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Sir,

**Sinonasal melanoma: an unusual cause of proptosis**

Malignant melanomas of the nasal cavity and paranasal sinuses are rare, accounting for less than 1% of all melanomas in the Western population. We report a case of sinonasal melanoma presenting with proptosis and a visible medial canthal mass.

*Case report*

An 85-year-old man presented to our department with a 3-month history of a pigmented lesion and mild swelling in his left medial canthus. His first symptom had been epiphora 9 months prior to presentation with some swelling of his medial canthus 3 months later associated with some scant epistaxis. Six weeks prior to presentation the epiphora had worsened and he had noticed some proptosis and diplopia.

The patient had worked as a French polisher for many years and was on 5 mg prednisolone daily for polymyalgia rheumatica. Although frail, he had no other specific medical problems.

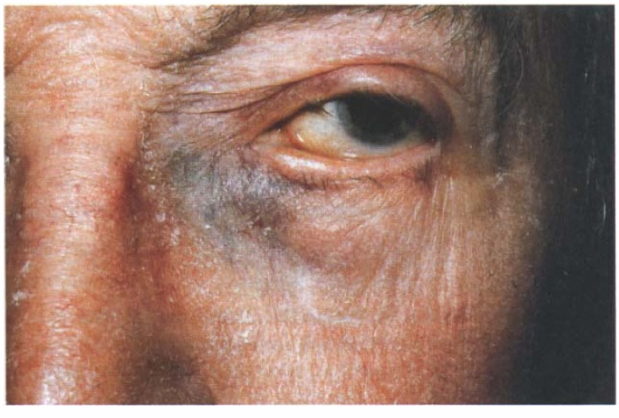
Examination revealed an obvious pigmented mass in the left medial canthus measuring approximately 15 mm × 15 mm. The mass was firm and non-tender, with no associated lymphadenopathy. There was 4 mm of non-axial proptosis with 6 mm of lateral globe displacement and markedly restricted extraocular movements (Fig. 1a). Fundoscopy revealed a raised lesion on the left inferonasal retina, but B-scan ultrasound indicated indentation of the left globe rather than a choroidal mass. MRI scan showed an extensive mass in the left ethmoid sinus eroding into the orbit and indenting the globe. No intraocular mass was evident, nor was there evidence of midline spread (Fig. 1b).

The patient was referred to an ENT surgeon who performed a transnasal biopsy, as the mass was accessible via this route. Histological examination confirmed a malignant melanoma of mixed cell type with positive staining for melanin and positive immunochemistry for S100 and HMB-45 (Fig. 2). Due to the extent of the tumour and the patient's frailty, it was decided that exenteration and block clearance was not indicated. He was therefore referred for palliative radiotherapy.

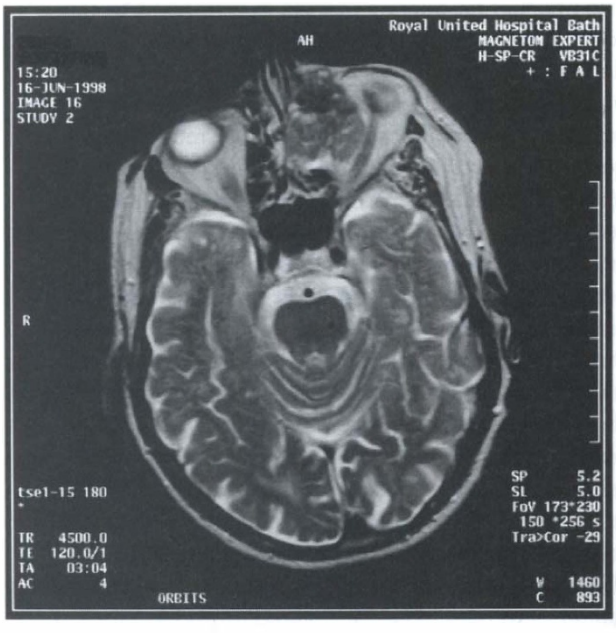
*Discussion*

Primary malignant melanoma arising from the sinuses and nasal mucosa is rare and carries a poor prognosis. It accounts for less than 1% of all melanomas in Western populations with an apparent higher incidence of 7–11% in Japanese populations.[1] It tends to occur more commonly in elderly people with a peak incidence in the sixth to eighth decades with no difference in sex prevalence.[2] Causal relations are not well established.

Presentation is often varied and occurs late in the natural history of the disease, contributing to a poor prognosis. The most common site of occurrence is the nasal cavity and patients may present with unilateral nasal obstruction and epistaxis.[3] Swelling of the nose and pain are less common presenting symptoms. Various studies have indicated a prevalence of 78% in the nasal cavity and only 5% in the ethmoid sinuses.[4,5]



(a)




(b)

**Fig. 1.** *(a) Slate-grey medial canthal mass causing proptosis and lateral displacement of the globe. (b) MRI scan showing a left ethmoid mass eroding into the orbit and indenting the left globe.*

