Giant dacryops in a patient with ocular cicatricial pemphigoid

EDITOR,—The term ‘dacryops’ was introduced by Schmidt in 1803 when he described cysts arising from the palpebral lobe of the lacrimal gland.1 Its formation is believed to be caused by occlusion of the lacrimal duct openings due to conjunctival inflammatory or traumatic changes followed by dilatation of the inflamed and weak duct walls. Conditions such as trachoma which cause conjunctival scarring have been reported as antecedents of ductal cyst formation.2 Ocular cicatricial pemphigoid (OCP), a relatively rare, chronic, progressive disease causing significant conjunctival scarring to our knowledge has not been documented as a cause of dacryops. We describe a patient with OCP who had a giant cyst on his left upper lid which was proved on histopathology to be a lacrimal duct cyst.

CASE REPORT

A 63-year-old male patient, diagnosed 16 months earlier as having OCP, developed massive swelling of the left upper lid over 1 month (Fig 1A). No improvement was noted with either antibiotic or steroid treatment. B-scan ultrasound demonstrated a cystic lesion which on aspiration revealed 6 ml of clear yellow fluid. The decompressed cyst reformed within 24 hours. A computed tomogram showed that the cystic mass, which measured 3-4 x 1-8 x 1-8 cm, was confined to the left preseptal space slightly displacing the left eyeball posteriorly (Fig 1B). The patient underwent total excision of the cyst through a lid crease incision. The cyst was noted to be posterior to the markedly attenuated levator aponeurosis and Müller’s muscle, adherent to the superior border of tarsus, and in a subconjunctival location. A portion of the palpebral lobe of the lacrimal gland to which the cyst was attached was also excised. Histopathology showed the cyst wall (Fig 2A) and dilated ducts of the lacrimal gland with surrounding acute and chronic non-granulomatous inflammatory reaction (Fig 2B) consistent with a diagnosis of lacrimal duct cyst with dacryoadenitis. No recurrence of the cyst was noted for the next 3 months.

COMMENT

Ocular cicatricial pemphigoid is a chronic disease characterised by conjunctival shrinkage, entropion, trichiasis, xerosis, and corneal opacification causing blindness.3 Fibrous occlusion of the lacrimal ducts in OCP can cause decreased aqueous tear production. The development of lacrimal duct cyst, however, has not been described as a common finding. Cyst formation as a result of collection of tears proximal to the obstruction does not readily occur because obliteration these ducts leads to gland atrophy and cessation of secretion.1

In this patient we believe that the dacryops originated from an inflamed main lacrimal gland duct containing tears produced by the

Figure 1 (A) A 63-year-old male patient with ocular cicatricial pemphigoid and a giant cyst on the left upper lid. His left eye was covered by the left upper lid cyst that caused stretching of both the lid skin and the upper palpebral conjunctiva. (B) Computed tomography scan demonstrates the preseptal location of the cystic mass (arrow) displacing the left globe posteriorly.

Figure 2 (A) Portion of the giant cyst wall with surrounding haemorrhagic connective tissue containing inflammatory cells. (Haematoxylin and eosin, × 15.) (B) Part of the main lacrimal gland showing moderate infiltration of lacrimal gland acini with acute and chronic inflammatory cells. (Haematoxylin and eosin, × 38.)
remaining normal main lacrimal gland acini. The cyst size and its rapid reaccumulation following aspiration supports this theory of its origin since the main lacrimal gland produces tears at a rate that can exceed 50 μl/min as opposed to the basic secretion from the glands of Kraus and Wolfring that produces less than 0.3 μl/min. In addition to ductal occlusion secondary to conjunctival scarring, we postulate that the inflammation around the ducts as seen histopathologically may have caused the ductal walls to weaken and more readily become ectatic forming this glandular cyst.

Complete cyst excision by the transconjunctival approach is usually recommended. 1, 2 In this particular patient, aside from the difficulty of exerting the lid to approach the cyst subconjunctivally, violating the conjunctiva in the setting of OCP is not advisable. Manipulation of the conjunctiva may provoke an acute flare of disease activity as observed in a previous report. 3 The lid crease approach also offers the lesion of the levator aponeurosis dehiscence which could be repaired primarily. Simple aspiration is not recommended since this will result in immediate recurrence.

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HEIDI D REMULLA
PETER AD RUBIN
Eye Plastics and Orbit Service,
Massachusetts Eye and Ear Infirmary,
Harvard Medical School,
Boston, USA

Correspondence to: Peter AD Rubin, MD, Division, Eye Plastics and Orbital Surgery, Massachusetts Eye and Ear Infirmary, 243 Charles Street, Boston, MA 02114, USA.

