Electron micrograph from the lesion described in case 2. Note adjacent to the nuclear membrane is a longitudinal section of a cilium ($\times 8500$). Insert shows a cilium in cross section ($\times 66000$).



Fig. 15 Carotid arteriogram, subtraction view, of histologically proved cavernous haemangioma showing late minor pooling of dye (between large arrowhead). Small arrowheads define the choroidal crescent.



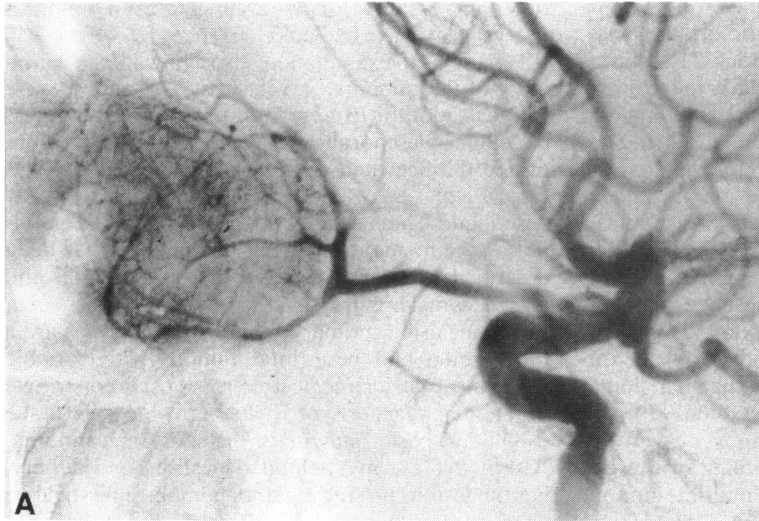


Fig. 16A *Early phase, carotid arteriogram, subtraction view, of histologically proved optic nerve meningioma demonstrating multiple tumour vessels.*

typical varicose, violaceous tumour vessels on their surface. Four nerves were specifically identified in association with their respective lesions, all of which were removed leaving the nerves of origin intact. The associated nerves were: superior division of the third, sixth, infraorbital, and superior division of the ophthalmic nerve.

Histology was variable but typical of schwannoma, including mixed Antoni type A and B patterns (Fig. 13A), tumour capsule, thick walled hyalinised vessels (Fig. 13B), perivascular lymphocytic cuffing, calcification (Fig. 13C), organoid arrangements of Schwann cells with Verocay bodies (Fig. 13B), loose collagenous stroma, and occasional nuclear pleomorphism. Electron microscopy confirmed the

schwannian origin in each of the tumours (Fig. 14A). Long spaced collagen was identified in one of the tumours only (Fig. 14B). An unusual electron microscopic feature noted in one of the tumours was the presence of a typical cilia (Fig. 14C).

Discussion

Primary schwannoma rarely occurs in the orbit. Although no single feature is pathognomonic, a multiplicity of clinical, radiographic, and surgical features point to the diagnosis of this lesion. It is a disease primarily of adults, has an insidious onset, is slow growing, and is noninvasive with minimal effect on other orbital structures. The range of clinical

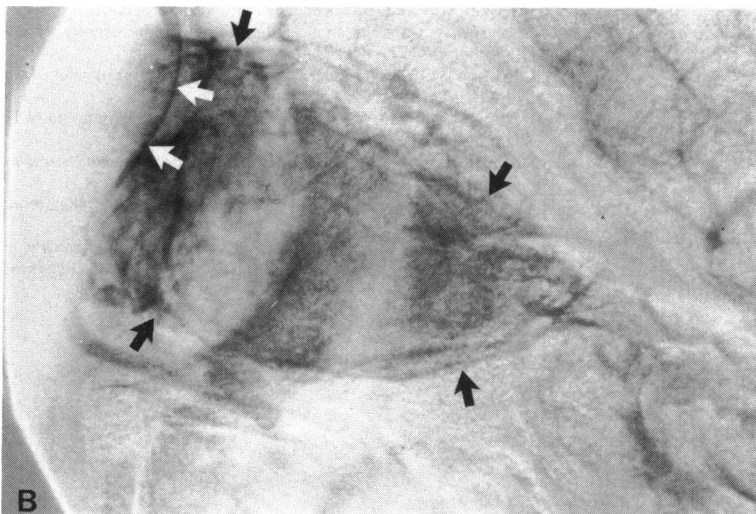


Fig. 16B *Same lesion, late phase, demonstrating tumour blush (black arrows); white arrows indicate choroidal crescent.*

