Sclerocornea

A term newborn delivered to a second gravida mother vaginally was noted to have an opacity in the right eye soon after birth. Eye examination revealed normal left eye and sclerisation of entire cornea on right side (Fig. 1). The thickness of opacity was more at periphery than at the center. Corneal diameter was 10 mm. The lid and adnexa were normal. Details of anterior chamber, iris and lens could not be made out. Margin of the pupil was hazy. The intraocular pressure was normal. There was no facial asymmetry. Systemic examination was normal. There was no family history of a similar problem. A diagnosis of total sclerocornea was made.

Sclerocornea, an uncommon developmental abnormality of anterior segment due to mesenchymal dysgenesis presents as a stationary congenital anomaly. It is usually seen as an isolated ocular abnormality involving both eyes. Occurs sporadically but may be familial or autosomal dominant in inheritance. Clinically, most often there is peripheral, white, vascularised 1-2 mm corneal rim that blends with sclera obliterating the limbus. The central cornea is generally normal. In total sclerocornea, the entire cornea is involved but the center of the cornea is clearer than periphery, which distinguish it from. Peters’ anomaly where opacity is more at center. The opacification affects the full thickness stroma that limits the visualisation of posterior corneal surface and intraocular structures. Histology includes disorganised collagenous tissue containing fibrils that is larger than normal. Potentially co-existing abnormalities include shallow anterior chamber, iris abnormalities and microphthalmos. Systemic abnormalities like limb deformities, craniofacial and genitourinary defects can also accompany. In generalised sclerocornea early keratoplasty should be considered in an effort to provide vision.

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