surgery and then radiotherapy. Two cases were treated with radiotherapy alone, and 2 cases received radiotherapy combined with chemotherapy. In the 2 cases treated with radiotherapy alone, 1 was alive at 6 years and the other died at 1 year because of direct extension of the tumour. The 2 cases treated with a combination of surgery and radiotherapy were alive at 30 months and 2 years. Of the 2 cases treated with radiotherapy and chemotherapy, 1 was alive at 4 years, and the other died at 19 months because of a contralateral orbital SCC. There are not enough cases of primary orbital SCC to compare with the outcomes of secondary orbital SCC, which in a study are 100% survival at 2 years but 19% by 9 years.[12]

Primary orbital SCC, although rare, must be considered in the differential diagnosis of orbital masses. The prognosis after exenteration and adjunctive radiotherapy may be better than after radiotherapy alone.

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Acute orbital sarcoidosis with preceding fever and erythema nodosum

A 53-year-old previously healthy Caucasian male presented to an emergency department with fever, well-circumscribed tender erythematous nodules of the shins, bilateral plantar pain, and bilateral ankle swelling and pain of 1-week duration. He was discharged without a diagnosis and his symptoms began to spontaneously resolve over the next 3 days. A month later, he developed new diplopia, left lower eyelid swelling, and a palpable orbital lump.

These new ocular findings prompted a referral to our ophthalmology clinic for further assessment. On examination, best-corrected visual acuities were 20/20, colour plates were full bilaterally, and there was no relative afferent pupillary defect. The left lower eyelid appeared full and an inferior orbital mass was palpable just posterior to the orbital rim with associated increased firmness on retropulsion. The left eye demonstrated limitation of depression in attempted down gaze. There was no proptosis with exophthalmometry or axial displacement of the globe. Findings of the remainder of the eye examination were normal.

Computed tomography of the head revealed a homogenous mass located in the inferior orbit of the left eye. The mass was continuous with the muscle tendons of the inferior oblique and inferior rectus muscles, displacing the globe slightly upward. There were no radiographic signs of bone or globe invasion.

During an orbital biopsy and surgical debulking procedure, the mass was found to encompass the inferior oblique muscle with adherence to the inferior rectus. Pathologic examination demonstrated extensive noncaseating granulomatous inflammation with negative stains for fungal, acid fast, and bacterial organisms, and was therefore felt to be consistent with sarcoidosis. Figure 1 shows the clinical appearance of the patient, imaging, and histology of the biopsy specimen.

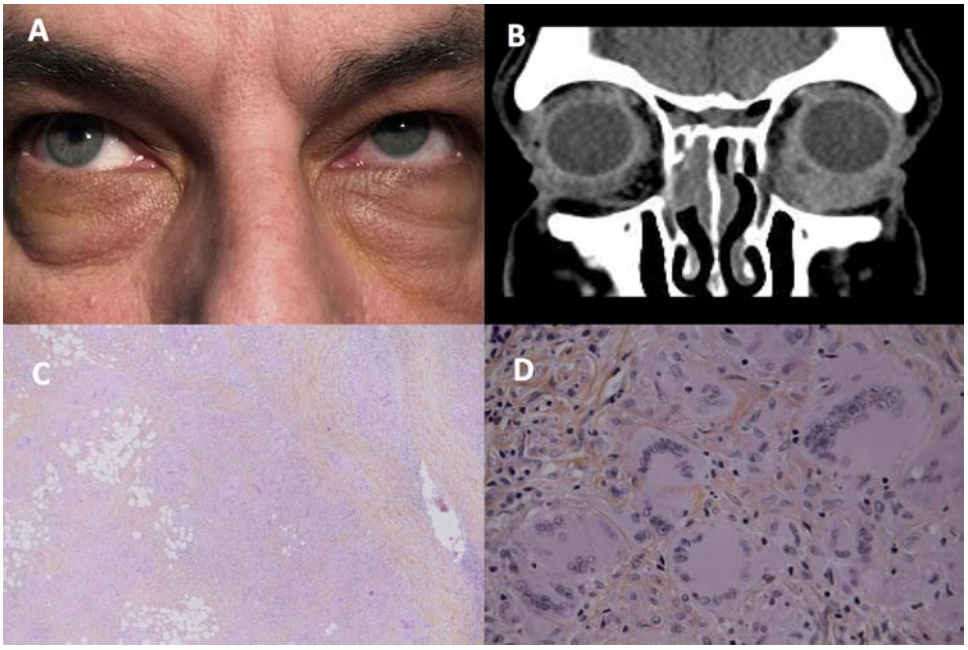

Fig. 1—(A) Clinical appearance of the patient, in upgaze, at time of presentation to our oculoplastic clinic. (B) Computed tomography imaging showing inferior orbital mass involving the rectus muscle. (C) Low-power (50×) and (D) high-power (400×) view of surgically obtained orbital specimen demonstrating replacement of orbital fat by noncaseating granulomatous inflammation and a granuloma with multinucleated giant cells, respectively. All specimens were stained with hematoxylin phloxine and saffron stains.

Blood work completed after the biopsy results yielded a mildly elevated angiotensin converting enzyme level (68 U/L) and C-reactive protein (3.4 mg/L) but normal liver function tests, calcium, and inflammatory markers. Findings of chest x-ray and pulmonary function tests were normal. A diagnosis of Lofgren's syndrome was made based on the constellation of acute onset arthritis, rash consistent with erythema nodosum, fever, and new extrapulmonary sarcoid mass.

