Developmental glaucoma in a patient of ectrodactyly-ectodermal dysplasia

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Key words: Developmental glaucoma, ectrodactyly-ectodermal dysplasia, glaucoma valve, keratoplasty

A 23-year-old Indian female, already on intraocular pressure (IOP) lowering eye drops was referred for control of IOP. Systemic involvement was marked for coarse facial features [Fig. 1a], dental and skeletal abnormalities [Fig. 1c-g] suggestive of ectrodactyly-ectodermal dysplasia (EED). The right eye was in exotropia with no perception of light. The left eye had generalized nebular corneal opacification [Fig. 1b], wide-angle recess with numerous iris processes on gonioscopy, and visual acuity of 20/800. Applanation IOP was 28 mm Hg in the right eye and 46 mm Hg in left eye. Pachymetry and axial lengths were 523 µm and 24.32 mm, respectively in the left eye. Hazy view of fundus revealed glaucomatous optic disc changes.

After lowering IOP with oral acetazolamide, Ahmed glaucoma valve FP7 (New World Medical, Inc CA) was implanted in superotemporal quadrant. The tube was placed in in the anterior chamber. At 4 months, full-thickness penetrating keratoplasty (PK) was done. In the early postoperative period vision was 20/1000 and IOP varied between 6 to 9 mm Hg [Fig. 2a]. At 7 months follow-up after PK, best-corrected visual acuity (BCVA) was 20/80 [Fig. 2b]. The corneal graft was clear but cataractous changes were noticed [Fig. 2c]. The IOP was 16 mm Hg, and the tube was well placed in the anterior chamber [Fig. 2d] with diffuse functional bleb [Fig. 2e]. The left eye fundus examination revealed average-sized disc with vertical cup-to-disc ratio of 0.75 and inferior rim thinning [Fig. 2f]. At 18-month follow-up, graft was clear, BCVA was 20/50, and IOP was 16 mmHg.

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Discussion

The reported ocular involvement in EED includes telecanthus, blepharophimosis, entropion, meibomian gland absence, atresia of lacrimal outflow tract components, ocular surface and tear film disorders, and keratopathy.[1,2] Infantile glaucoma has been reported with ectodermal dysplasia.[3] The absence of iris anomalies and presence of ectrodactyly differentiate this case from Axenfeld-Rieger syndrome.[4]

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Nil.

Conflicts of interest

There are no conflicts of interest.

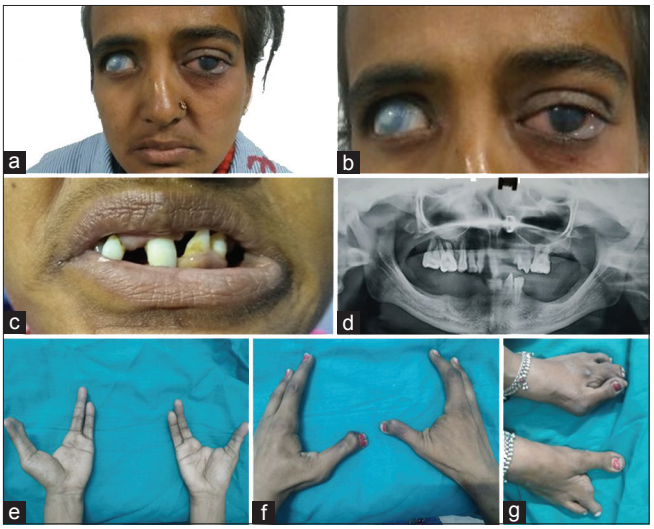

Figure 1: (a) Coarse facial features. (b) Dense corneal opacity in the right eye and the left eye with hazy cornea. (c) Oligodontia. (d) Orthopantomograph showing oligodontia with underdeveloped alveolar bone. (e and f) Ectrodactyly of hands. (g) Ectrodactyly and syndactyly of feet

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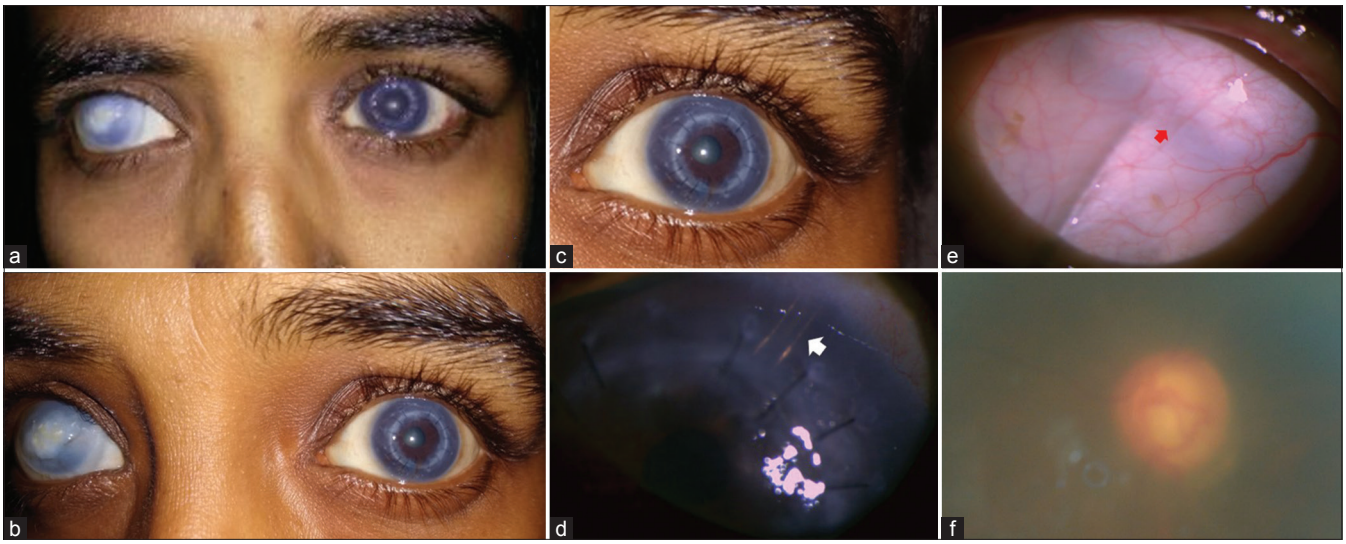

Figure 2: (a) Some edema of the graft 1-week post-keratoplasty (b) Clear graft at 7-month follow-up (c) Clinically significant cataract. (d) Ahmed glaucoma valve FP7 tube *in situ* (white arrow) in the anterior chamber. (e) Bleb (red arrow). (f) Fundus photo depicting glaucomatous cupping in the left eye

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