A healthy 43-year-old officer of a merchant ship at sea developed pain, redness, and photophobia in his right eye. During the next 2 weeks, he noted the presence of a band of opacity spreading from his temporal limbus toward his central cornea. His episcleral vessels were engorged in a distribution contiguous with the peripheral, sectorial, fleck-like corneal opacities. The opacity had progressed during topical and systemic antibiotic therapy, but halted with use of topical corticosteroids. Systemic evaluation showed mild IgM monoclonal gammopathy. Transmission electron microscopy of a corneal biopsy specimen revealed electron-dense fibrils identified as immunoprotein. To our knowledge, this is the first report of a case of acute unilateral deposition of corneal immunoprotein in a patient with monoclonal gammopathy. Clinicians should begin with a broad differential diagnosis when evaluating patients with corneal opacity.

Immunoprotein deposition is a rare cause of corneal opacity. In general, the immunoprotein deposits occur bilaterally, with gradual onset. We report a case of immunoprotein deposition in the right cornea of a 43-year-old British sailor, 2 weeks following the onset of redness and pain in that eye. We believe that this is the first reported case of acute-onset unilateral corneal immunoprotein deposition in a patient with a monoclonal gammopathy.
The deposits can be initial or early manifestations of these systemic illnesses. Deposits generally develop gradually and involve both eyes. Although some patients experience no reduction in visual acuity, many require treatment of the underlying disease with cytotoxic chemotherapy or corticosteroids to restore visual function, and penetrating keratoplasty may be required in severe cases. Based on initial evaluation, the patient in this case seemed to have monoclonal gammopathy of unknown significance.

Close examination of the distribution of engorged ocular surface vessels depicted in Figure 1, left, suggests a connection between the inflammatory process and the immunoprotein deposits. The inferotemporal and inferonasal quadrants show significant hyperemia, with dilated conjunctival and episcleral vessels, suggesting bisectional episcleritis. The bands of immunoprotein are widest in the locations where there is significant hyperemia. In this patient, dilated, permeable vessels may have allowed exudation of large IgM molecules. Although some researchers argue that the tear film is a transport medium for immunoproteins into the cornea, this case is most consistent with recent reports indicating the paralimbal vascular arcades as the principal route of influx.

The response of this patient to topical corticosteroid therapy also suggests an inflammatory leakage mechanism. Topical prednisolone apparently halted the central progression of the corneal deposit, and spared the patient significant loss of visual acuity. The rapid progression of this patient’s corneal deposition, although not documented by an ophthalmologist, is likely to be correct since the patient was the ship’s medical officer and provided written records of his course prior to evaluation. This case widens the spectrum of findings of corneal immunoprotein deposition, and reminds us to begin with a broad differential diagnosis when evaluating patients with corneal opacity.

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REFERENCES


Figure 1. Slitlamp photographs of a patient with corneal immunoprotein deposition. Left, Before corneal biopsy. The inferotemporal band of deposition is clearly visible. There is a subtle asymmetry in the episcleral and conjunctival hyperemia, which is greater in the area contiguous with the inferotemporal band. Right, After corneal biopsy. The slitlamp beam indicates the posterior stomal location of the deposition. The nasal band of deposition is also visible.

Figure 2. Transmission electron micrograph of the corneal biopsy specimen. Electron-dense fibrils with a faint periodicity at 16.6 nm are present.

IgM level was 2.2 g/L (normal range, 0.6-2.1 g/L).

A right inferotemporal corneal biopsy was performed. One portion of the tissue was fixed in formalin for light microscopy. A second portion was fixed in a buffered mixture of 1% glutaraldehyde and 4% formaldehyde for transmission electron microscopy.

The part of the biopsy specimen processed for light microscopy showed no corneal deposits. Transmission electron microscopy revealed electron-dense fibrils with a faint periodicity of 16.6 nm (Figure 2). These fibrils had the characteristic appearance of immunoprotein.

COMMENT

Diffuse or focal corneal immunoprotein deposits are a relatively uncommon manifestation of several systemic illnesses, including multiple myeloma,1 monoclonal gammapathy of unknown significance,2 Waldenstrom macroglobulinemia,3 certain lymphoproliferative disorders,4 and leukemia.5 The deposits can be initial or early manifestations of these systemic illnesses.1,3-5 Deposits generally develop gradually and involve both eyes.1,4 Although some patients experience no reduction in visual acuity, many require treatment of the underlying disease with cytotoxic chemotherapy or corticosteroids to restore visual function, and penetrating keratoplasty may be required in severe cases. Based on initial evaluation, the patient in this case seemed to have monoclonal gammopathy of unknown significance.

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