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Grade III Corneal Dermoid: Good Aesthetic Results After Surgery in Childhood

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Abstract

We report of an initially 4-year-old boy with grade III corneal dermoid. Enucleation, a surgical challenge, was performed and histopathologically the diagnosis epibulbar dermoid was given. This case demonstrates that a satisfactory aesthetic result can be achieved by careful planning though preconditions were complex facing this rare entity.

Case Report

The mother reported of a thin cornea that impressed at birth. A forthcoming scar formation and deformation of the cornea was early observed followed by staphyloma formation resulting in an overall axial length of about 35 millimeters.

The initial clinical examination revealed a visual acuity of 12/20 at his right eye, and no light perception at the left eye. The slit lamp and fundus examination of the right eye presented a regular result. On the left eye a white, cherry-sized, opaque protrusion was seen (**Figure 1A**). There were no intraocular details visible, but ultrasound revealed no intraocular pathology. MRI (done elsewhere before) confirmed the axial enlargement of the eye with a small lens. Furthermore, a hydrocephalus was known.

Therapeutic options were discussed in detail with the parents and eventually the left eye was enucleated after careful dissection revealing limited access to Tenon's space and the eye muscles (**Figure 1B**). A 20 mm alloplastic implant was muscle fixated inserted. A temporary glass conformer was applied which was replaced by a regular prosthesis after a few weeks.

32 months after surgery the parents reported on no complaints and a good aesthetic result (**Figure 1C**).

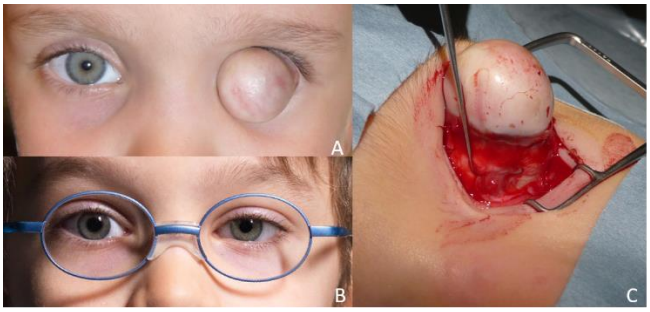

Figure 1: (A) Initial presentation: left eye with white opaque cherry-sized protrusion. **(B)** Final result with prosthesis. **(C)** Intraoperative situs: Conjunctiva opened; medial rectus muscle on squint hook.

The histological examination presented a 32 x19 x 20 mm sized eyeball (**Figure 2A**). A white deformation instead of the cornea and a tiny lens were seen macroscopically.

Microscopically a dense cornea with vascularization impressed. The epithelium was keratinized. Bowman's layer- and Descemet's membrane were absent except some Descemet's membrane remnants in the very periphery near

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by the chamber angle rudiments. There was pigment epithelium at the back of the cornea as well as in the anterior chamber where some pigment epithelium with a basal membrane between the epithelial layers was observed. There was a rudimentary iris visible (**Figure 2B**). The retinal ganglion cell layer and internal limiting membrane were thickened; the ganglion cell layer was also degenerated.

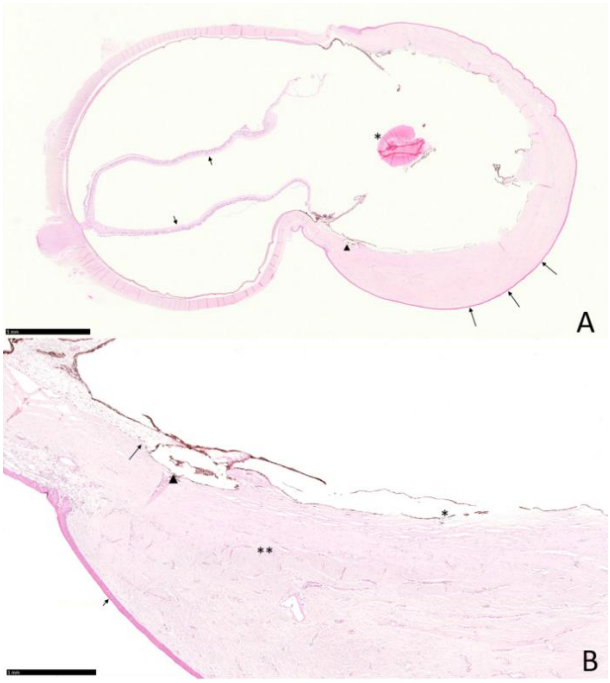

Figure 2: (A) The enucleated eye presenting with diffuse corneal thickening (long arrows) and a small malformed lens (*). Artificial retinal detachment (short arrows). Malformed posterior cornea lacking Descemet's membrane (triangle) (Hematoxylin-Eosin, magnification bar = 5 mm). **(B)** Magnification of the anterior segment: corneal dermoid consisting of dense, vascularized connective tissue covered by epidermis (short arrow), unsharp transition into corneal stroma (**). Aplasia of the iris, iridocorneal synechiae (long arrow). Pigment epithelium next to the inner corneal surface, missing Descemet's membrane (*). Triangle indicating identical site as in **figure 2A** (Hematoxylin-Eosin, magnification bar = 1 mm).

