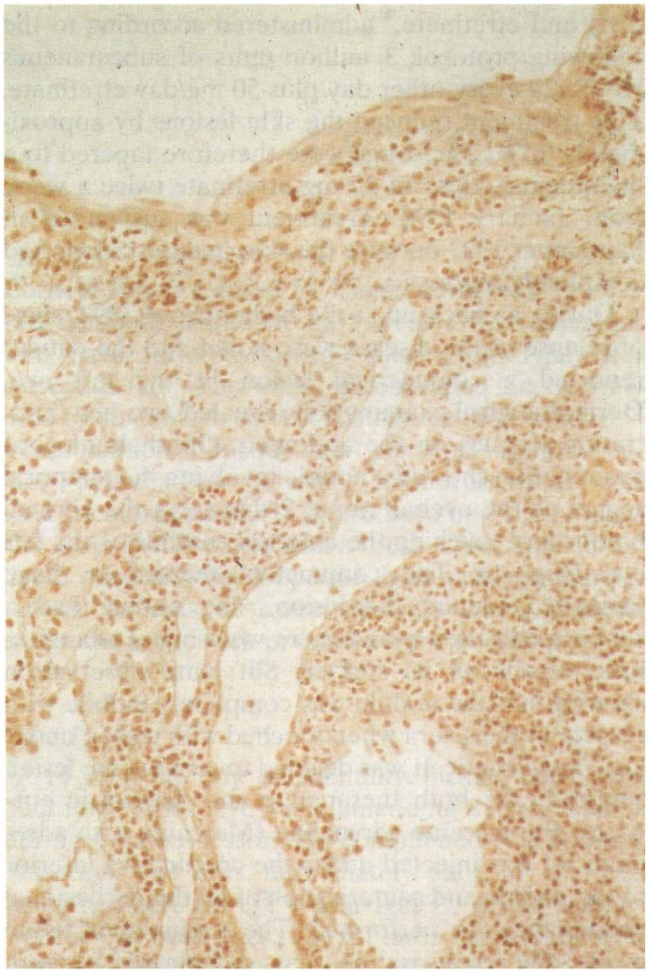

Fig. 2. *Sheets of medium-sized atypical lymphoid cells involving the dermal stroma and Pautrier microabscesses in the epithelium (haematoxylin-eosin, ×15).*

higher magnification, some of these lymphocytes showed large convoluted nuclei. Immunohistochemical study of the tissue showed that the infiltrating lymphocytes were of T-cell origin ($CD3^+$ and $CD4^+$).

These data led to the diagnosis of conjunctival localisation with mycosis fungoides. Therapy with low doses of rINF-α2a (3 million units 3 times a week) and 25 mg etretinate 3 times a week was reinstated and led to a gradual regression of cutaneous plaques. A year later, the patient is still on this therapy and no eye or skin relapses have occurred.

Comment

Eye anomalies are common in patients with mycosis fungoides.[2] The most frequent are seborrhoeic blepharoconjunctivitis, cicatricial ectropion, diffuse punctate epithelial defects, conjunctivitis and meibomianitis. However, it is rare to observe a primitive localisation of mycosis fungoides in the eye.[1,2,5] In a review on the subject,[1] concerned with 36 consecutive mycosis fungoides patients with histologically documented eye involvement, the authors reported that the eyelids were the most commonly affected region and only six cases had a direct conjunctival localisation. Unfortunately the authors did not report how these lesions were treated. In another report of conjunctival involvement with mycosis fungoides, O'Day *et al.*[3] suggested that superficial X-ray therapy can be used to treat these conjunctival localisations of mycosis fungoides, although the authors recognised that side effects such as cataract formation should be anticipated.

We report this case because of its rarity and because of the excellent results obtained with surgery and low doses of rINF-α2a plus etretinate. When possible, this treatment is preferable to X-ray therapy, being just as efficacious and without significant side-effects for the eye.

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Sir,

Hormonal Treatment of Bilateral Optic Nerve Meningioma

Bilateral optic nerve sheath (ONS) meningiomas are rare. To date, of the approximately 500 cases of primary ONS meningiomas reported,[1] only 5% have been bilateral. Neurofibromatosis type II is associated with ONS meningioma and a case of bilateral

ONS meningioma associated with neurofibromatosis was recently reported.[2] In the present case thorough physical examination revealed no evidence of neurofibromatosis. Genetic studies were not performed.

