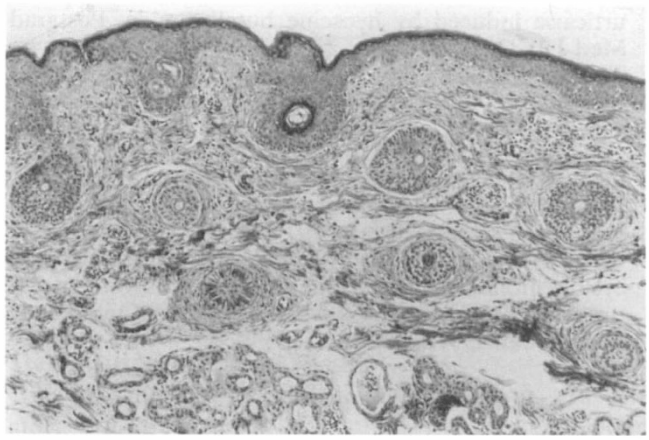

Fig. 2. *Numerous well-differentiated hair follicles were found in the dermis in a tangential section (haematoxylin and eosin, ×60).*

serial sections revealed no central dilated infundibulum, the histological diagnosis was trichofolliculoma because of apparent increased density in hair follicles. Follow-up examination 6 months later revealed no sign of recurrence.

Comment

Follicular neoplasms, which comprise the largest class of cutaneous adnexal tumours, exhibit a wide spectrum of differentiation. The most organised naevoid tumour related to the entire hair follicle has been appropriately called trichofolliculoma; these tumours are classified as highly structured hamartomas. The lesion was considered by Kligman and Pinkus[4] to be of intermediate differentiation between a hair follicle naevus, a congenital simple hyperplasia of the hair follicle, and trichoepithelioma, which usually lacks mature hair follicles. Histologically, the dermis contains a large cystic space that is lined by squamous epithelium and contains horny material and frequently also fragments of birefringent hair shafts.[5] Radiating from the wall of these primary hair follicles, one sees many small but usually fairly well differentiated secondary hair follicles. These fine hairs are visualised best where the secondary hair follicles appear in cross-section, as shown in our case.

Clinically, trichofolliculoma occurs in adults as a solitary, skin-coloured, dome-shaped papule or nodule, sometimes with a central pit.[5] If such a central pit is present, a wool-like tuft of immature, usually white hairs may be seen emerging from it – a highly diagnostic clinical feature.[5] The lesion occurs most commonly on the face, scalp and neck; however, very seldom is it found on the eyelid. Although one case of perineural invasion has been reported,[6] the tumour is considered benign and complete excision is curative. There have been no reported cases of malignant transformation. Simpson *et al.*[1] reviewed benign eyelid tumours derived from hair follicles, such as trichoepithelioma, trichofolliculoma, trichilemmoma and pilomatrixoma, and have stressed the importance of making a tissue diagnosis, because these tumours are rare but frequently misdiagnosed as basal cell epithelioma, which is the most common malignant neoplasm on the eyelid (Table I). Dermatologists and ophthalmologists should be aware of this neoplasm because it can occur on the skin around the eyelid.

Table I. Incidence of benign tumours and basal cell epithelioma on the eyelid[1]

Basal cell epithelioma	95.4%
Pilomatrixoma	3.6%
Trichoepithelioma	0.7%
Trichilemmoma	0.2%
Trichofolliculoma	<0.1%

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Sir,

A Rare Case of McCune–Albright Syndrome Associated with Glaucoma Retinal Degeneration and Arteriovenous Malformations

McCune–Albright syndrome (MAS) is characterised clinically by unilateral localised bone lesions termed fibrous dysplasia, cutaneous pigmented areas and endocrine dysfunction associated with precocious puberty in females.[1] Craniofacial bones are affected in 50–70% of all cases, and there have been reports of proptosis, diplopia, epiphora and even visual loss caused by optic nerve compression.[2–4] The MAS patient described here was found to have dysgenic iridocorneal angles with increased intraocular pressure, retinal degeneration, and an arteriovenous