Discussion

Epibulbar dermoids are choristomas that are defined as overgrowth of normal tissues in an abnormal location. The incidence of epibulbar choristomas is 1-3 per 10.000. Choristomas are the most common epibulbar and orbital tumors in children [1].

Epibulbar dermoids can affect the conjunctiva, limbus and cornea [2]. They are solid tumors of collagenous connective tissue, covered by squamous epithelium. They may contain hair follicles, gland tissue, adipose tissue and nerve tissue that were lacking in our case [1]. They are associated with Goldenhar-syndrome, Schimmelpenning-Feuerstein-Mims phakomatosis, epidermal nevus syndrome and oculoauriculovertebral dysplasia, but can also present isolated [3]. Autosomal dominant, recessive, X-chromosomal and multifactorial inheritance patterns are described. They can occur uni- and bilateral as well as a single lesion or multiple lesions [2,4]. In addition, there was a severe anterior segment anomaly with lacking Descemet's membrane, iridocorneal synechiae and lens anomaly which reminds on Peters anomaly.

Epibulbar dermoids are thought to arise from an early developmental anomaly resulting in metaplastic transformation of the mesoblast between the rim of the optic nerve and surface ectoderm [1].

In 1937, Ida Mann introduced the current three-grade classification of corneal dermoids depending on the depth of involvement [5]:

Grade I dermoids are superficial, typically located at the limbus and measuring less than 5 mm. Grade II dermoids involve deeper structures up to the stroma though the Descemet membrane is intact. Grade III dermoids are the rarest type. They cover the whole cornea and can involve the anterior segment up to the pigment epithelium of the iris. Grade III dermoids are associated with microphthalmos and posterior segment anomalies [1,3].

In our case, we present a grade III dermoid with staphyloma. There is no sign of microphthalmos, but posterior segment anomalies exist. In addition, severe anterior segment malformation reminding on Peters anomaly were present.

Even though one case of spontaneous partial regression is described, [6] surgery typically is the therapy of choice. Surgical options for a grade III corneal dermoid include anterior segment reconstruction, corneal transplantation, Keratoprothesis in case of staphyloma formation, enucleation or evisceration [2] and excision followed by scleral graft patching [4].

In our case, the fellow eye had good visual acuity, but the affected eye had no light perception. Since a resulting orienting vision of this eye after corneal transplantation or keratoprothesis was not expectable, our primary concern was the cosmetic improvement. At the age of four, the facial bones are almost mature so that facial asymmetry is no major concern anymore [7]. In consequence, we favored enucleation. The application of a ball-shaped alloplastic implant ensures an adequate basement for adapting a prothesis and preserves partial motility.

A proper preoperative planning takes numerous parameters like age of the patient, growth of the facial bones, size of the bulb, staphyloma formation and visual acuity into consideration. Thus, a carefully chosen strategy improves the outcome regarding a pleasing cosmetic result.

References

1. Mansour AM, Barber JC, Reinecke RD, Wang FM (1989) Ocular choristomas. *Surv Ophthalmol* 33:339-358.
2. Pirouzian A (2013) Management of pediatric corneal limbal dermoids. *Clin Ophthalmol* 7:607-614.
3. Stergiopoulos P, Link B, Naumann GO, Seitz B (2009) Solid corneal dermoids and subconjunctival lipodermoids: impact of differentiated surgical therapy on the functional long-term outcome. *Cornea* 28:644-651.
4. Şimşek A, Bilak Ş, Balyen L, Erdoğdu İH (2015) Conservative Surgical Management of a Grade III Corneal Dermoid. *Cornea* 34:1318-1320.
5. Mann I (1957) Developmental Abnormalities of the Eye. 2d ed. Philadelphia: Lippincott 357-364.
6. Rončević MB, Dorešić JP, Md LD (2011) Large congenital corneal dermoid with spontaneous partial regression: the first report. *Cornea* 30:219-221.
7. Hintschich C, Zonneveld F, Baldeschi L, Bunce C, Koornneef L (2001) Bony Orbital Development After Early Enucleation in Humans. *Br J Ophthalmol* 85:205-208.