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Sir,

Accelerated growth of a primary orbital schwannoma during pregnancy

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Primary orbital schwannomas are rare, accounting for about 1% of orbital tumours.[1,2] There is a known association with neurofibromatosis.[2–4] Typically, they arise from the first division of the trigeminal nerve (supraorbital and supratrochlear branches).[1,2,5] A typical presentation may occur as a single cystic lesion in the orbit[6] or as a schwannoma arising from the optic nerve itself.[7] We present a case of a primary orbital schwannoma with accelerated growth during pregnancy, which was positive for progesterone receptors. To the best of our knowledge, this is the first time hormone receptors have been found in such tumours.

Case report

A 38-year-old Caucasian lady presented initially with a 1- to 2-year history of a slowly progressive right-sided proptosis, which worsened significantly during her pregnancy. On examination, she had 11 mm of axial proptosis with normal optic nerve function. She had no trigeminal or other cranial nerve involvement. Systemic examination was normal and there was no family history of neurofibromatosis. MRI scan showed an extensive, moderately enhancing lesion, which masked the optic nerve (Figure 1a and b).

An orbital biopsy was subsequently carried out, histology of which suggested a diagnosis of schwannoma/neuroleimoma. Surgical treatment with the associated risks were discussed with the patient.

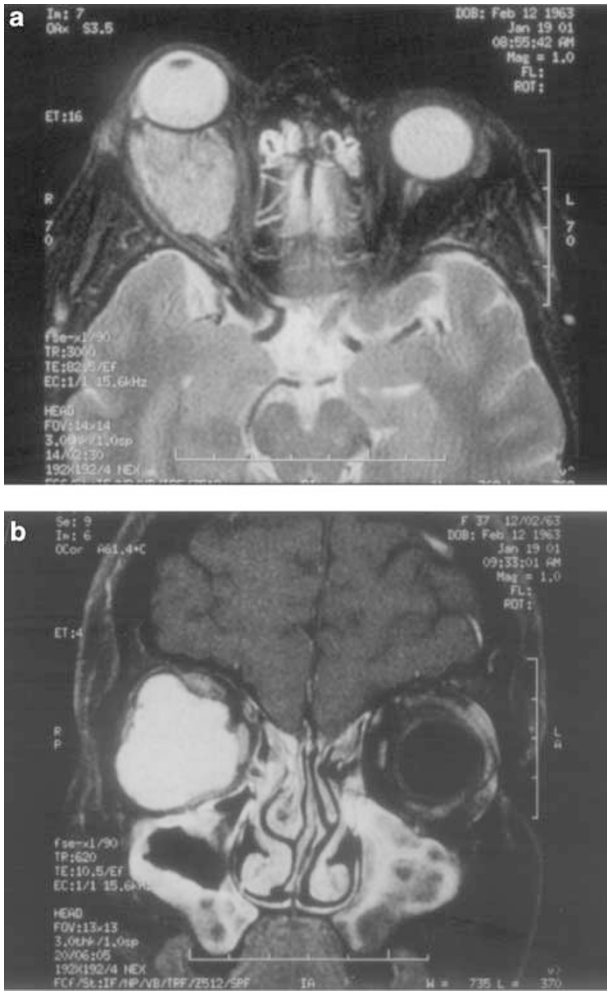

Figure 1 (a and b) Axial and coronal MRI (T2-weighted) scans showing a large enhancing mass in the right orbit. The optic nerve is masked by the lesion.

A lateral orbitotomy was performed. The tumour was found to be poorly encapsulated and friable, making complete excision difficult. Further histological examination confirmed the initial diagnosis of a primary orbital schwannoma. Histology showed abundant spindle cells in a myxoid, collagenous background (Figure 2a), which was positive for S-100 and vimentin. The history of accelerated tumour growth during pregnancy, prompted us to perform immunohistochemical examination for hormone receptors. The tumour was found to be diffusely positive for progesterone (using Dako antibody, clone PGR 636), but not oestrogen receptors (clone 1D5), see Figure 2b.

Her postoperative course was uneventful, with significant resolution of her proptosis and preservation of her optic nerve function. Currently, she has a moderate abduction deficit, which continues to improve. We continue to monitor this patient, as complete surgical excision was not histologically confirmed.