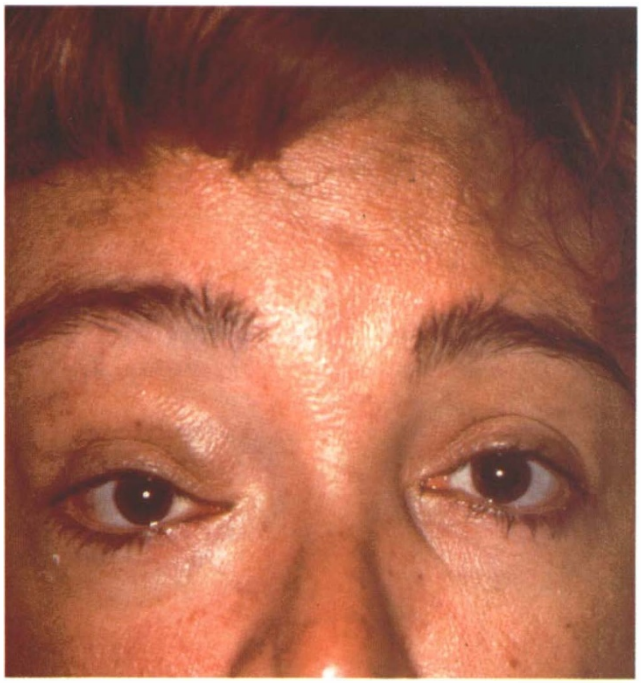


Fig. 1. *The right eye is displaced inferolaterally. Note the angioma located at the 2/3 position on the right upper lid.*

malformation involving the palpebra and orbit; none of these has previously been reported in association with this disease.

Case Report

A 36-year-old woman was referred for ophthalmic evaluation. Café-au-lait macules were present on the trunk at birth. When she was 3 years old a femoral bone biopsy had revealed fibrocystic lesions. MAS was diagnosed at the age of 4 years when menarche occurred and secondary sex characteristics appeared.

The physical examination revealed facial asymmetry and an angioma involving the right upper eyelid (Fig. 1). Best corrected visual acuity in both eyes was 20/20. Intraocular pressures (IOP) were 27 mmHg in the right eye (RE) and 21 mmHg in the left eye (LE). Visual field examination showed bilateral enlargement of the blind spot. The iridocorneal angles in both eyes were open in all four quadrants. The trabecular meshwork appeared dysgenic with angular microbridges extending from the root of the iris, which was inserted more posteriorly than normal, towards the sclerocorneal trabeculae and Schwalbe's line (Fig. 2). The optic discs were asymmetrically cupped with cup/disc ratios of 0.5 (RE) and 0.1 (LE). Areas of lattice degeneration and perivenous pigment deposits were noted in the extreme peripheries of both fundi.

Computed tomography revealed mild right exophthalmos caused by an osteodystrophic neoplasm involving the sphenoid and frontal bones and the right ethmoidal cells; this was even more evident on magnetic resonance imaging. The optic nerve and intra-orbital muscles were unaffected. Magnetic resonance angiography revealed that the tumour was supplied by an abnormal vascular structure arising from the external carotid artery and apparently linked to the palpebral angioma (Fig. 3).

