LETTERS TO THE EDITOR

Microcystic adnexal carcinoma masquerading as a chalazion

EDITOR,—Microcystic adnexal carcinoma (MAC) is a locally aggressive cutaneous tumour first described by Goldstein et al in 1982.1 This tumour has a predilection for the face2 with only a few cases reported involving the periocular region.3,4 It is a low grade tumour of eccrine sweat gland derivation which behaves aggressively in its growth pattern and recurrence rate. It has also been known as a sclerosing sweat duct carcinoma (SSDC). We report a case of MAC initially masquerading as a benign chalazion.

CASE REPORT
A 66 year old white woman attended the accident and emergency department at the Western Eye Hospital with a 2 week history of red sore eyes. Bilateral blepharitis was diagnosed and a chalazion noted at the medial end of the left lower eyelid, adjacent to the punctum. The only unusual comment made was that the chalazion had “granulation tissue” on its conjunctival surface.

The blepharitis was treated with topical antibiotic and eyelid hygiene and she was listed for minor surgery. However, she had incision of her chalazion elsewhere and did not attend the accident and emergency department for another 18 months when she returned with recurrent symptoms of blepharitis.

On examination, a distorted reddened lower eyelid lump was observed in the exact location of her previous “chalazion” which had apparently remained despite incision. There was some puckering of the anterior conjunctiva associated with a fairly dense fibrous stroma. Focal dural differentiation was evident with occasional keratocysts present.

The patient’s past medical history was notable for systemic lupus erythematosus complicated by renal failure necessitating allograft renal transplantation with prednisolone and cyclosporine immunosuppression for 18 months before her initial presentation with an apparent chalazion.

HISTOPATHOLOGY
The tumour was composed of narrow cords of cells with round or oval vesicular nuclei associated with a variety of synonyms which are now in disuse; sclerosing carcinoma of sweat ducts, malignant syringoma, sebaceous gland carcinoma with syringomatous features, and sclerosing sweat duct (syringomatous) carcinoma (SSDC).5 Clinically it presents in the fourth to seventh decade of life with no sex predilection. Up to 90% of these tumours are facial.6,7

Table 1 Clinical features of malignant eyelid tumours

<table>
<thead>
<tr>
<th>Tumour Characteristics</th>
<th>Treatment</th>
<th>Spread</th>
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<tbody>
<tr>
<td>Basal cell 90% nodular/морфеаформ, ulcerates/bleds, slow growth</td>
<td>Excision biopsy, Mohs’ excision (2–3 mm margin), radiotherapy, cryotherapy</td>
<td>Local extension, neglected tumours may spread to sinus and brain</td>
</tr>
<tr>
<td>Squamous cell 10% ulcerates/bleds/horn, vascular/velvety surface, rapid growth</td>
<td>Mohs’ excision for best cure (3–4 mm margin), relatively radioresistant</td>
<td>Local extension to orbit and sinuses</td>
</tr>
<tr>
<td>Sebaceous cell 1% firm nodular or pagetoid mass, inflamed eyelid, slow growth</td>
<td>Wide excision (6 mm margin), exenteration if eye involved</td>
<td>Local and distant to lymph nodes, lung and liver</td>
</tr>
<tr>
<td>Malignant melanoma 1% lentigo maligna or nodular melanoma, bleeds, medium growth</td>
<td>Wide excision (6 mm margin), prognosis depends on depth of lesion, may have skip lesions</td>
<td>Distant spread to lymph nodes, lung, liver, brain</td>
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Figure 1 (A) Appearance of left lower eyelid before incisonal biopsy. The contour is a distorted by the fibrotic lesion at the medial end of the lid. It is pale with telangiectatic vessels and has indistinct margins. The tumour has no cicia on it. (B) The tumour involves the full thickness of the lid including the posterior lamella. It surrounds the lower punctum and has obliterated its lumen.