We report a patient with bilateral optic nerve sheath meningiomas whose unresectable tumours were treated by hormonal manipulation.

Case Report

A 51-year-old woman presented to the Royal Victoria Eye and Ear Hospital, Dublin, in October 1992, with a 5-year history of painless bilateral periorbital swelling and gradual deterioration of the vision in her right eye. She complained of redness and discomfort in both eyes, and noticed that the left eye had started to swell and was becoming more prominent than the right.

On examination, visual acuity was hand movements in the right eye and 6/5 in the left. There was no colour perception in the right eye (Ishihara score 0/15), and significantly reduced colour perception in the left (Ishihara score 7/15).

She had a grade 3 right relative afferent pupillary defect, a right divergent strabismus with a right hypertropia, and limitation of eye movements in all directions of gaze in both eyes. Intraocular pressures were 22 mmHg in the right eye and 14 mmHg in the left, increasing on upgaze to 30 mmHg and 25 mmHg respectively, which indicated a restrictive element to the ophthalmoplegia. Fundoscopy showed right optic atrophy and a swollen left optic disc. No opticociliary shunts or choroidal folds were seen.

Radiological Investigations. Plain radiographs of the optic foramina showed no evidence of enlargement. A CT scan revealed non-contiguous bilateral posterior orbital intraconal lesions with no muscle swelling. There was some enhancement with intravenous contrast and the lesions were noted to surround and encase the optic nerves. A small area of high signal intensity was noted in the left intraconal mass, suggestive of calcification. Magnetic resonance imaging (MRI) of the orbits showed bilateral optic nerve swellings with large posterior expansions of the lesions at the orbital apex (Fig. 1). This particular feature has been reported to be a frequent finding in optic nerve sheath meningioma.[3] Railroad tracking, another feature thought to be associated with ONS meningioma, was also demonstrated on the MRI scan. STIR (i.e. MRI with fat saturation) sequences after the administration of gadolinium DTPA were performed. The lesions were noted to extend to the anterior aspect of the optic canals on either side, but did not appear to extend intracranially through the canals. The optic chiasm and sphenoid ridges were normal. The patient underwent right lateral orbitotomy, and a biopsy was taken from the tumour which was isolated from the intraconal space.

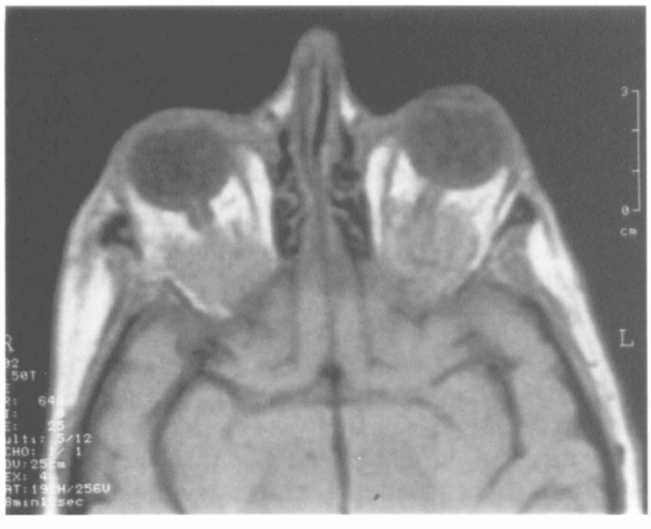

Fig. 1. *MRI scan showing bilateral posterior orbital intraconal tumours with posterior expansion of the lesions at the orbital apex. No connection is seen between the two orbital tumours. A distal perioptic cyst is seen surrounding the optic nerve.*