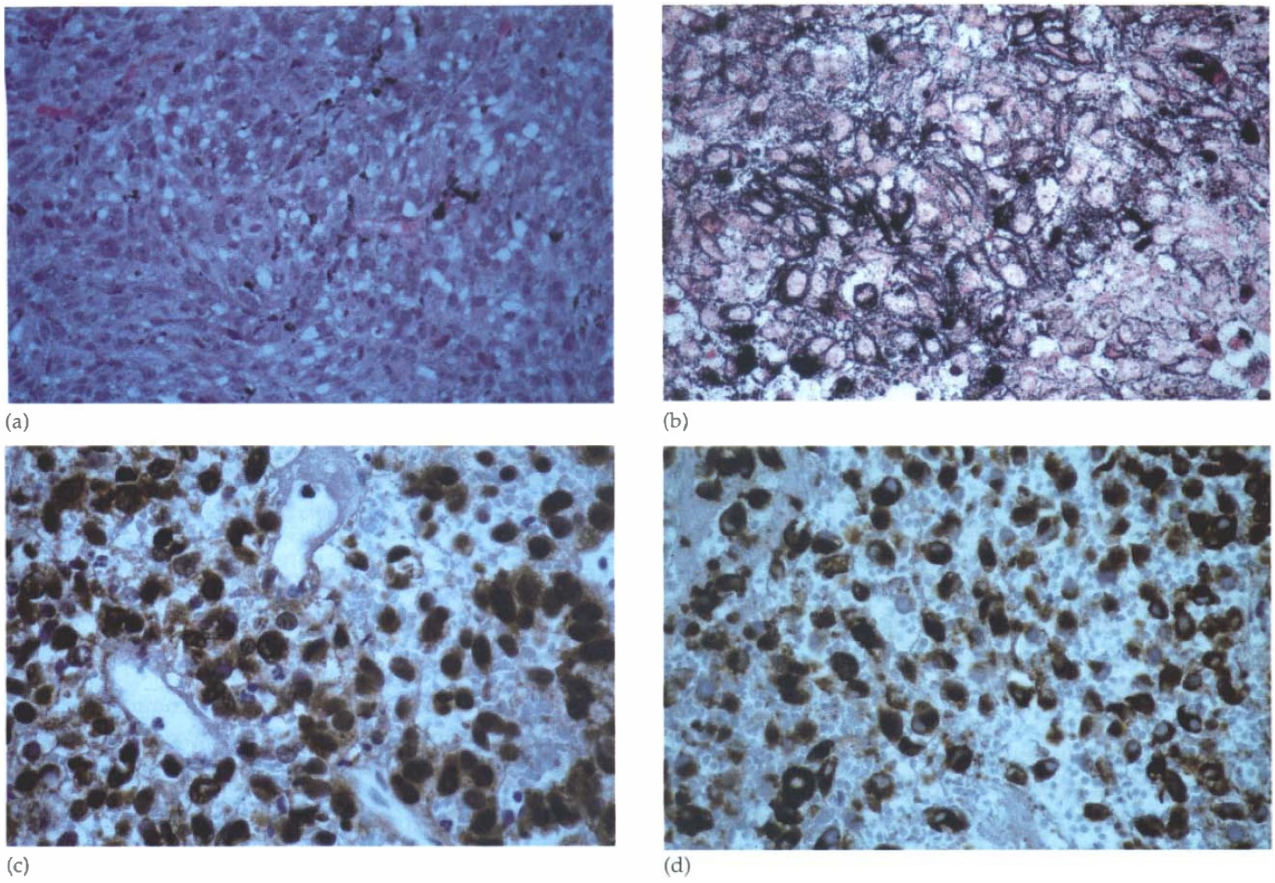



**Fig. 2.** *(a) Photomicrograph of H&E stain. Scale 1:200. (b) Photomicrograph of Masson-Fontana stain illustrating melanin staining black. Scale 1:200. (c) S100 and (d) HMB-45 immunoreactivity. Scale 1:200.*

Diagnosis can be difficult, especially in amelanotic lesions, which show no evidence of melanin on staining. Immunochemistry confirms the diagnosis, as melanomas characteristically are immunoreactive for S100 protein and HMB-45 (melanin-specific antigens). They are immunonegative for cytokeratin, lymphoid and neuroendocrine markers.

Approaches to treatment vary, but surgical resection with radiotherapy seems to provide the best prognosis.[4] Radiotherapy alone may be effective in controlling local tumour spread and limiting size but is not effective in completely eradicating the tumour. It is therefore more often used in conjunction with surgical resection and in this context may improve survival.[4] The value of adjunctive chemotherapy and immunotherapy has not been well proven.[1]

Survival rates of patients with mucosal malignant melanoma arising in the nose and paranasal sinuses are poor with a mean survival of between 2½ and 3½ years. Less than 1% of patients survive for 10 years.[6,7] Prognosis seems unrelated to the size, site or pigmentation of the tumour and death is usually from local invasion or systemic metastases.[7]

In summary, although sinonasal melanoma is rare, it should nonetheless be considered in the differential diagnosis of both a medial canthal mass and non-axial proptosis.

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Sir,

**Recurrent Optic Disc Edema with a Macular Star (ODEMS)**

Neuroretinitis is a term which encompasses a heterogeneous group of disorders characterised by reduced visual acuity, optic disc oedema and peripapillary exudates which often form a macular star.[1] The majority of cases are of unknown aetiology and typically there is spontaneous improvement in visual