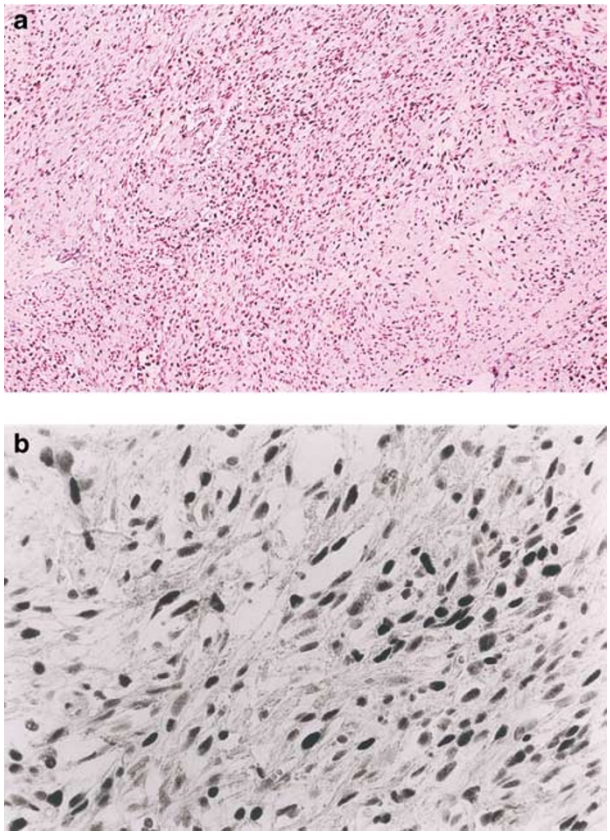

Figure 2 (a and b) Haematoxylin and eosin stain showing spindle cells in a myxoid collagenous background (top). Lesion stained with Dako antibody (clone PGR 636) shows the presence of progesterone receptors (bottom, at $\times 40$ magnification).

Comment

Schwannomas are more common in females than males.[1,2] Proptosis, visual dysfunction, diplopia, and paraesthesia/pain in the trigeminal divisions summarise the most common presenting features of this condition. Neuroimaging typically shows a well-circumscribed, lobulated, round to oval lesion in the superior orbit, which enhances mild to moderately with contrast.[2,8] Cyst formation within the tumour is characteristic.[1,2,9,10]

The differential diagnosis includes other slow-growing tumours including cavernous haemangioma, meningioma, and haemangiopericytoma,[1,2,5] all of which show greater contrast enhancement radiologically than schwannomas[5,8] in general.

Cockerham *et al*[11] described in their series one patient presenting 2 months postpartum. Our patient clearly described a significant increase in proptosis during her pregnancy, suggesting a possible hormonal influence in the growth of a subgroup of schwannomas. The confirmed presence of progesterone receptors in this case helps support this hypothesis. Progesterone levels are known to increase from 2 weeks gestation onwards until term, which may explain the accelerated growth during pregnancy.[12] This may also explain the increased prevalence in females, who have higher levels of progesterone, particularly in the luteal phase of the menstrual cycle (18.1–89.4 nmol/l) compared with males (0–1.2 nmol/l).[13] Hormonal manipulation may therefore be a future option in the treatment of schwannomas.

Histological features can vary, with the Antoni A and B type cells classically but not universally identified. A true capsule of perineural tissue surrounds schwannomas and if intact confirms complete excision.[1,2] Immunohistochemical examination, for example, with S-100 and vimentin gives additional supportive data when classical histology is not present.[1,2,11]

Surgical excision is the treatment of choice in symptomatic patients. Surgical friability can complicate tumour excision and, although not common, has been described.[1,5,11] In this case, the previous biopsy, which disrupted the capsule, may have resulted in operative friability of the tumour. Local recurrences may occur many years later, but malignant transformation is extremely rare.[1,2,5]

Conclusion

The presence of progesterone receptors may explain the higher incidence of primary orbital schwannomas in females and the influence of pregnancy on its growth.

This also opens the possibility of hormonal manipulation in treating such tumours.

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Sir,

Scedosporium apiospermum keratomycosis with secondary endophthalmitis

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Scedosporium apiospermum, the asexual form of *Pseudallescheria boydii*, is a saprophytic filamentous fungus found in soil, manure, and polluted water. In the tropics, it is a common cause of mycetoma and systemic infections have been reported in immunocompromised patients.[1] Ocular infection is frequently associated with trauma and generally the prognosis is poor. We report the treatment of a case of keratomycosis caused by *S. apiospermum* that progressed to endophthalmitis. Although infection was controlled, vision was ultimately lost as a result of secondary glaucoma.

Case report

A 56-year-old female contact lens wearer became aware of a foreign body sensation in her right eye while walking in a wooded area of southern England. A corneal abrasion was treated in A&E with topical chloramphenicol 0.5% drops. After 1 week, the patient presented with a central corneal abscess and hypopyon. A sample of corneal tissue was taken for microscopy and culture. Treatment was modified to topical ofloxacin 0.3% hourly. There was a rapid clinical improvement and the patient was discharged on topical ofloxacin and prednisolone (0.3% q.i.d.). On review, 1 week later, there was an increased corneal infiltrate and the hypopyon had recurred. *S. apiospermum* was cultured from the initial corneal scrape material and, therefore, the topical steroid was withdrawn and treatment changed to topical clotrimazole (1%, hourly), oral fluconazole (200 mg/daily) and intravenous liposomal amphotericin B (1 mg/kg/day). This was later modified to hourly topical econazole (1%) and amphotericin B (0.15%) with oral itraconazole (200 mg/daily) when the results of *in vitro* susceptibility testing became available. No donor corneal material was available and despite continued intensive topical therapy, the cornea perforated after 4 days (Figure 1a).