

ptosis, arched eyebrows with lateral thinning of eyebrows, and prominent eyelashes (Figure 1). There were also fetal fingertip pads and abnormally short fifth digits in both hands (Figure 2). The anterior segment examination was normal. The fundus examination showed right prepapillary gliosis, bilateral tortuous retinal vessels, foveal irregular pigmentation, and tilted discs (Figures 3 and 4). The axial lengths of right and left eyes were 24.42 and 24.67 mm, respectively. The updated refraction showed $-2.00 -0.50$ at 100 (right eye) and $-2.50 -1.00$ at 100 (left eye).

Comment

Amblyopia, refractive errors, strabismus, nystagmus, colobomas, microcornea, corneal opacities, blue sclera, cataracts, nasolacrimal duct obstruction, jaw-winking ptosis, caruncle lipoma, cornea pannus, retinal telangiectasia, and retinal pigmentation have all been reported in Kabuki syndrome.⁵⁻⁹ To the best of our knowledge, prepapillary gliosis and tortuous retinal vessels have not been reported (Table 1).

References

- 1 Niikawa N, Matura N, Fukushima Y, Ohsawa T, Kajii T. Kabuki make-up syndrome: a syndrome of mental retardation, unusual facies, large and protruding ears, and postnatal growth deficiency. *J Paediatr* 1981; **99**: 565-569.
- 2 Kuroki Y, Suzuki Y, Chyo H, Hata A, Matsui I. A new malformation syndrome of long palpebral fissures, large ears, depressed nasal tip, and skeletal anomalies associated with postnatal dwarfism and mental retardation. *J Pediatr* 1981; **99**(4): 570-573.
- 3 Niikawa N, Kuroki Y, Kajii T. The dermatoglyphic pattern of the Kabuki make-up syndrome. *Clin Genet* 1982; **21**: 315-320.
- 4 Klujit I, van Dorp DB, Kwee ML, Toutain A, Keppler-Noreuil K, Warburg M *et al*. Kabuki syndrome-report of six cases and review literature with emphasis on ocular features. *Ophthalmol Genet* 2000; **21**: 51-61.
- 5 Turner C, Lachlan K, Amerashinge N, Hodgkins P, Maloney V, Barber J *et al*. Kabuki syndrome: new ocular findings but no evidence of 8p22-p23. 1 duplications in a clinically defined cohort. *Eur J Hum Genet* 2005; **13**(6): 716-720.
- 6 Anandan M, Porter NJ, Nemeth AH, Blair E, Downes SM. Coats-type retinal telangiectasia in case of Kabuki make-up syndrome (Niikawa-Kuroki syndrome). *Ophthalmic Genet* 2005; **26**(4): 181-183.
- 7 Emmert-Buck LT, Preslan MW, Kathuria SS. Jaw-winking ptosis in a patient with Kabuki syndrome. *J Pediatr Ophthalmol Strabismus* 2004; **41**(6): 369-372.
- 8 Evans SL, Kumar N, Rashid MH, Hughes DS. New ocular findings in a case of Kabuki syndrome. *Eye* 2004; **18**(3): 322-324.
- 9 Ming JE, Russell KL, Bason L, McDonald-McGinn DM, Zackai EH. Coloboma and other ophthalmologic anomalies in Kabuki syndrome: distinction from charge association. *Am J Med Genet A* 2003; **123**(3): 249-252.

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Sir,
Corneal epithelial dysmaturation: case report

Corneal epithelial dysmaturation is clinically similar to primary corneal epithelial dysplasia or corneal intraepithelial neoplasia without a prominent limbal lesion. We report a case of successful treatment of an unusual case of unilateral central corneal epithelial dysmaturation.

Case report

A 67-year-old man visited the ophthalmology department for an opaque corneal opacity in his right eye, which had developed 1 year before and had gradually increased in size. Slit-lamp examination showed an individual island of opalescent corneal epithelial lesion, 2×2.4 mm in size, located in the central cornea, without neoplastic fibrovascular corneal pannus (Figure 1). Best-corrected visual acuity was 20/40 in the right eye and 20/20 in the left eye. Other ocular findings were normal. A decision was made to remove the lesion and subject it to cytological examination. The lesion was carefully removed by simple superficial excision with a knife. Cytological examination of the removed lesion showed normal nuclear/cytoplasmic ratio and hyperplastic epithelium with no atypia (Figure 2). Corneal epithelial dysmaturation was diagnosed based upon the clinical and cytological results. Topical antibiotic and steroid were used for 4 weeks with tapering of the steroid. Visual acuity improved to 20/20



Figure 1 Right anterior segment view at the initial examination. It shows an individual island of opalescent corneal epithelial lesion, 2×2.4 mm in size, located at central cornea, without neoplastic fibrovascular corneal pannus.

