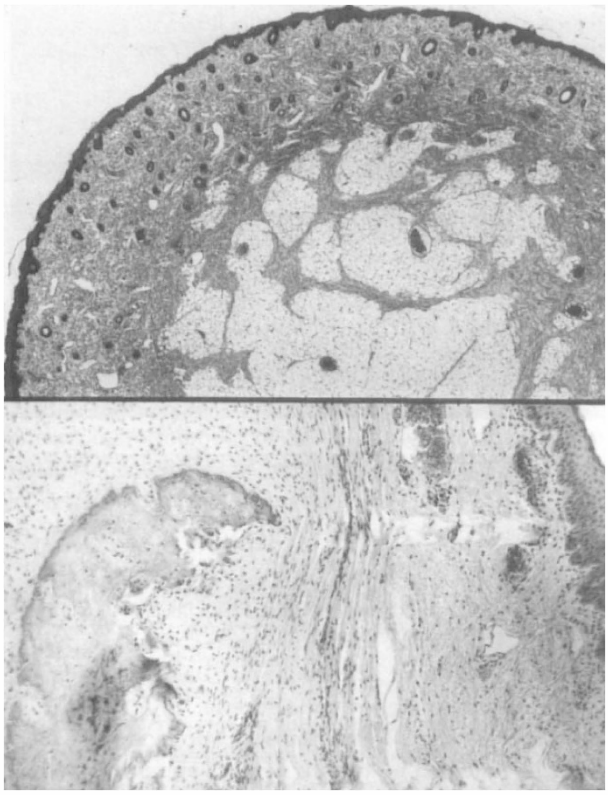

Fig. 2. *Upper: Section of dermoid choristoma covered by skin with pilosebaceous apparatus and sweat glands. The centre of the specimen (left lower corner) shows mature adipose tissue, intersected by fibrous septae. H&E, ×20. Lower: A nodule of peripheral nerve tissue with central cancellous bone surrounded by osteoclasts. H&E, ×80.*

syndrome).[1] Rumbaur[3] in 1920 mentioned the possibility of epibulbar dermoids protruding through the palpebral fissure where they may become pedunculated. A similar case of a pedunculated epibulbar complex choristoma has been reported by Pe'er and Ilsar[4] in association with naevus sebaceous and associated ocular anomalies, viz. choroidal colobomas and corneal vascularisation. Casey and Garner,[5] in their case reports on epibulbar choristomas with microphthalmos, have suggested that intraocular involvement is quite a common association with these masses.

The possible pathogenesis of such an unusual pedunculated episcleral lesion is uncertain since it is poorly understood. The only hypothesis we could offer is that the evolution of this choristoma might have preceded the embryological fusion of the eyelids in the 12th week of gestation. The rationale for such an assumption is that the mass was lying outside the eye with a solitary communication with the temporal conjunctiva through the peduncle. This could suggest that once the palpebral fissure began to close, the lack of available space within the confines of the developing eyeball socket resulted in this mass protruding through the palpebral fissure. The outcome was a pedunculated mass, which continued to grow outside the eye. The presence of a notch in the lateral canthus of the left eye might suggest that the peduncle was lodged here, probably providing the necessary nourishment during development.

After reviewing most of the available literature we concluded that this case was unique in its clinical presentation, since it was an isolated anomaly with no ocular or systemic associations and it had an unusual histological picture.

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Sir,

Conjunctival lymphangioma. A can of worms?

We report a case of conjunctival lymphangioma that was unique in that its appearance was similar to *Loa loa* infestation. Excision and biopsy confirmed the nature of the lesion and there has been no recurrence to date. A review of lymphangiomas is presented.

Case report

A 42-year-old machinist presented to the Coventry Eye Unit with a right corneal foreign body that was removed under topical anaesthetic. An asymptomatic white curvilinear lesion was noted beneath the bulbar conjunctiva of the contralateral eye (Fig. 1). The patient

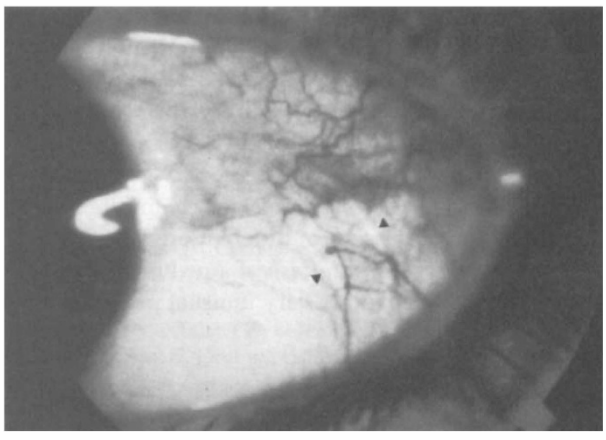

Fig. 1. *Photograph showing the lesion beneath the temporal conjunctiva of the left eye.*