presentation in our patients included optic neuropathy, proptosis, diplopia, anterior orbital mass, and sinusitis. Of available investigations CT scan was found to be the most useful, particularly in localising the lesion, whether within the orbit or extending into it from periorbital soft tissues or paranasal sinuses. The configuration varies from round to oval, and the tumour generally has a well-defined margin and, if intraconal, is well demarcated posteriorly. On CT scan most lesions enhanced to a mild degree, but on carotid arteriography there is no uptake of dye. Ultrasonography confirmed the presence of a discrete, encapsulated, solid-mass lesion. Other slow growing orbital tumours to be considered in the differential diagnosis are cavernous haemangioma, meningioma, and haemangiopericytoma. In our experience, on carotid arteriography, cavernous haemangiomas show late minor pooling of the dye (Fig. 15), while meningiomas have multiple tumour vessels and a late blush (Fig. 16A, B). Haemangiopericytomas have an early florid blush. These 3 lesions when compared with schwannomas showed more marked enhancement with contrast on CT scanning. In addition, on CT scanning schwannomas tend not to follow a muscle or mimic muscle enlargement as is seen in thyroid ophthalmopathy, nor do the lesions extend into and obscure the apex of the orbit, as may be seen in orbital pseudotumour or intrinsic optic nerve lesions.⁴⁻⁷ Although previous series⁸⁻¹¹ all describe lesions of large size, a high index of suspicion, accurate localisation, and determination of size by the methods described here allow small, symptom-causing lesions to be treated early and successfully, as demonstrated in case 2. Primary orbital schwannomas are typically benign, encapsulated, noninvasive tumours. Thus surgical excision as a definitive therapy is indicated.

The surgical approach, whether anterior, lateral, or combined lateral-orbitotomy and frontal-craniotomy, was dictated by the location of the lesions as determined by preoperative investigations. Surgical removal may be total or subtotal, in piecemeal fashion, or by evacuation of the tumour from within its capsule. The type of removal is dictated by the location and attachments to adjacent structures. Because of the peripheral outpouching characteristic of schwannomas they may be successfully stripped of the nerve of origin by microscopic techniques. Like

other authors, in a follow-up ranging from 6 months to 5 years we have had no recurrences.⁸⁻¹⁰

Histological examination also showed the variability of the tumours.^{2,3} Antoni types A and B were found in addition to more fibrous types and some areas of cellular pleomorphism. Electron microscopy confirmed the Schwann cell origin in all cases and was typical of Schwann cell tumours previously described.² The unusual finding of cilia in one case is not a specific feature of schwannomas but has been described in many tumours of both epithelial and mesenchymal origin.¹²

In conclusion, primary orbital schwannomas are a rare orbital and periorbital tumour with variable anatomical and histological expressions. Preoperative differential diagnosis of a slow-growing soft tissue orbital or periorbital mass includes schwannoma. Although CT scan is helpful in localising and identifying the lesion, no single pathognomonic investigative procedure or feature exists. Rather a constellation of features point to this diagnosis. Surgical excision is indicated and to date has been successful.

References

- 1 Schatz H. Benign orbital neurilemmoma. *Arch Ophthalmol* 1971; **86**: 268-73.
- 2 Jones IS, Jakobiec FA. *Diseases of the Orbit*. Hagerstown: Harper and Row, 1979.
- 3 Harken JC, Reid RJ. *Tumours of the Peripherical Nervous System: Atlas of Tumour Pathology*. Second Series. Fascicle. Washington, DC: Armed Forces Institute of Pathology, 1969.
- 4 Nugent RA, Rootman J, Robertson WD, Lapointe J, Harrison PR. CT features and classification of acute orbital pseudotumours. *Am J Neuroradiol* in press.
- 5 Wing SD, Hursaker JN, Anderson RE, Van Dyle HJ, L, Osborn A. Direct sagittal computed tomography in Graves' ophthalmopathy. *J Computer Assisted Tomography* 1979; **3**: 820-4.
- 6 Enzmann D, Donaldson SS, Marshall WH, Kriss JP. Computed tomography in orbital pseudotumour (idiopathic orbital inflammation). *Radiology* 1976; **120**: 579-601.
- 7 Trokel SL, Hilal SK. Recognition and differential diagnosis of enlarged extraocular muscles in computed tomography. *Am J Ophthalmol* 1979; **87**: 503-12.
- 8 Rottino A, Kelly AJ. Specific nerve sheath tumour of orbit. *Arch Ophthalmol* 1941; **26**: 478.
- 9 Alman M, Frayer WC, Hedges TR. Orbital neurilemmoma. *Ann Ophthalmol* 1977; **9**: 1409-13.
- 10 Schmitt E, Spoerri O. Schwannomas of the orbit. *Acta Neurochir (Wien)* 1980; **53**: 79-85.
- 11 Duke-Elders S. *System of Ophthalmology*. London: Kimpton, 1974; **13** (2).
- 12 Font RL, Jakobiec FA. The role of electron microscopy in ophthalmic pathology. In: Trump BF, Jones RT, eds. *Diagnostic Electron Microscopy*. New York: Wiley, 1979; **2**.