SY Shen and BCC Liang

Department of Ophthalmology
United Christian Hospital
130 Hip Wo Street
Kwun Tong
Kowloon
Hong Kong

Correspondence: SY Shen
Tel: +852 2379 4509
Fax: +852 2775 3157
E-mail: drphoebus@yahoo.com

Sir,

Allergic eye disease associated with mastocytosis
Eye (2003) **17,** 788–790. doi:10.1038/sj.eye.6700467

Mastocytosis is a rare disease characterized by an abnormal proliferation of normal, active mast cells. The disease can present at any age. Symptoms of mastocytosis occur when pharmacologic or physical stimuli cause mast cell degranulation with release of histamine, prostaglandin D2 (PGD2), leukotrienes, heparin, and proteolytic enzymes. Ocular involvement in mastocytosis has been previously documented.[1–3] We report a patient with severe allergic eye disease associated with the condition. To the best of our knowledge, this has not been reported before.

Case report

A 47-year-old lady presented with 2 weeks' history of increasing lid swelling, redness, watering, and itching of both eyes. Her symptoms initially started and were more severe on the left side. She had been seen at the onset of her symptoms by an ophthalmologist when a diagnosis of herpes zoster ophthalmicus was made, and she was commenced on oral acyclovir. Her symptoms worsened and she presented to our department.

On examination, the patient was found to have diffuse facial erythema and oedema. She had bilateral lid swelling, diffuse conjunctival injection with papillary hypertrophy, and watery discharge (Figure 1a). Anterior and posterior segment examination was otherwise normal. The patient had been previously diagnosed with systemic mastocytosis on the basis of clinical signs and symptoms and a positive bone marrow biopsy (Figure 2), and was being treated with systemic antihistamines and cromolyn sodium.

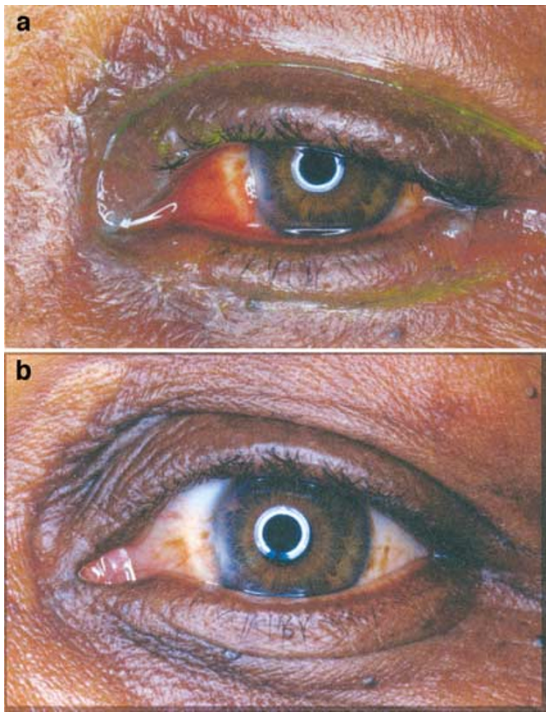

Figure 1 (a) Photograph of the left eye showing lid oedema, conjunctival injection, and watering. (b) Post-treatment photograph of the patient's left eye showing resolution of all signs.

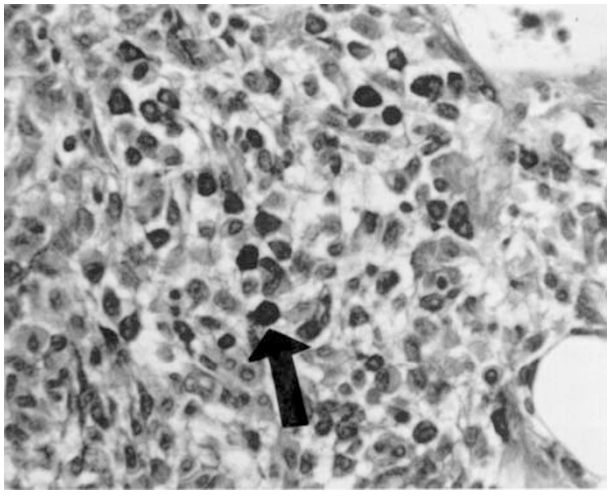

Figure 2 Specimen of a bone marrow biopsy from the patient, stained with Giemsa stain, at $\times 400$ magnification. Note the abundance of mast cells (increased compared to normal) with dark blue staining cytoplasm (arrow).