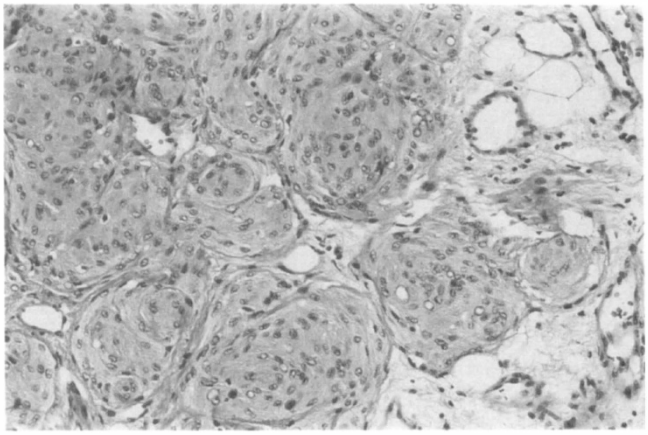

Fig. 2. *Photomicrograph showing typical whorls of bland meningothelial cells infiltrating orbital fat.*

Pathological Findings. Microscopic examination revealed the presence of a typical meningothelial meningioma in which polygonal cells were arranged in whorls and sheets (Fig. 2). The tumour infiltrated orbital fat. Psammoma bodies and mitotic figures were not seen. The tumour (500 mg) was analysed for the presence of hormone receptors by radioimmunoassay. It was found to be negative for oestrogen receptors but progesterone receptors were present at the very high concentration of 850 fmol/g tissue.

Approximately 2 weeks post-operatively the patient complained of backache and was found to have an abdominal mass. Ultrasound revealed a large intrauterine mass (15 cm). Hysterectomy and salpingo-oophorectomy were carried out. A benign intrauterine leiomyoma was removed.

On detailed questioning, the patient now gave a history of visual deterioration prior to menstruation, and improvement with the onset of menstruation, for several years prior to admission. This history, which correlates with the normal premenstrual progesterone rise, together with the progesterone receptor content of the tumour, strongly suggested that she has a progesterone-responsive tumour. Since the meningioma(s) in this case was considered surgically unresectable, and the history of visual deterioration and resolution corresponding with the peak and trough of progesterone levels during the menstrual cycle, together with the high progesterone receptor content of the tumour, suggested a possible hormonal effect, there was a strong possibility that this tumour could be hormonally sensitive and thus susceptible to endocrine manipulation. Meningiomas have long been known to grow more rapidly during the menstrual cycle or during pregnancy.[4] There is also a higher incidence of meningiomas in individuals who have hormonally mediated tumours such as breast cancer, endometrial or ovarian cancer.[5] An association with uterine leiomyomata has not previously been reported.

High titres of non-oestrogen/oestrogen receptor regulated progesterone receptors (PR) are present in over 70% of meningioma cytosols,[6] and meningiomas of meningothelial type are known to have the highest PR values.[7] By contrast, oestrogen receptors are positive in only 31%. Grunberg *et al.*[8] recently reported an objective response of partial tumour regression in 5 of 14 patients who had unresectable intracranial meningioma and were treated with the progesterone antagonist Mifepristone. We have used the synthetic antiprogesterone hormone gestrinone (ethylnorgestrienone or $C_{21}H_{24}O_2$) to treat this patient. The patient has been taking 2.5 mg of this drug twice weekly for 3 years with no side effects, and she has no further redness or discomfort in either eye. Repeat MRI scan shows no change in tumour size after 3 years (however, these tumours grow extremely slowly), and field analysis shows no significant deterioration. An antiprogesterone agent is still necessary in this patient despite bilateral salpingo-oophorectomy, as progesterone is produced in the subcutaneous tissues and adrenal glands. She will remain on gestrinone, and will be reviewed at 6 monthly intervals. Surgery will only be performed should the tumour extend intracranially, as at the present time tumour resection would render her totally blind. This case demonstrates stabilisation of bilateral optic nerve meningioma with hormonal manipulation.

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Sir,

Cytomegalovirus Retinitis in the Presence of a Normal Helper T-cell Count

Cytomegalovirus (CMV) infection is seen in severely immunodeficient patients with carcinomatosis, following organ transplant, or on cytotoxic or immunosuppressant therapy. CMV retinitis now occurs most often in patients with the Acquired Immune Deficiency Syndrome (AIDS), usually in the presence of a helper T-cell ($CD4^+$) count of less than 0.05×10^9/l.