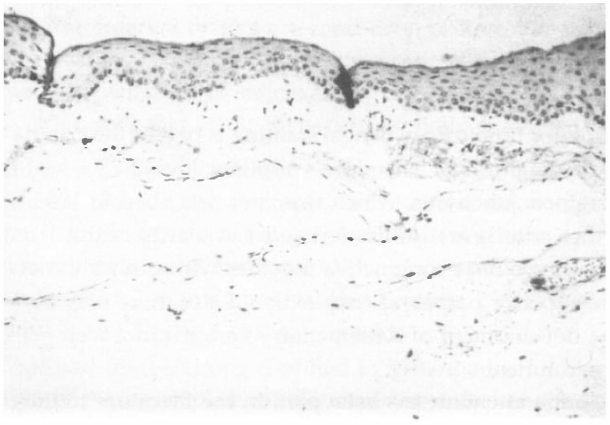

Fig. 2. *Micrograph showing the histology of the lesion. H&E stain.*

was not aware of the lesion or any previous symptoms in that eye and, therefore, there was no evidence of the lesion being progressive. Corrected visual acuity was 6/9 right eye and 6/5 left eye. The lesion was non-pulsatile and compressible. There was no proptosis. Routine examination of the anterior and posterior segments was otherwise normal.

The appearance of the lesion suggested that the patient had a *Loa loa* infestation, although the patient denied having ever travelled to areas where this infestation is endemic. No microfilariae were seen in the anterior chamber. Because of the unusual appearance of the lesion and the curiosity of the medical staff and patient, a decision was made to remove the lesion in order to reach a diagnosis.

The lesion was removed under topical anaesthetic. The specimen was fixed in formalin, sectioned and stained with haematoxylin and eosin. Histology showed a network of empty bloodless channels lined by flattened endothelium. There was no capsule around the lesion. There were no cystic spaces or germinal centres (Fig. 2). A diagnosis of conjunctival lymphangioma was made.

The patient was reviewed and he did not have any cutaneous, nasal or palatal abnormality. The conjunctival wound healed uneventfully and there has been no recurrence of the lesion to date.

Discussion

Lymphangiomas are benign tumours that may affect the orbits, lids or conjunctiva. They tend to enlarge slowly and progressively over a number of years until they reach a relatively static size. Spontaneous or traumatic haemorrhage into the lymph-filled space may cause sudden enlargement. Coexisting lymphangiomas of the face, nasal sinuses, nasal cavity or palate may suggest the diagnosis.[1] Lymphangiomas may also involve the skin elsewhere on the body. Peachey *et al.*[2] described three groups of lymphangioma of the skin: (1) classical lesions involving larger skin areas with subcutaneous muscle-coated cisterns, (2) superficial lesions involving smaller areas without subcutaneous lesions and (3) spongy lesions where skin and mucosa are interwoven with striated muscle, such as on eyelids, tongue and lips.

Conjunctival lymphangiomas are rare.[3] Dilated irregular lymphatic channels appear in the bulbar conjunctiva more commonly than on the tarsal conjunctiva. The average age at presentation is 25 years (range from birth to 65 years), with an average 3 year duration of symptoms at presentation.[4] Lymphangiomas have been histologically divided into capillary, cavernous and cystic types. In a review of 13 cases of orbital-adnexal lymphangiomas, Rootman *et al.*[5] classified these lesions into superficial (as in this case), deep and combined. Superficial lesions consisted of isolated multicystic vascular abnormalities of cosmetic significance only. The deep and combined lesions were found to be more symptomatic, for example presenting with spontaneous haemorrhages and acute proptosis. Histopathologically, lymphangiomas showed evidence of both vascular and lymphatic channels, with the authors questioning the separate classification of tumours of venous and lymphangiomatous origin. Therefore, they concluded that lymphangiomas represent a spectrum of vascular hamartomas. These lesions were characterised by relative haemodynamic isolation.