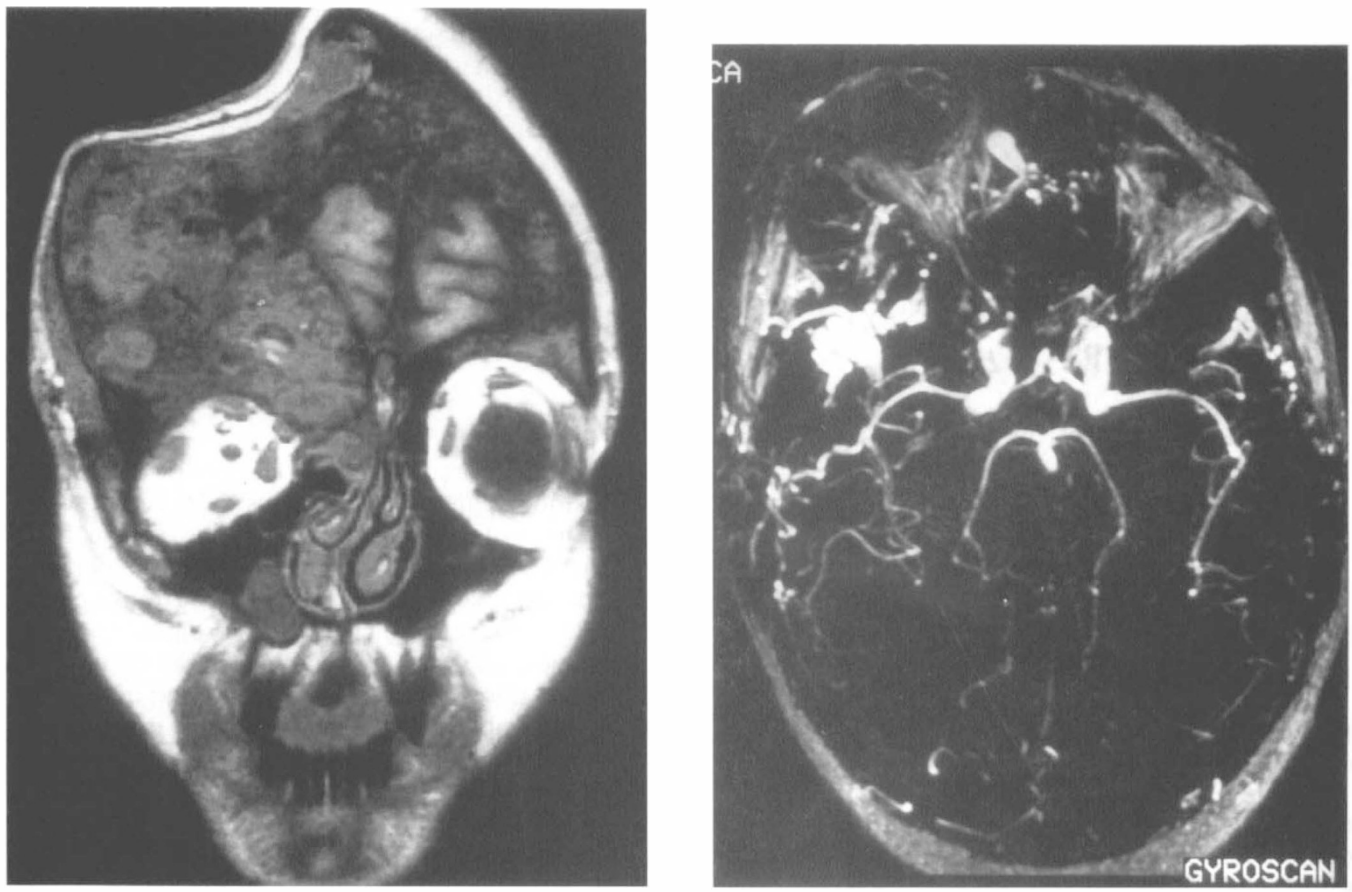
The retinal lesions were treated with laser photocoagulation. Excision of the orbital bone lesions was deferred since the tumour was not compressing the optic nerve; the glaucoma is being controlled with timolol maleate 0.5%. However, surgery may be necessary if the compression becomes more severe.

Discussion

MAS is a sporadic disease classically defined by polyostotic or monostotic fibrous dysplasia, café-au-lait skin spots ipsilateral to the side of the bone lesions, sexual precocity and autonomous hyperfunction of multiple endocrine systems.[1] The dysplastic bone lesions are characterised by progressive replacement of the normal medullary architecture with abnormal fibro-osseous tissue, composed of spindle-shaped fibroblastic cells and minimal extracellular matrix, surrounding trabeculae of immature metaplastic woven bone.[5]

Various non-endocrine developmental abnormalities have been described in association with MAS,

Fig. 2. *Right eye. Gonioscopic view demonstrating angle dysgenesis.*



(a) (b)

Fig. 3. *(a) T1-weighted spin echo magnetic resonance (MR) image acquired in the coronal plane. Neoplastic tissue is seen involving the frontal bone and expanding medially and downwards, invading the frontal sinus, the ipsilateral ethmoidal region and displacing the nasal septum contralaterally. Signs of previous surgery can be noted on the right parietal bone. (b) MR angiography (2D ToF; Maximum Intensity Projection reconstruction in the axial plane) demonstrating the high vascularity of the osteo-fibrous tissue arising from the external carotid artery.*

including congenital biliary atresia, gastrointestinal polyps, hyperplasia of the thymus, spleen and pancreatic islet cells, microcephaly and failure to thrive,[6] but this is the first report of iridocorneal angle dysgenesis in a patient with this disease.