Figure 2 (A) In this field there are two keratocysts. Narrow strands of epithelium are embedded in a dense fibrous stroma (haematoxylin and eosin, original magnification × 25). (B) Ductal differentiation in the form of small cysts are present (haematoxylin and eosin, original magnification × 25).

Figure 3 Satisfactory lower eyelid appearance 4 months after tumour excision and reconstruction with Hughes tarsoconjunctival flap and free skin graft to medial half of lower eyelid.

Wide excision by Mohs’ micrographic surgery is indicated8 as the tumour is locally aggressive with perineural, intraneural, and direct spread into adjacent dermis, subcutaneous tissue, muscle, periosteum, and bone.9,10 Solid nests of dark or clear staining cells form nests, strands, or cords in a sclerotic stroma. Large biopsies are necessary for these features to be conserved for evaluation at low magnification; small biopsies have been shown to contribute to misdiagnosis.

The aggressive nature of MAC is not initially recognisable from its slow growth and benign clinical and histological appearance. It is more infiltrative with perineural invasion, than basal cell carcinoma.
Simple excision alone is insufficient as local recurrences are common (recurrence rate over 40%). Late recurrence has been reported, even 30 years after simple excision. Recurrences are greatly reduced with augmented excision and Mohs’ micrographic surgery.

This case of MAC differs from others in that it was mistaken clinically by several ophthalmologists for a chalazion. In particular, our patient had been immunosuppressed following renal transplantation, and it is conceivable that this had a role in its pathogenesis. The association of radiotherapy at a distant site and the development of MAC has been reported. In the past, MAC has variably been thought to be derived from the hair follicle, the eccrine sweat gland, or both. Nowadays, however, the tumour is firmly classified within the spectrum of eccrine sweat gland malignancies. A coherent finding in this tumour is the presence of ductal differentiation which can be made more obvious with the periodic acid Schiff reaction or by EMA/CEA immunohistochemistry.

Other malignant tumours occurring around the eyelids are summarised in Table 1. MAC is a rare tumour which may be differentiated clinically from other eyelid tumours by its slow growth, dense fibrous appearance, and slightly indistinct margin without skin ulceration. It is less common than sebaceous cell carcinoma which may also masquerade as a chalazion. Breast carcinoma metastases do occur in the eyelid and should be considered as a clinical differential diagnosis; however, the histological differential diagnosis does not include breast sweat gland carcinoma metastasis.

Clinicians should have a low threshold for submitting curettages for histopathology from any apparent chalazion which appears solid on incision. In particular, clinicians should be suspicious of solid recurrent chalazions, even if they have a benign clinical appearance which gives very little indication of the tumour’s identity and aggressive behaviour. If in doubt, we recommend a large incisional biopsy for histopathological diagnosis; then wide excision biopsy with Mohs’ micrographic surgery once the histopathology has been confirmed, to prevent recurrence.

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Optic disc dimensions, body length, and body weight

EDITOR—In the optic disc, all visual afference originating in the photoreceptors of more than 1000 mm−2 retina is concentrated on an area of about 2 mm2 to 3 mm2. The retinal ganglion cell axons are nowhere else so densely and tightly packed as in the optic nerve head. It explains the importance of the optic disc for anomalies and diseases of the optic nerve. Regarding the marked interindividual variability of the size of the optic disc,1 we undertook the present study to evaluate whether the dimensions of the optic disc are correlated with the length and weight of the whole body.

CASE REPORT
The study included 517 white subjects (243 women, 274 men) with a mean age of 46.6 (SD 13.0) years (range 8–87 years) and a mean refractive error of less than 2 dioptres. The subjects came to the eye hospital for diagnosis and treatment of glaucoma (n=244), or for diagnostic reasons (n=244) or for dis- cernment of the real size of fundus objects from the premium objective.2

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The same held true when only eyes with a myopic refractive error of less than 4 dioptres were taken into account, or when we considered only eyes with a myopic and hyperopic refractive error of less than 4 dioptres, or eyes with a myopic and hyperopic refractive error of less than 2 dioptres. Dividing the total study group into women and men, both sexes differed significantly (p=0.0001; Mann–Whitney test) in body length and body weight. They did not vary significantly (p=0.45) in optic disc area (2.74 (SD 0.72) mm2 in women versus 2.80 (0.72) mm2 in men).