After consultation with the patient's haematologist, she was treated with a short course of oral steroids, topical antihistamine (emedastine 0.05% eye drops), mast cell stabilizer (sodium cromoglycate 2% eye drops), and

topical steroid (prednisolone sodium 0.05% eye drops) with a remarkable improvement in her symptoms and signs (Figure 1b). Her symptoms are controlled on a maintenance dose of topical antihistamine and sodium cromoglycate 2% eye drops. Alternative topical mast cell stabilizers like lodoxomide and nedocromil were not employed initially as most acute-phase symptoms settled with the use of systemic steroids, and subsequent control of symptoms was achieved with the above medications. A biopsy of the conjunctiva was felt to be unjustifiable as the patient's initial and subsequent symptoms were controlled on the above regime.

Comment

Mastocytosis is a disorder characterized by an abnormal proliferation of tissue mast cells. There are cutaneous and systemic forms of the condition. The increase in mast cells may be (1) generalized, (2) discrete as in urticaria pigmentosa, or (3) present as a single collection of cells in solitary mastocytosis. Systemic mastocytosis is caused by the accumulation of mast cells in the tissues and can affect organs such as liver, spleen, bone marrow, and small intestine.

The prevalence of mastocytosis in the general population is unknown but it is generally considered to be an 'orphan disease' (200 000 or fewer people in the US). Mastocytosis occurs in all races and there is no sex predilection. The peak incidence is during infancy and early childhood with a second peak in middle age.

Mast cells originate from bone marrow progenitor cells and are distributed in the connective tissues. They are concentrated in the skin and peripheral nerves, and adjacent to blood and lymphatic vessels. Activation by immunoglobulin E or other stimuli causes the mast cells to degranulate and release preformed mediators of inflammation that initiate the acute and delayed hypersensitivity reactions associated with the allergic mechanism cascade and the various cutaneous and systemic manifestations of mastocytosis.

The symptoms in patients with mastocytosis are generally related to the increased release of mast cell-derived mediators such as histamine, PGD2, peptide leucotrienes, platelet-activating factor, heparin, and proteolytic enzymes. Chemicals released by mast cells cause physiological changes that lead to typical allergic responses such as hives, itching, abdominal cramping, bone pain, nausea, vomiting, diarrhoea, hypotension, or even anaphylactic shock.[1]

The primary event responsible for mast cell proliferation in mastocytosis is largely unknown, but a derangement of the network involving c-kit receptor and its natural ligand, the stem cell factor that promotes mast cell growth, and differentiation in humans is the likely cause.[1]

Ocular involvement in mastocytosis has been described as solitary mastocytoma of the eyelid,[2] painful orbital lesions,[3] and nyctalopia caused by the malabsorption of Vitamin A.[4]

Diagnosis of the cutaneous forms of the disease can be through the abnormally high concentrations of mast cells in the skin. Likewise, the diagnosis of systemic mastocytosis can be made by a biopsy showing an increase in mast cells in the affected organ with the use of special stains such as giemsa and toludine blue. Plasma tryptase and histamine levels are persistently elevated, and urine may contain high levels of histamine and PGD2 metabolites in these patients.[1]

The treatment of mastocytosis is largely symptomatic. An array of drugs is used, including antihistamines (H1- and H2-receptor blocking agents) to control itching, skin complaints, and pathological gastric hypersecretory conditions. Severe flushing and hypotension are treated prophylactically with antihistamines and with epinephrine after symptoms begin. Cromolyn sodium helps to stabilize mast cell membrane. Nonsteroidal anti-inflammatory agents and sometimes steroids are used to inhibit the formation of PGD2. Interferon and photochemotherapy with psoralen and ultraviolet A irradiation (PUVA) are being tried in the management of this condition. In rare cases where mastocytosis is malignant, chemotherapy is necessary.

References

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A Shaikh and L Benjamin

Department of Ophthalmology
Stoke Mandeville Hospital, Mandeville Road
Aylesbury, Buckinghamshire HP218AL, UK

Correspondence: A Shaikh
Tel.: +44 1296 315034
Fax: +44 1296 315037
E-mail: asifasshaikh@aol.com

Sir,

Surgical removal of sequential epiretinal and subretinal neovascular membranes in a patient with traumatic choroidal rupture

Eye (2003) **17,** 790–791. doi:10.1038/sj.eye.6700472

Fibrocellular epiretinal membrane formation and subretinal neovascularisation (SRN) are documented sequelae of traumatic choroidal rupture. We herein report a case that illustrates the development of these complications at different time points following injury and the successful surgical management of both pathologies.