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Angiolympoid hyperplasia with eosinophilia (Kimura's disease) of the conjunctiva

EDITOR—Kimura's disease is a rare chronic vascular inflammatory disorder occurring predominantly in orientals, with occasional cases in white and black people. 1 The disease first appeared as 'hypertrophic lymphadenopathy' in the Chinese literature, but the disorder is commonly known as Kimura's disease, following the report of Kimura et al in the Japanese literature. 2 Kimura's disease predominantly involves the head and neck region, and orbital involvement has been reported. 1 The lesions may be isolated or multiple and vary in clinical appearance.

Histopathologically, the lesions are composed of vascular hyperplasia with plump endothelial cells accompanied by varying degrees of mixed cellular infiltrate dominated by lymphocytes and eosinophils. 3 Conjunctival Kimura's disease, to our knowledge, has not been described in the English language published reports.

CASE REPORT

A 24-year-old white girl presented in March 1994 with a painless ulcerated white lesion of the bulbar conjunctiva of the right eye. There was no history of trauma. She was otherwise in good health.

On examination visual acuity was 6/4 bilaterally. An ulcerated, mobile firm swelling was noted in the inferotemporal aspect of the bulbar conjunctiva, with no associated lymphadenopathy. Systemic examination was normal.

Routine investigations full blood count, sedimentation rate, serology, and angiotensin converting enzyme were normal. Serum concentrations of immunoglobulin IgA and IgM were elevated. The peripheral blood eosinophil level was normal.

 Conjunctival scrapings for bacteria, viruses, chlamydia, and fungi were negative. A chest radiograph was also normal. Computed tomography demonstrated an anterior orbital soft tissue swelling with no evidence of a foreign body within the lesion.

Treatment with topical steroids and systemic antibiotics led to no improvement in the clinical condition.

The lesion became increasingly uncomfortable, and extended into the inferior tarsal conjunctiva. Figure 1 shows the yellow-white nodular mucosal plaque measuring 15 mm by 10 mm. Biopsy and debulking of the lesion was performed. The histopathological examination revealed a tan-coloured nodule which measured 15×10×7 mm. Ulceration of the conjunctiva was present with underlying prominent vascular inflammatory stroma.

The base of the ulcer showed inflammatory granulation tissue and included neutrophils, eosinophils, and lymphocytes. Lymphoid aggregates with reactive germinal centres were also present (Fig 2 top). Abundant blood vessels lined by plump endothelial cells and surrounded by lymphocytic and eosinophilic infiltration were also prominent features (Fig 2 bottom). Histiocytic granulomata were not seen. Special stains showed no organisms within the ulcer. The histopathological appearance was that of angiolympoid hyperplasia with eosinophilia.

After a course of topical steroids there was no change in the residual lesion and further debulking with intralecular steroids was performed.

COMMENT

Kimura's disease is an inflammatory process of unknown aetiology, with a predilection for the head and neck region. Controversy exists as to whether it represents an allergic response to a continuing antigenic stimulus or a benign lymphoid neoplastic process. Lymphadenopathy, peripheral blood eosinophilia, and elevated serum immunoglobulin IgE are features of Kimura's disease. 2 In this patient an elevated serum immunoglobulin IgA and IgM was noted. Systemic associations include asthma and nephrotic syndrome. 3 The long term natural history of the disease is uncertain. Whether Kimura's disease and angiolympoid hyperplasia with eosinophilia represent two distinct clinicopathological disorders, 4 or different appearances of the same disease is still debated and is beyond the scope of this report.

Kimura's disease may present as a solitary or multiple lump(s) of insidious onset. Orbital disease may be preceded by other foci of involvement, such as the parotid gland. 4 The first orbital case was recorded in 1976, and in 1983, a series of five orbital and three adnexal cases was published. 3 The mean age of patients with orbital disease was higher than the mean age at other sites. The lesions were solitary, and the location of the lesions varied from the eyebrow to the roof of the orbit. Kimura's disease occurring in association with an Iowa enucleation implant has also been reported. 2 Conjunctival Kimura's disease has not been reported in the English language published reports.

The histopathological features of Kimura's disease include endothelial cell proliferation, eosinophilic infiltration, lymphoid aggregates with florid germinal centres, fibrous sclerosis, and giant cells of the Warthin-Finkeldey type. The pathological differential diagnosis of Kimura's disease includes inflammatory pseudotumour but the eosinophils in these cases are not usually such a conspicuous feature. Other disease processes such as Hodgkin's disease can be differentiated by the presence of Reed-Sternberg cells and bacillary angiomatosis by proliferating endothelial cells in association with clumps of bacteria.

Previously reported cases of periocular Kimura's disease were successfully treated with excisional biopsies, though excessive bleeding during the surgery was noted. 3 The systemic use of corticosteroids in orbital disease is also recognised though there may be recurrence. 6 Skin disease has been successfully treated with intralesional triamcinolone, carbon...