Recent investigations suggest an aetiological role for embryonic somatic cell mutation that leads to the substitution of His or Cys for Arg at amino acid 201 of the Gs alpha-subunit, which stimulates cAMP formation.[6–8] This alteration has been observed in dysgenic endocrine and non-endocrine tissues from patients with MAS, but not in normal-appearing tissues,[6–8] which is consistent with the mosiac distribution of abnormal cells.[9]

It is possible that the abnormal embryological development observed in the iridocorneal angles of our patient may be related to the presence of mutant cells; in that case their bilaterality would confirm that the dysgenesis in this syndrome occurs at an early stage of embryogenesis. The increases in intraocular pressure were probably caused by this malformation, although compression of the bulb and/or increased venous resistance within the right orbit caused by the bone tumour may have contributed to the more significant increase in the RE. This patient also presented a complex vascular malformation and retinal degeneration that have never been reported in patients with MAS. The aetiology of our patient's retinal lesions is unclear, and they may, in fact, be unrelated to the MAS.

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Sir,

Bilateral Endogenous *Candida* Endophthalmitis and Chorioretinitis following Toxic Megacolon

Endogenous *Candida* endophthalmitis has been reported with increasing frequency in hospitalised patients with recognised risk factors. Confirmation of the diagnosis is often difficult and delayed. Treatment must proceed on the basis of the clinical appearance, behaviour and history. Intravitreal antifungals should be given at the time of vitreous biopsy. Vitrectomy is not always necessary but may considerably accelerate recovery and should be considered early. High-dose oral fluconazole, if tolerated, brings about a slow but steady resolution. Final prognosis is good if no significant chorioretinal scarring occurs at the macula and the patient recovers from any concurrent systemic illness.

Case Report

A 61-year-old woman developed *Salmonella* gastroenteritis with persistent diarrhoea while on holiday in Mallorca. She was admitted to the medical ward but within 2 weeks required a total colectomy and ileostomy for toxic megacolon. She also underwent bilateral radical oophorectomy for ovarian tumours, later confirmed histologically as moderately well differentiated serous cystadenocarcinoma. She received vancomycin, clarithromycin, metronidazole and ciprofloxacin intravenously or orally over a period of 4 weeks.

Visual symptoms were of a steady deterioration of left vision over 1 week with accompanying floaters. Corrected visual acuity on presentation was 6/12 in the right eye and counting fingers in the left. There was mild anterior segment activity in the right eye and moderate activity in the left with flare, 3+ cells and posterior synechiae. Vitritis was marked (3+) in the left eye and a focal fluffy area of active chorioretinitis was noted at the left macula (Fig. 1).

The possibility of fungal aetiology was considered at presentation because of the history and appearance. She was admitted and investigations showed negative serology for *Toxoplasma*, *Toxocara* and syphilis, as well as negative blood cultures. However, the wound swab from her ileostomy site was positive for *Candida albicans.* She underwent left vitreous biopsy and intravitreal injection of antibiotics and antifungal (amikacin 0.4 mg and vancomycin 1 mg, both in 0.1 ml of normal saline and amphotericin B 5 μg in 0.1 ml of 5% dextrose solution) to cover the likely causes of endogenous endophthalmitis including the suspected fungal cause. She was also started on oral fluconazole (100 mg b.d.), topical steroids and a mydriatic. Prompt microscopy was unable to detect any fungal mycelia and cytology failed to detect any fungal elements. Over the next 5 days

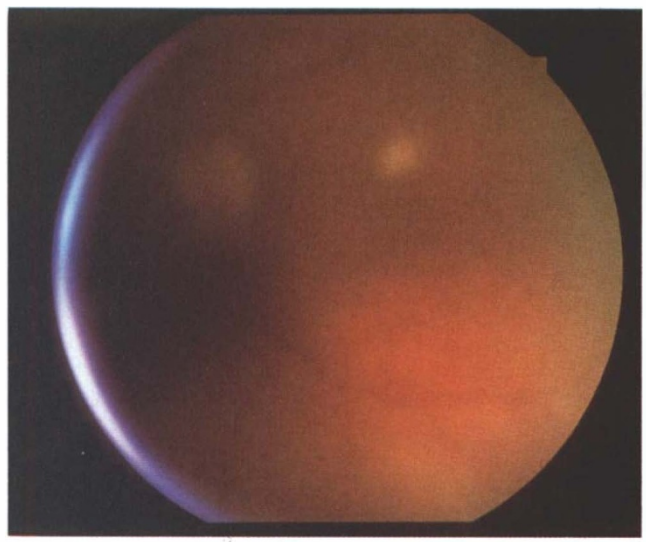

Fig. 1. *Left fundus at presentation. A dense focus of active chorioretinitis is visible in the central macula through considerable overlying vitritis.*