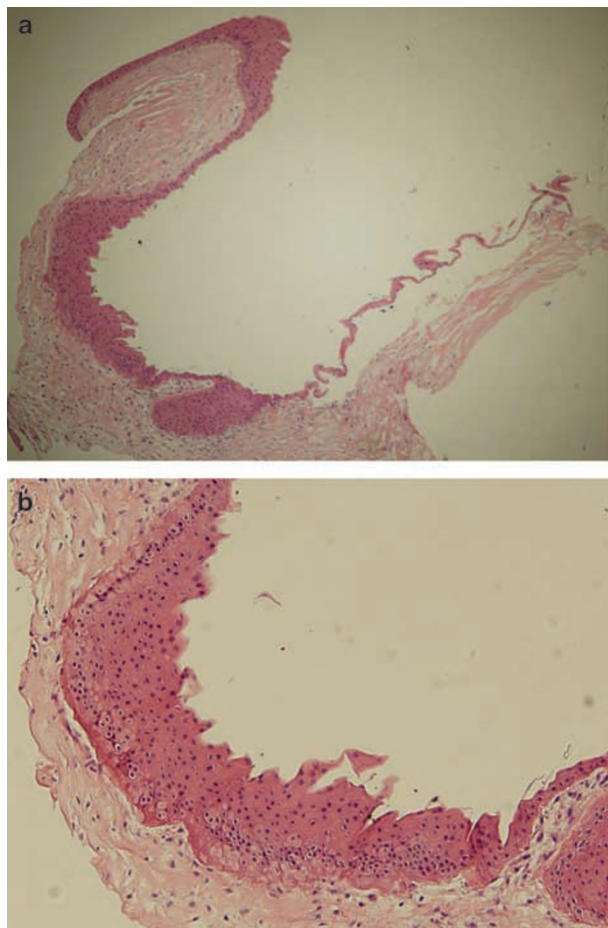


Figure 2 (a) Histopathology of the excised corneal tissue shows increased epithelial cells without surface maturation. There is no inflammatory reaction within the epithelial or stromal layers (haematoxylin–eosin staining; $\times 40$ magnification). (b) Histopathology of the excised corneal tissue reveals a slight loss of polarity of the epithelial cells; however, neither cellular atypia nor dyskeratosis is noted (haematoxylin–eosin staining; $\times 100$ magnification).

in the right eye. After 3 years of treatment, the lesion had completely regressed and did not recur per slit-lamp examination (Figure 3).

Comment

Corneal epithelial dysmaturation tends to exclusively involve the corneal epithelium or involved it in an amount disproportionately large compared with the conjunctiva or limbus.^{1,2} Unilateral central corneal epithelial dysmaturation is an extremely unusual group of conditions, which are characterized by the involvement of corneal epithelium only or a disproportionate involvement of the corneal epithelium from a small limbal lesion that has preferentially spread towards the cornea. Epithelial dysmaturation is a benign and indolent process. Lesions may be unilateral or bilateral, stationary or slowly progressive, and may wax and wane spontaneously. Sometimes the opalescent islands and fingers spread over the corneal epithelium

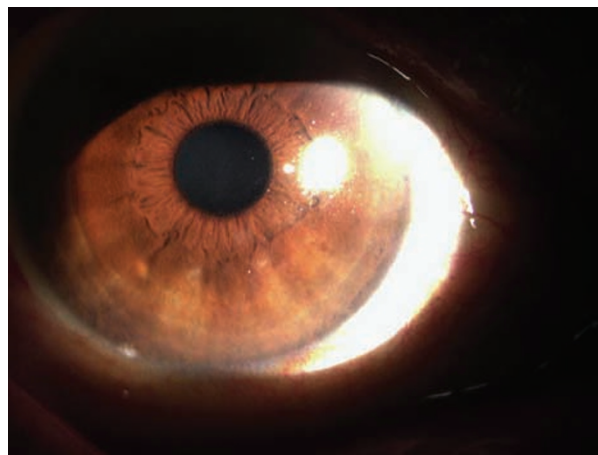


Figure 3 Right anterior segment view at the last visit. At 3 years after treatment, the lesion had completely regressed and showed no sign of recurrence.

and can be mapped geographically over the course of months to years.¹ Fortunately, these lesions are indolent and can be treated with simple corneal scraping and, if needed, wide excision of limbal components as with CIN. We report an unusual case of corneal epithelial dysmaturation. It appears that simple superficial excision may be an effective treatment for corneal epithelial dysmaturation.

References

- 1 Waring GO, Ross AM, Elkins MB. Clinical and pathologic description of 17 cases of corneal intraepithelial neoplasia. *Am J Ophthalmol* 1984; **97**: 547.
- 2 Campbell RJ, Bourne WM. Unilateral central corneal epithelial dysplasia. *Ophthalmology* 1981; **88**: 1231.

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**Sir,
Iris naevus with recurrent spontaneous hyphema simulating an iris melanoma**

Hyphema associated with an iris naevus suggests malignant transformation.^{1,2} Here, we report a case of iris naevus with recurrent spontaneous hyphema simulating an iris melanoma.

Case report

A 58-year-old female patient presented with blurred vision in the right eye because of a recurrent hyphema.