Superficial reticular degeneration of Koby

HENRY D. PERRY¹ AND HAROLD G. SCHEIE²

From the ¹Department of Ophthalmology, University of Pennsylvania School of Medicine and the Scheie Eye Institute, Philadelphia, Pennsylvania 19104, and ²Long Island Jewish-Hillside Medical Center, New Hyde Park, New York 11042, USA

SUMMARY A 36-year-old white man with congenital ichthyosis and a history of a posterior uveitis beginning at age 8 presented with an unusual central corneal opacity. At age 26 bilateral central corneal opacities were noted consisting of fine interlacing white lines forming a reticulum at the level of Bowman's membrane. A faint brown background tint of the basal epithelium was also present. Over the next 10 years the opacity progressed only slightly, with some mild superficial scarring of the underlying corneal stroma. The visual acuity remained good. This corneal pattern appeared virtually identical to the superficial reticular degeneration of Koby. As described by Koby in 1927, this rare corneal degeneration is characterised as a painless, slow, progressive development of a central fine, white reticulum at the level of Bowman's membrane. This rare corneal degeneration probably represents an atypical form of band keratopathy.

Koby¹ described a progressive, superficial corneal degeneration in a middle-aged woman which led to a painless loss of vision. This condition was characterised by the appearance of a central fine white reticulum at the level of Bowman's membrane. The epithelium was thickened and bedewed, giving the appearance of a faint opalescence.¹⁻³ This slightly brownish discoloration provided a striking contrast to the white reticulum. There were no signs of inflammation. The loss of vision was progressive but rarely beyond 20/40.

After Koby's original description there was some disagreement concerning the true occurrence of such a case.³⁻⁶ Some authors thought that Koby's case represented lattice dystrophy,⁴ while others held that it was a distinct entity.^{3,5,6} This report presents a patient with the clinical signs of the superficial reticular degeneration of Koby.¹ The authors support the occurrence of this disease as a distinct entity which clinically resembles lattice dystrophy but lacks a positive family history, severe visual loss, and the presence of recurrent erosions.⁷ It is likely that the superficial reticular degeneration of Koby¹ represents an atypical form of band keratopathy.⁸

Case report

An 8-year-old white boy with the diagnosis of chronic vitreitis was referred to one of us (H.G.S.) in June 1950. There was a family history of congenital ichthyosis, but other past medical and family history was negative. The patient's entire body was affected except for both hands and face. At initial presentation a 6-month course of severe ocular inflammation had resulted in opacification of vitreous, which caused substantial visual loss. Fortunately the patient experienced only mild discomfort.

He was admitted to the University of Pennsylvania Hospital in June 1950, and a complete medical survey failed to reveal any other abnormalities. Ocular examination at that time showed that visual acuity was 6/30 in the right eye and 6/60 in the left eye. The intraocular pressure was normal in both eyes. The anterior segment was unremarkable. Both vitreous cavities showed milky-white, stringy opacities. The view of the posterior pole was blurred but seemingly normal. At the extreme periphery of the retina, especially temporally, large, yellow-white exudates extended from the retina into the vitreous. The clinical impression at the time suggested a chronic vitreitis, possibly of pars planitis type. Treatment was begun with adrenocorticotrophic hormone intramuscularly, and the opacities began

Correspondence to Dr Henry D. Perry, Long Island Jewish-Hillside Medical Center, New Hyde Park, New York 11042, USA.

to clear. In July 1950 the patient was referred to the Wilmer Institute, where another exhaustive medical and ophthalmological examination was performed. At that time the ACTH therapy had improved his vision to 6/9 in the right eye and 6/18 in the left eye. The large, yellow-white exudates were still present.

Over the next few years the patient was followed up at intervals never greater than 1 year. The visual acuity remained in the 6/12 to 6/18 range, and the episodes of inflammation became increasingly rare. Since 1964 there has been only 1 episode of inflammation.

In January 1963 a bilateral roughening of the corneal epithelium was noted. This was the first sign of any corneal change. At this time the visual acuity was 6/15 in both eyes, and there were no signs of any other ocular change. By March 1967 the cornea was noted to have a definite central opacity with a mosaic pattern, which was interpreted as being secondary to chronic inflammation and was thought to be a form of band keratopathy. At this time the visual acuity was 6/15 in the right eye and 6/18 in the left eye. By April 1968 the opacity had become more marked and the visual acuity had remained at the same level as the preceding visual acuity, 6/12 in the right eye and 6/15 in the left eye.