Treatment of superficial lesions of the skin by surgical excision is 91–100% curative; the larger classical lesions tend to recur.[6,7] Cautery, cryotherapy, radiotherapy and carbon dioxide vaporisation have all been employed as treatment modalities for skin lesions. However, none of the methods prevents recurrence of classical lesions.[8] In Rootman *et al.*'s series[5] only 1 of the 13 patients had a superficial lesion and this was in the lower conjunctival fornix. The treatment was surgical excision, which was uneventful and without recurrence.[5]

The important differential diagnosis of the conjunctival lesion includes vascular lesions such as haemangioma, venous malformation and Kaposi's sarcoma,[9] and non-vascular lesions such as conjunctival dysplasia, conjunctival intra-epithelial neoplasia, and benign and malignant lymphoid hyperplasia.[1] Capillary haemangioma is unlikely since these regress well before adulthood. Furthermore, histological examination did not reveal proliferation of endothelial cells. The lesion was unlikely to be a cavernous haemangioma because of the general absence of blood within the vascular spaces, absence of a capsule and smooth muscle cells and no evidence of secondary changes such as calcification and chronic inflammation. There was no history of proptosis with positional change to suggest venous malformation. The lesion in our case was clinically different from Kaposi's sarcoma, which manifests as red-blue vascular nodules. The histological findings did not support dysplastic or cancerous changes and there was no evidence of lymphocytic proliferation.

Loa loa is a microfilarial infestation endemic to the rain forests of West and Central Africa.[10] It is transmitted by the bite of infected flies of the genus *Chrysops*. The larvae may take years to develop into the adult form, which

may enter many organs and become symptomatic by exciting an allergic response. The commonest clinical manifestations are cutaneous and ocular. Recurrent ('Calabar') swellings may surround the adult worm as it slowly migrates beneath the skin. The adult form may be seen as a 3–7 cm lesion moving slowly through the subconjunctival space. Microfilariae may sometimes be seen free in the anterior chamber. *Loa loa* parasites migrate when alive and cause inflammation when they die. Conjunctivitis, keratitis, anterior uveitis and chorioretinitis have all been described in loiasis.[11]

We have described a case of asymptomatic conjunctival lymphangioma that had been present for an indeterminate period of time. We are sure that it was not a case of infestation with the *Loa loa* worm, but it is interesting that the appearance was so similar.

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Sir,

Diagnosis of Theodore's superior limbic keratoconjunctivitis

Superior limbic Rose Bengal staining is part of the criteria for a diagnosis of Theodore's superior limbic keratoconjunctivitis, which was first described in 1963.[1] Other criteria are infiltration and vascularity of the superior bulbar conjunctiva together with infiltration of the superior palpebral conjunctiva. Later there may be the development of a filamentary keratitis in a wet environment.

Little attention has been paid in the literature to the symptoms of this disease. A special interest of one of us in the disease, over many years, has revealed that the great majority of patients complain of actual pain. There may be feelings of discomfort on awakening in the morning, but the sensation soon passes. Pain then develops and increases as the day progresses. It usually reaches its maximum intensity in the late afternoon, in an office worker. Stress greatly increases the severity of the pain so that a waiter who serves at a busy lunch may have to go and lie down before it is finished. The pain can disrupt lives and lead to thoughts of suicide. It can be said, however, that the pain never interferes with sleep – once the patient's head is on the pillow there is never any problem falling asleep.

The symptoms often bear no relation to the severity of the signs. Minor degrees of superior limbic staining can be accompanied by major symptoms and massive superior limbic disease, where the superior bulbar conjunctiva overhangs the cornea, can be asymptomatic.

Bengal Rose staining of the external eye is not a pleasant procedure. Patients experience a stinging sensation when the drug is inserted, which may last a long time. It is not unknown for a patient to return after the consultation and ask for it to be washed out. Accordingly, the dye is not routinely used in ophthalmic examination. Without its use and without the proper eliciting of symptoms the diagnosis of superior limbic keratoconjunctivitis is usually not entertained. Instead, a diagnosis of dry eyes, lid margin disease or chronic conjunctivitis of the upper palpebral conjunctiva may be made and inappropriate medication may be prescribed. These diseases may be uncomfortable, but are seldom painful.

We were interested to know the incidence of Rose Bengal superior limbic staining in a miscellaneous group of eye patients. Accordingly, we chose to stain 95 consecutive patients attending the morning Eye Casualty at St George's Hospital. Bearing in mind that the degree of Rose Bengal staining is dose-dependent,[2] we laid down a strict methodology for the instillation of the dye. It was done as follows. With the upper lid retracted by the thumb of one hand and the patient looking down, a drop of Rose Bengal was instilled in the area above the superior limbus, taking care not to touch the eye with the applicator. This was a Minim of Rose Bengal (Chauvin).