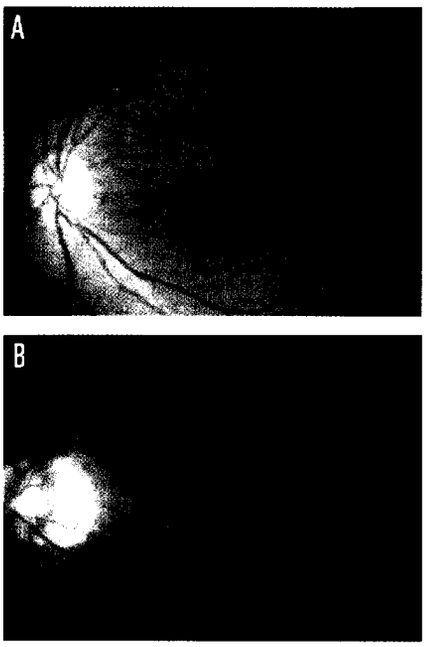

Figure 1 Fundus photographs of cases 1 (A) and 2 (B) showing essentially normal findings.

COMMENT

Focal macular cone ERG is the key to the diagnosis of OMD.[1 3] In our two patients, m-ERG activity was markedly diminished in a relatively small circumscribed area in the macula, suggesting limited functional defects in the fovea. Since no abnormality was found by ICG-V, or FA, the intact RPE may have a barrier effect to the underlying choroidal circulation.

The pathology of OMD may involve the macular cone system without affecting the RPE and the choroid. m-ERG can be useful for the differential diagnosis of OMD and can help map the topography of cone activity loss more precisely. m-ERG may aid in characterising the functional retinal topography in the near future.

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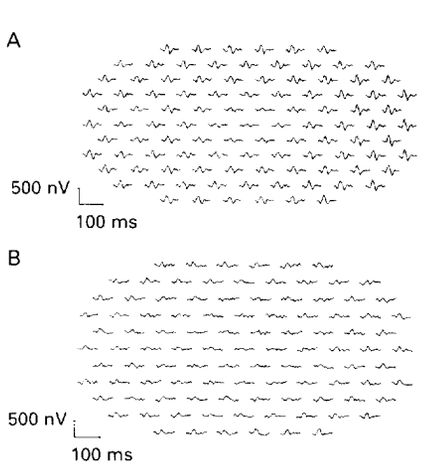

Figure 2 m-ERG of cases 1 (A) and 2 (B) showing decreased amplitudes of the wave patterns in the central 8° and 12° of the macula, respectively.

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Kimura's disease: no evidence of clonality

EDITOR,—Kimura's disease is a chronic inflammatory disorder of unknown aetiology.[1] Patients usually present with recurrent painless swellings in the subcutis of the head and neck region, increased serum IgE levels, and peripheral eosinophilia. The disease is described as reactive and data on clonality is absent.

Here we describe a patient with Kimura's disease involving the orbits. Clonality studies were performed by polymerase chain reaction (PCR) for immunoglobulin heavy chain (IgH), T cell receptor gamma (TCR-γ), and delta (TCR-δ) gene rearrangements.[2-4]

CASE REPORT

A 20 year old man presented with a 2 × 3 cm right eyelid swelling in 1986 with normal visual acuity and absence of diplopia. In 1993, he presented with progressive swelling in the right upper eyelid, which subsided with a short course of prednisolone (50 mg/day × 1 week). He was lost to follow up until May 1997 when he developed recurrent swelling of the right upper eyelid. Excision biopsy of the right upper eyelid mass showed changes consistent with Kimura's disease. DNA was extracted from lacrimal gland biopsy tissue.[5] Gene rearrangements for IgH gene, TCRγ, and TCRδ genes were tested by PCR[2-4] but no clonal gene rearrangement was identified (Fig 1). In June 1998, he had recurrence of the right upper eyelid mass without any local or systemic symptoms (fever, night sweats, weight loss) and multiple left cervical lymph nodes measuring 1–2 cm in diameter. A complete blood examination showed haemoglobin 14.1 g/l, platelets 282 × 10^9/l and leucocytes 18.4 × 10^9/l (differential: eosinophils 3.68 ×10^9/l, neutrophils 9.2×10^9/l, lymphocytes 1.5×10^9/l, and monocytes 4.02×10^9/l) (normal range: leucocytes 4–11×10^9/l; eosinophils 0.1–0.4×10^9/l). The IgE level was 10 328 IU/ml (normal range: <100 IU/ml). IgG, A, and M levels were normal. Urea, creatinine, albumin, and transaminase levels were within normal limits. Serology for HIV was negative. Magnetic resonance imaging (MRI) showed an oval mass in the right lacrimal gland and thickening of the right superior rectus muscle (Fig 2). After 3 weeks of prednisolone (50 mg/day), there was almost complete resolution of the lacrimal mass and IgE level and eosinophil count went down to 2860 IU/ml and 1.2 × 10^9/l respectively. Computed tomography scan of the abdomen revealed absence of intra-abdominal lymphadenopathy or organomegaly.