After the orbital debulking procedure, the patient's ocular symptoms resolved with the exception of very mild diplopia in far upgaze. Repeat imaging demonstrated a residual mass at the surgical site that was felt to represent scar tissue but persistent disease is a possibility. Given his lack of symptoms, no treatment has been initiated to date. His clinical course has remained stable for the last 11 months and he continues to be followed closely for any sign of disease progression.

Discussion

Ocular and ocular adnexal involvement with sarcoidosis is well known and estimated to affect 25%–60% of people with the disease.[1] Of the ocular adnexal structures, the lacrimal gland is the most likely to be affected.[1] Löfgren syndrome is an acute form of sarcoidosis, characterized by the triad of erythema nodosum and/or inflammatory ankle arthritis, bilateral hilar lymphadenopathy, and fever.[2] A systematic review of the last 20 years identified only a single case of Löfgren syndrome with ocular adnexal involvement that was reported to affect the eyelid.[3]

Löfgren syndrome is considered a distinct clinical entity affecting up to 20%–30% of Caucasians and 4% of Asian[4] and Indian[5] adults with sarcoidosis.

Recognition of the classic symptom pattern is key to the diagnosis. The course of disease is characterized by a prompt and spontaneous resolution of symptoms (erythema nodosum and fever) within approximately 6 weeks to 6 months. Steroids are rarely required in the treatment of this syndrome, and nonsteroidal anti-inflammatory drugs (NSAIDs) are often used if symptomatic relief is necessary. Some experts reason that pathologic confirmation of disease is unnecessary if all clinical diagnostic criteria are met and the symptom resolution proceeds as expected.[6] Common extrapulmonary manifestations include anterior uveitis, lymphadenopathy, and skin granulomas.[1]

Our patient presented with acute orbital sarcoidosis with preceding fever and erythema nodosum. The diagnostic criteria of Löfgren syndrome were not met in the case presented because we failed to detect hilar lymphadenopathy on chest x-ray. Of note, the x-ray was completed several months after onset of initial presentation of symptoms. For a large portion of patients, the resolution of lymphadenopathy occurs within 1 year[7] and up to 50% of patients with suspected sarcoidosis can have a negative chest x-ray but positive bronchoalveolar lavage.[8] Therefore, hilar lymphadenopathy may have initially been present and/or gone undetected for the case described here.

Because of the acute onset of orbital symptoms and overlap in symptomatology with Löfgren syndrome, we elected to withhold steroid treatment and instead treat symptomatically.

To our knowledge this is the first case of acute orbital sarcoidosis to manifest as part of a suspected acute sarcoidosis syndrome. Given the frequently benign course of Löfgren syndrome and lack of need for aggressive treatment, in contrast to other manifestations of ocular adnexal sarcoidosis, it is an important association for ophthalmologists to be aware of.

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Squamous cell carcinoma arising from syringocystadenoma papilliferum of the eyelid

Syringocystadenoma papilliferum (SCAP) is an uncommon sweat gland tumour that is generally considered benign and usually arises in the head and neck region.[1] Typically, it is a pinkish pedunculated nodular lesion with a keratotic or verrucous centre.[2–4] This appearance means that it is often misdiagnosed clinically as a basal cell carcinoma (BCC).[2] We present the first reported case of SCAP in the eyelid that developed into squamous cell carcinoma (SCC).

Case Report

A healthy 50-year-old caucasian female was referred to the oculoplastic team with a painless left lower eyelid lesion of 1-year duration. Examination revealed a solitary 4.5-mm nodular lesion on the left lower eyelid (Fig. 1). The lesion was firm, nonpigmented, and associated with lash loss. The rest of the ocular examination was unremarkable; in particular, there were no associated lymphadenopathy or periorbital sensory changes. Given the findings, a BCC was suspected and the patient consented to an excisional biopsy. She underwent a 2-stage surgical procedure, stage 1 being excision of the lesion with a 3-mm margin of healthy skin and stage 2 being the reconstructive procedure (performed 1 week after stage 1, after complete clearance of the tumour was confirmed histologically).

Histology demonstrated an exophytic and endophytic tumour, with duct-like structures extending as invaginations into the underlying dermis and papillary projections extending into the lumen (Fig. 2A). It was glandular with apocrine differentiation, expressed by decapitation secretions (Fig. 2B). These histological features were in keeping with SCAP. It had a squamous component that was extensively connected to the epidermis and neighbouring hair follicles. The glands appeared fairly bland but were colonized by atypical squamous cells that descended from the overlying component, which displayed full-thickness cytological atypia (Fig. 2C). Nests of atypical squamous cells were also seen to be invading the underlying stroma,

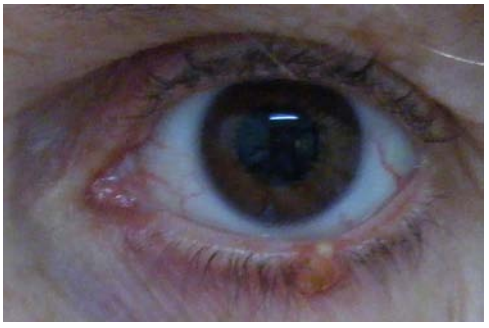

Fig. 1—Photograph demonstrating the clinical appearance of the left lower lid lesion.