

this organism to cause severe orbital disease and to emphasize the need for early aggressive debridement and treatment with a prolonged course of antibiotics which are effective against anaerobic bacteria.

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Sir,

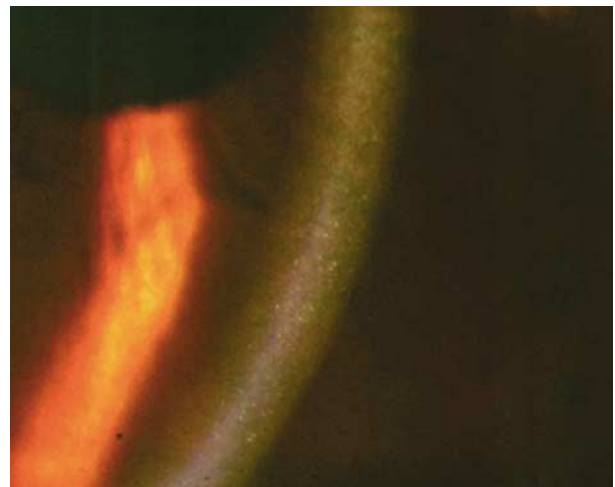
## Corneal crystalline deposits as the initial manifestation of IgA-kappa multiple myeloma

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Corneal crystalline deposits of immunoglobulin origin have been reported to occur in a variety of hypergammaglobulinaemic states, including multiple myeloma, benign monoclonal gammopathy, cryoglobulinaemia, Waldenström's macroglobulinaemia, rheumatoid arthritis,<sup>1</sup> other associated neoplasms,<sup>2</sup> and after immunoglobulin therapy.<sup>3</sup> The most frequently reported monoclonal gammopathy associated with corneal crystalline deposits is of the IgG-kappa light chain. We report an unusual case, initially presenting with corneal crystalline deposits, who developed IgA-kappa multiple myeloma 2 years later. To the best of our knowledge, only one case of corneal crystalline deposits associated with IgA-kappa monoclonal gammopathy<sup>2</sup> has been described in the literature. However, bone marrow pathology of that patient was normal.

## Case report

A 62-year-old woman was examined in March 1999 because of dry eye symptoms. Her best-corrected visual acuity was 20/25 OU. Slit-lamp examination disclosed tiny, colourless to white, crystalline deposits distributed throughout both corneas and within all layers of the stroma (Figure 1). The Schirmer test result (under topical anaesthesia) was 4 mm OU. Fundoscopic examination was normal. The remainder of the ocular examination was unremarkable. The systemic workup disclosed



**Figure 1** Slit-lamp examination disclosed tiny, colourless to white, crystalline deposits distributed throughout both corneas and within all layers of stroma.

pancytopenia (white-cell count, 1690/mm<sup>3</sup>; red blood cell count, 2.69/mm<sup>3</sup>; platelet count, 24 000/mm<sup>3</sup>) and splenomegaly. Electrolytes were normal. Uric acid, lipid profile, renal function, and liver function were normal. Urine for Bence-Jones protein was negative. Bone marrow biopsy showed increased histiocytes. She underwent splenectomy in July 1999. During the ensuing years, the crystalline deposits increased progressively and became polychromatic. Serum immunoglobulin levels were checked in June 2001 and showed an elevation of IgA to 1400 mg/dl (normal 158–358 mg/dl). The IgG level was 911 mg/dl (normal 1188–1800 mg/dl). The IgM level was 39.5 mg/dl (normal 72–216 mg/dl). Skull and thoracic spinal X-rays revealed no osteolytic lesions. Urinalysis for Bence-Jones proteins was now positive. Serum immunoelectrophoresis revealed a monoclonal gammopathy involving IgA-kappa light chain. A repeat bone marrow biopsy demonstrated increased plasma cells to 15%. The diagnosis of IgA-kappa multiple myeloma was made in November 2001. Systemic therapy was initiated with melphalan and prednisolone. Serum IgA/IgG/IgM levels were 1210/818/25.4 mg/dl, respectively, 13 months after chemotherapy. Her best-corrected vision remained at 20/25 OU. The corneal crystalline deposits persisted. She rejected a biopsy of the cornea because her vision was not affected.

## Discussion

Corneal crystalline deposits associated with multiple myeloma were first described by Burki in 1958.<sup>4</sup> Since then, a variety of immunoprotein deposits have been described in the cornea, among which, IgG-kappa light-chain deposition is most frequently reported. Only one case of IgA-kappa chain monoclonal gammopathy associated with deposition has been reported previously.<sup>2</sup> That patient's best-corrected vision was reduced to counting fingers in the right eye and 20/80 in the left eye because of macular drusen in both eyes. The authors suggested a specific association between IgA monoclonal gammopathy and macular drusen, because drusen were not found in IgG monoclonal gammopathy. In our case, however, we did not observe macular drusen.

Presenting initially as crystalline keratopathy has also been reported to occur in patients of IgG benign monoclonal gammopathy.<sup>5</sup> One study showed that 19% of 241 patients with a diagnosis of benign monoclonal gammopathy developed myeloma, or a related disorder when followed for 10 years or more.<sup>6</sup> Morphological changes of the corneal crystals were noted previously in a patient of IgG-kappa monoclonal gammopathy.<sup>7</sup> The deposits changed from dot-like opacities to polychromatic crystals, at which time multiple myeloma

was diagnosed. Similar alteration of the corneal crystals and malignant transformation of the underlying disease were observed in our patient. The corneal crystals in the previously reported case of IgA benign monoclonal gammopathy appeared colourless to slightly white,<sup>2</sup> which perhaps indicated an early stage of the disease process. Ophthalmologists should be aware of these manifestations and the potential for developing malignancies.

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Sir,

## The varix of angular vein

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These cases describe an unusual cause for a paranasal swelling, the lesion mimicking a lacrimal sac mucocele,