COMMENT
The results suggest that, in white people, the size of the optic disc is independent of the dimensions of the whole body. Although the Littmann method may underestimate optic disc measurements in myopic eyes,1 one may arrive at this conclusion, since the correlations between disc area and body length and weight also remained statistically insignificant when only eyes with minor refractive errors were taken into account. The result of this study corresponds with the finding that men and women, although varying in body length and weight, did not differ in optic disc area. It agrees with other morphometric studies in which women and men did not vary significantly in retinal surface area and number of retinal photoreceptors and optic nerve fibres while optic disc area was significantly corre- lated with the count of optic nerve fibres and retinal photoreceptors and retinal surface area.3 The finding of the present study that the size of the optic disc is independent of the dimensions of the whole body may be important for optic nerve anomalies and diseases, such as optic disc drusen and non-artic- teric anterior ischaemic optic neuropathy, the frequencies of which are correlated with optic disc size.4

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Orbital Wegener’s granuloma resulting from direct extension of nasal disease through a surgical rhinostomy

EDITOR,—Wegener’s granulomatosis is a multisystem granulomatous disease of unknown aetiology but known to have numerous ocular complications including orbital inflammatory disease. We report a patient with “limited Wegener’s granulomatosis” (Wegener’s disease without renal involvement) who developed an orbital inflammatory mass by direct extension of nasal inflammatory disease, in a patient with known orbital inflammatory disease due to nasal inflammatory disease.

CASE REPORT
A 46 year old man, with a 7 year history of quiescent bilateral nasal sinus disease due to nasal mucosal biopsy proved Wegener’s granulomatosis, was seen in the lacrimal clinic at Moorfields Eye Hospital. He was maintained on daily oral prednisolone 10 mg and azathioprine 150 mg. He had a 7 month history of bilateral epiphora and lacrimal irrigation demonstrated obstruction of the nasolacrimal ducts. Biopsy of the lacrimal sac mucosa at the time of left dacryocystorhinostomy was clear of granulomatous disease and he received no additional postoperative immunosuppression. Postoperatively all healed well and computed tomographic scan (CT scan) showed no orbital disease. At 11 months, while still on the original dose of immunosuppression, he presented with left retrobulbar ache. Visual acuity was unimpaired (Snellen 6/6 right eye, 6/9 left eye) and there was no relative afferent pupil defect. The left globe was displaced superolaterally by 2 mm and there was 3 mm of relative proptosis with no restriction of left ocular movements (Fig 1). An ill defined inferomedial orbital mass, of soft tissue attenuation and in continuity with the nasal cavity at the site of rhinostomy, was shown by CT scan (Fig 2). The proptosis and orbital signs resolved after a 6 month course of reducing dose of oral prednisolone 60 mg and cyclophosphamide 250 mg and the lacrimal drainage fistula remains patent. A right dacryocystorhinostomy was performed 3 months later, while the patient was on an increased maintenance dose of oral prednisolone 17.5 mg and cyclophosphamide 100 mg, and his postoperative course has been uneventful to 24 months of follow up.

COMMENT
Nasolacrimal duct obstruction has been reported in 7% of patients with Wegener’s granulomatosis and dacryocystorhinostomy in these patients may be associated with postoperative wound necrosis and the formation of nasocular fistulae. Although dacryocystectomy has been suggested as a treatment for nasolacrimal duct obstruction in this disease, others have reported more encouraging results with dacryocystorhinostomy.