Over the next 6 years there was a slow but progressive bilateral increase in the opacities, and in 1975 the patient had well-defined corneal reticular patterns with some evidence of pigmented cells, which at the time were noted on the endothelium. The visual acuity was 6/9 in the right eye and 6/18 in the left eye. A new increase in the uveitis, limited to the left eye, was noted, as was the development of bilateral posterior subcapsular cataracts. In December 1978 at the time of his last examination the visual acuity was 6/12 in both eyes and applanation tension was 17 mmHg in both eyes. External examination showed no evidence of ichthyotic changes of the lids or periorbital. The result of the Schirmer's test with anaesthetic was 12 mm of wetting in each eye; without anaesthetic, wetting measured 16 mm in both eyes. Rose bengal and fluorescein patterns were normal, as was corneal sensation. A bilateral central corneal opacity with a faint brownish colour was noted at the level of the basal epithelium (Figs. 1 and 2). In this central area there was a well-defined, discrete, netlike pattern of interlacing fine white lines at the level of Bowman's membrane (Figs. 3–6). By scleral scatter the fine white lines appeared light brown (Figs. 4 and 6). The underlying superficial stroma appeared slightly scarred. Descemet's membrane was normal, and there was a mild amount of pigment deposited on the endothelium. The lens and retina could easily be seen through the opacity, which was limited to

the central cornea in both eyes. There was no evidence of peripheral band keratopathy. The lens showed early posterior subcapsular cataracts in both eyes. The vitreous showed numerous large opacities in both eyes, and the posterior pole appeared normal. It is our impression that this patient had the superficial reticular degeneration of Koby,¹ which may represent an atypical form of band keratopathy.

Discussion

Information on superficial reticular degeneration of Koby¹ is scarce, especially in recent publications,⁵ which leads to scepticism in terms of the occurrence of this entity as a true corneal dystrophy. Only one pedigree has been reported, a full 20 years after the original description.⁵ The possibility that this patient may have represented a pedigree of lattice dystrophy is likely in view of the finding in several other family members of recurrent erosions, a prominent feature of lattice dystrophy⁷ but one that has not been described in the superficial reticular degeneration of Koby.^{1,3} We are then left with several isolated occurrences, most of which were associated with chronic ocular inflammation. All these factors suggest that we are dealing with an unusual type of corneal degeneration. This is also likely in view of the fact that most patients with this type of corneal change have either been middle-aged or had long periods of chronic ocular inflammation. Furthermore, in his original description Koby¹ used the term degeneration. For this and other reasons we support the designation of this entity as the superficial reticular degeneration of Koby¹ and have used this term throughout this report. As to the specific type of degeneration, it appears clinically to resemble an atypical form of band keratopathy.⁸

The white linear appearance at the level of Bowman's membrane suggests an atypical form of band keratopathy. This is in keeping with the occurrence in some eyes of longstanding inflammation. The opalescence of the brown background tint of the epithelium is more difficult to explain. In cases of recurrent erosions we find irregular iron lines as evidence of chronic tear film disruption. These iron lines impart an irregular yellow-brown tint to the central epithelium. This tint is usually discrete and linear. In this unusual degeneration, we noted an accumulation of pigment at the level of the basal epithelium similar to that seen in Fabry's disease or chloroquine toxicity^{9,10} except that the distribution of the pigment is not whorl-like but reticular and diffuse. This brownish pigment extends just beyond the limits of the reticulum and is

Fig. 1 Right eye. Reticular pattern covering pupillary axis.

Fig. 2 Left eye. Central reticular pattern appearing identical to that of right eye.

Fig. 3 Right eye. Slit-lamp view showing superficial location of reticulum at Bowman's membrane with mild underlying stromal scarring.

Fig. 4 Left eye. High-power scleral scatter showing fine reticular pattern appearing light brown.

Fig. 5 Right eye. Red reflex view of reticulum in central cornea showing limits of corneal involvement.

Fig. 6 Left eye. High-power slit-lamp photograph highlighting white-appearing reticulum with direct light and light brown pattern with indirect light.

unique to this corneal degeneration. We are unable to explain the way in which this pigment distribution occurred or its exact nature.

One of our chief initial speculations was that this particular corneal pattern might represent a manifestation of congenital ichthyosis. Numerous authors have reported cases of various corneal manifestations of congenital ichthyosis. These fall into four main groups:¹¹⁻¹⁴ (1) superficial, round, elevated nodules which have been likened to Salzmann's nodular degeneration and indeed in 1 case were shown to be histopathologically identical to this entity;¹³ (2) forms of typical band keratopathy which are usually secondary to exposure due to the cicatricial ectropion commonly found in patients with the facial manifestations of congenital ichthyosis;¹¹ (3) a superficial, discrete, subepithelial opacity which may or may not be accompanied by a form of typical band keratopathy;¹² and (4) a discrete, deep, pre-Descemet's membrane opacity that is linear and seedlike in appearance, and usually found as an isolated occurrence.¹⁴ This form is more commonly found in sex-linked ichthyosis. No previous article on ichthyosis reports any change similar to that which occurred in our patient. Therefore it seems unlikely that this patient's corneal appearance could be ascribed to congenital ichthyosis.

Clinically the differential diagnosis must include lattice dystrophy, since the pattern of this delicate network of white lines mimics the typical lattice pattern. It is only with careful slit-lamp examination that the lines in superficial reticular dystrophy of Koby¹ can be noted to be limited to the level of Bowman's membrane and lack the birefringence seen in lattice lesions. Of course the lack of family history, severe progressive visual loss, and symp-

toms of recurrent erosions all argue against the diagnosis of lattice dystrophy.

In conclusion, we surmise the superficial reticular degeneration of Koby¹ to represent an atypical band keratopathy.

We thank William C. Nyberg for ocular pathology and S. Flavia Blackwell for editorial assistance.

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