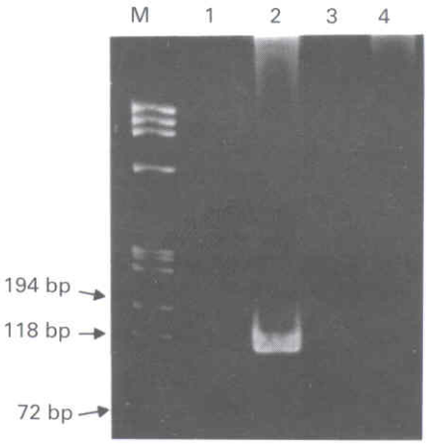

Figure 1 PCR amplification of TCRδ gene rearrangement showing absence of clonal rearrangement in 6% polyacrylamide gel. Lanes: M-ΦX174, HaeIII digest; 1, reagent blank; 2, positive control (B cell lymphoma); 3, water only; 4, patient's lacrimal gland sample.

COMMENT

Neoplasia is characterised by clonal proliferation of cells and is most often demonstrated in cases of malignant diseases. However, monoclonality has also been demonstrated in some "benign" or "reactive" lymphadenopathy such as angioimmunoblastic lymphadenopathy and Castleman's disease, both of which are associated with a tendency to aggressive lymphoma.[6 7]

Kimura's disease runs an indolent course and has been described as a chronic inflammatory process reactive to some "unknown" stimuli.[1] Our patient had a typical clinical presentation with recurrent lacrimal swelling and lymphadenopathy in the head and neck region. It ran an extremely indolent course and, despite the recurring nature of the disease, our patient remained so asymptomatic that he was lost to follow up for years. Interleukin-5 has been shown to be constitutively expressed and explains some of the features of the disease such as eosinophilia and elevated IgE level.[8]

TCRδ gene has been shown by PCR amplification to be rearranged not only in clonal T cell disorders, but also in 73% of clonal B cell disorder.[2] The PCR based methods for the IgH gene rearrangement is positive in 55%–100% of various types of clonal B cell disorders.[4] In our patient, the absence of

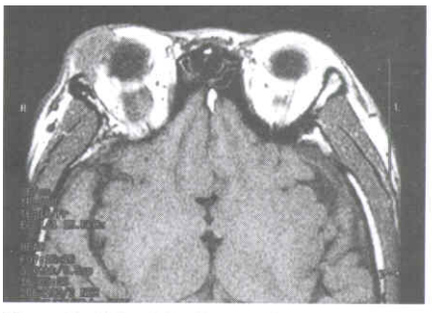

Figure 2 T1 weighted magnetic resonance image of the orbit showing a 2.5 cm diameter mass in the right lacrimal gland with thickening of the right superior rectus muscle.

clonal TCR and IgH gene rearrangements is consistent with the reactive nature of the disease. However, despite the relatively high sensitivity of these PCR based techniques to detect clonality, the finding should be confirmed by testing larger numbers of patients and Southern hybridisation with appropriate probes if DNA from fresh tissue is available.

In conclusion, our patient illustrates the typically indolent, recurring nature of the disease with lymphadenopathy and swelling confined to the head and neck region. The failure to demonstrate clonality is consistent with the reactive nature of the entity and the lack of report of malignant lymphoma transformation.

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Correction of the inadequate lower fornix in the anophthalmic socket

EDITOR,—A well formed inferior fornix in the anophthalmic socket requires an adequate amount of conjunctival tissue and a deep recess. Obliteration of the fornix might occur either secondary to conjunctival shrinkage or as a result of obliteration of the inferior recess and inadequate fixation of the abundant conjunctiva.[1] In the latter condition, the subject of this report, prolapse of the forniceal conjunctiva will result in anterior rotation of the inferior edge of the prosthesis and secondary laxity of the lower lid.

This report presents a modification of the technique described previously by Neuhaus and Hawes for the correction of the above condition.[1]

CASE REPORT

Twenty patients with an inadequate inferior fornix and sufficient amount of conjunctival tissue who presented over a period of 5 years were managed. They all complained of easy prolapse of the lower edge of the prosthesis and sagging of the lower lid.

The procedure was done under local anaesthesia except in the two patients who, in addition, underwent secondary insertion of an orbital implant. A canthotomy and cantholysis were performed. An infratarsal conjunctival incision was carried out along the length of the lower lid. Dissection was then performed in the retro-orbicular plane till the orbital rim was reached (Fig1A). The posterior edge of the conjunctival incision was fixed to the periosteum about 5 mm posterior to the inferior orbital rim with interrupted 5-0 Vicryl sutures. The posterior surface of the lower lid was left to heal by secondary intention (Fig 1B). An adequately sized conformer was inserted. A tarsal strip procedure was performed to tighten the lower lid horizontally.[2] A prosthesis was fitted at about the second postoperative month. The follow up period ranged from 5 months to 3 years. Nineteen patients had a satisfactory result with adequate retention of the prosthesis, and good lower lid level. Inspection of the lower fornix revealed a deep recess with a well epithelialised inner lower lid surface, and no evidence of symblepharon (Fig 2). One patient had loosening of the tarsal strip suture noted at the 1 week postoperative follow up and was later corrected. However, the final lid level remained unsatisfactory.