Extension of nasal Wegener’s granulomatosis into the orbit after dacryocystorhinostomy has not previously been reported, although non-contiguous involvement, probably due to reactivated orbital disease, has been recorded at 1 month after a dacryocystorhinostomy in Wegener’s granulomatosis. The disease is known to involve ocular and orbital tissues and several papers report the coexistence of orbital and nasosinus disease, with erosion of intervening bone. In the case reported here, orbital involvement was the direct result of the disease process passing through an iatrogenic bony opening created during dacryocystorhinostomy and, in view of this, we suggest that such patients should be monitored postoperatively for signs of orbital disease and be considered for additional perioperative systemic immunosuppression.

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Rhodococcus keratitis

EDITOR,—Ocular infection with Rhodococcus is rare. In previous reports Rhodococcus species caused endophthalmitis and keratitis which failed to respond to treatment. We present a case of Rhodococcus keratitis which was successfully treated.

CASE REPORT
Eleven weeks after simple pterygium excision, an otherwise healthy 56 year old woman developed a corneal ulcer. No organisms were identified from corneal scrapes. Resolution had not occurred after 5 weeks’ treatment with topical cefturoxime, gentamicin, and econazole. The patient was referred to Moorfields


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CASE REPORT

An 11 year old black boy was seen because of transient episodes of monocular loss of vision in both eyes. The patient had a long standing history of migraine usually triggered by fatigue, anxiety, hunger, and some foodstuffs. In most of the episodes he complained of severe throbbing hemicranial headache with no visual symptoms, but occasionally he noticed blurred vision in association with the pain. He used to take aspirin or paracetamol (acetaminophen) for pain relief. He also complained of recurring transient loss of vision in the left eye with no headache during intensive physical exercises such as playing soccer and doing gym at school. Four months earlier he had had an episode of headache in the morning followed by decreased sight in his left eye which persisted even after relief of the pain. He was seen at another hospital where a left optic disc oedema was observed and he was given prednisone 60 mg a day for 14 days with a partial recovery.

Ophthalmic examination revealed his best corrected visual acuity to be 20/20 in the right and 20/60 in left eye. The right eye was unremarkable but the patient could read none of the Ishihara plates with the left eye, had a dense centrocaecal scotoma, and a pale optic disc in this eye, as well as a left relative afferent pupillary defect. There was no retinal abnormality.

Laboratory examination including complete blood count, blood chemistry, platelet count, prothrombin time, partial thromboplastin time, protein C, protein S, antithrombin III, and factor V activities were normal. Tests for anti-phospholipid antibodies and for collagen vascular diseases, echocardiogram, chest x-rays, cranial magnetic resonance imaging, and cerebrospinal fluid analysis with immunoelectrophoresis were all normal. A visual evoked response study showed increased latency and reduced amplitude in the left eye. Haemoglobin electrophoresis revealed a double band of A and S mobility. The S haemoglobin was 38%, A haemoglobin 62%, and the red cells sickled under classic conditions. Careful systemic evaluation for possible complications of sickle cell trait was unrevealing.

COMMENT

Infarction in sickle cell disease is probably a very complex and multifactorial process. Its intricate mechanisms still remain to be elucidated but a number of disturbances such as intimal hyperplasia due to abnormal adhesive and procoagulant properties of sickled red blood cells, time to gelation, and platelet changes may play a role.19 Altered vascular reactivity and vasospasm may be also important. Although complications of sickle cell trait are rare they occur under extreme conditions such as vigorous exertion at high altitudes. Reported neurological complications of AS haemoglobinopathy include seizures4 and spinal cord and cerebrovascular thrombosis.5 Migraine with resulting vasoconstriction and spasm may be a factor in triggering vascular occlusion in some of these patients.6 Complicated migraine may even be found in higher prevalence among people with sickle cell trait.7 Posterior ischaemic optic neuropathy has been reported in a patient with SC sickle cell disease and migraine,8 and in a case of SC haemoglobinopathy.9 Our patient presented loss of vision during a migraine attack. Extensive investigation did not suggest demyelinating disease or haematological disturbances.
other than sickle cell trait. The present case is the first to be described with AION in the set of AS haemoglobinopathy and migraine. It is possible that sickle cell trait may increase the risk of AION in patients with migraine.

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