Case report

An 18-year-old male sustained a left closed-globe contusion injury in a road traffic accident, and developed vitreous haemorrhage. He was referred for vitreoretinal management 3 months later following initial observation at a local hospital. His visual acuity was 6/12 with marked distortion. Fundoscopy revealed a crescent-shaped choroidal rupture concentric to the optic disc and an epiretinal membrane extending from the rupture (Figure 1). His vision returned to 6/6 after vitrectomy and surgical peeling of this membrane, despite a faint subretinal haemorrhage. Fluorescein angiography confirmed a subretinal neovascular membrane extending from the rupture towards the fovea (Figure 2a). Given his good visual acuity, and previous experience of spontaneous neovascular membrane regression, we elected to observe his progress. After 4 months, his visual acuity was 6/12 and the membrane had extended subfoveally (Figure 2b). Surgical removal of the membrane was planned, prior to which his visual acuity had deteriorated to 6/24 with clinical evidence of membrane extension and increased subretinal haemorrhage. Following surgical removal via a small retinotomy nasal to the choroidal rupture, his vision improved and was stable at 6/9 (with no significant distortion) 3 months after surgery, without membrane recurrence. Histological examination of the subretinal neovascular membrane demonstrated RPE cells on one surface and within a fibrovascular core, and no evidence of photoreceptors or Bruchs membrane components.

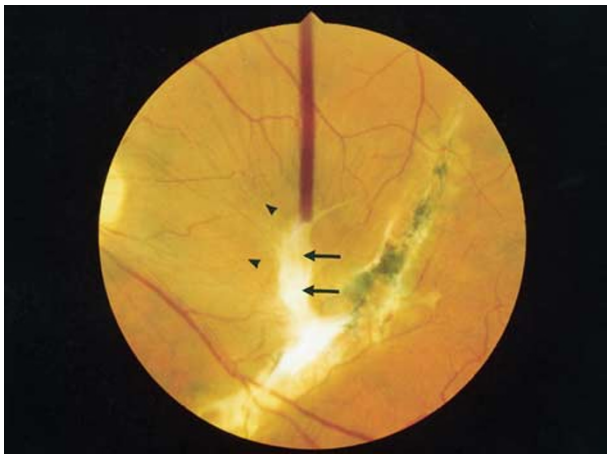

Figure 1 Left eye fundus demonstrating choroidal rupture extending to the macula, with epiretinal membrane formation (arrow) and a striated appearance of adjacent neuroretina secondary to traction (arrowheads).

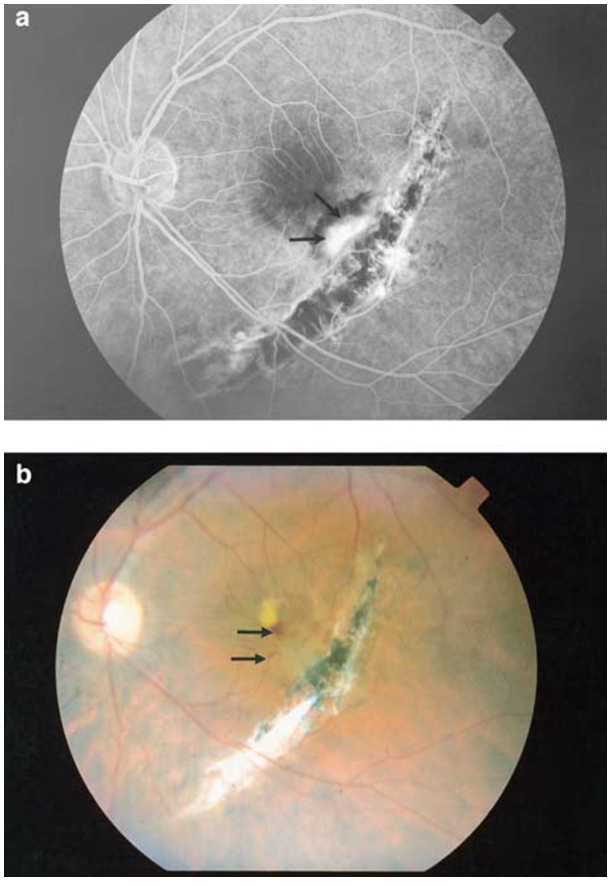

Figure 2 (a) Fluorescein angiogram confirming the presence of a neovascular membrane nasal to the choroidal slar (arrows). (b) Left eye 7 months after injury: the neovascular membrane has extended subfoveally, note subfoveal haemorrhage (arrows) and lipid deposit.