COMMENT

Adequate retention of the prosthesis in the anophthalmic socket requires a well formed inferior fornix, which in turn requires sufficient conjunctival length and a deep recess.

Obliteration of the inferior fornix might occur despite having a good amount of conjunctival tissue.[1] This occurs possibly because of dehiscence of lower lid retractors, or development of scar tissue in the inferior recess that ultimately results in prolapse of the forniceal conjunctiva and anterior rotation of the lower edge of the prosthesis. The long term effect exerted by the weight and pressure of an improperly accommodated prosthesis will result in secondary laxity of the lower lid.

The traditional solution to the above condition consisted of a lateral canthal tendon tightening and a fornix reformation using an externalised suturing technique in addition to alloplastic stenting material.[3] Skin erosion and infection necessitated early removal of the externalised sutures and increased the risk of recurrence.

Another method of repair was described by Neuhaus and Hawes[1] for the correction of the inadequate inferior cul de sac. It consisted of a transconjunctival inferior fornix incision used to gain direct exposure of the periosteum of the inferior orbital rim. Direct suture fixation of the edges of the conjunctival incision to the periosteum is then achieved. Externalised sutures and stents were not required. Out of 12 patients reported in the above paper, two developed mild lower lid retraction and two developed mild lower lid entropion. This is because the vertical length of the conjunctival tissue is not always sufficient to allow for fixation of both edges of the incision down to the periosteum. Lower lid retraction or entropion occurs whenever the anterior edge of the incision is forced down and sutured under tension.

The technique described in this report makes the conjunctival incision just at the infratarsal border so as to save the maximum length of conjunctiva for the posterior flap. By this, the inner lid surface is left to heal by secondary intention. The tarsal strip procedure performed during the surgery aims to eradicate the lower lid laxity and sag.[2]

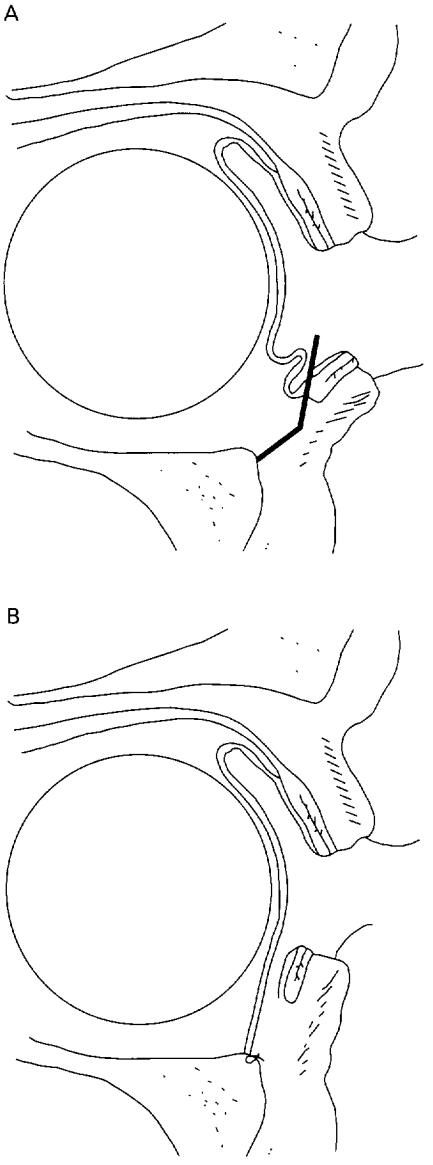

Figure 1 (A) The bold line marks the dissection plane. (B) The posterior conjunctival edge sutured down to the periosteum.

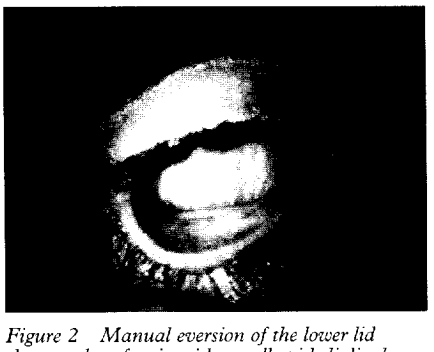

Figure 2 Manual eversion of the lower lid shows a deep fornix with a well epithelialised inner lid surface and no symblepharon.

In conclusion, this modified technique allows the use of internal fixation to correct the lower fornix while minimising the risk of lower lid retraction or entropion.

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