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Original Research Paper

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CONGENITAL UPPER EYELID COLOBOMA:2 CASES

Dr Angshupriya Bhattacharjee* MS Ophthalmology *Corresponding Author

Dr Saurabh Gupta Mch, Plastic Surgery

ABSTRACT The goal was to describe our experience in the surgical management and treatment of 2 patients with congenital upper eyelid colobomas.

KEYWORDS :

INTRODUCTION

Congenital eyelid coloboma is an uncommon, unilateral or bilateral, partial or full-thickness eyelid defect. It is caused by failure of fusion of the mesodermal lid folds . It may be isolated or associated with other ocular or systemic anomalies. Immediate attention at an early age through corneal protection, surgical repair of the eyelid defect, and monitoring of the visual development are essential to prevent complications: corneal leukoma, symblepharon,and amblyopia .

This report summarizes our experience in the surgical management and treatment of two patients with congenital eyelid colobomas.

METHODS

A detailed clinical history was collected, including information about personal and family history, exposure to drugs or diseases during pregnancy, and a complete ophthalmic examination at the time of referral, at subsequent check-ups, and in the postoperative period.

An initial examination was performed to assess the visual acuity (VA), site, and size of the eyelid defect and ocular motility as well as to determine the presence or absence of ocular anomalies (by biomicroscopic examination, examination of the ocular fundus, and ocular ultrasonography) .

RESULTS

This study included 2 patients with eyelid coloboma not associated with other ocular and systemic pathologies .

The mothers of the patients had no history of infections or other illnesses during pregnancy and there was no exposure to drugs or medicines.

Case Reports

Case 1. A 17-day-old male infant was referred to us for congenital upper eyelid coloboma of medial location involving more than two-thirds of the eyelid margin. The patient had telecanthus, involving more than 80% of the corneal surface in both eyes.

Ultrasonography of both eyes was normal and reconstruction of OU using Tenzel's semicircular flap and lateral canthotomy and cantholysis were performed.

Currently, aafter1 years of surgery, the patient has 0.5 mm of lid lag

Fig1 case1.preoperative and post operative picture of 17 days infant

Case 2. A 2 month-old female patient was referred due to congenital
upper eyelid coloboma affecting the medial one-half of the upper eyelid. She had undergone surgery sliding taraocunjunctival flap with lateral cantholysis.

DISCUSSION

There are several theories that attempt to explain the cause of congenital upper eyelid colobomas. Tessier considers coloboma of the eyelid to be a form of facial cleft .

Other authors suggest that these colobomas are associated with intrauterine factors such as amniotic bands, abnormal fetoplacental circulation, or radiation . They are usually unilateral, generally located at the medial one-third of the upper eyelids (90%), and may vary from a small notch to complete defects of the eyelid. Lower eyelid colobomas usually affect the outer one-third of the eyelid .

Colobomas are also associated with facial clefts, Goldenhar syndrome, Treacher Collins syndrome, Charge syndrome, or frontonasal dysplasia .

They may present other ocular and orbital anomalies such as conjunctival or limbal dermoid tumors, conjunctival chondroma, symblepharon, corneal opacities, macular or optic nerve colobomas, and strabismus.

Eyelid reconstruction at the right time is essential in these patients. This will depend on the size of the defect and on the presence of corneal exposure.

If the defect is small and there is no corneal exposure, surgery could be delayed until the age of 3-4, when there is an increased amount of eyelid tissue. Otherwise, surgery should be done as soon as possible to avoid corneal lesions .

The surgical technique will depend on the size of the defect. Defects up to 25% can be closed directly. Defects between 25%

and 50% can be repaired by direct closure with canthotomy and cantholysis or by Tenzel's semicircular flap.

In case of defects greater than 50% of the eyelid, functional and cosmetic results will be difficult to achieve. In these cases, several techniques could be used such as Tenzel's semicircular flap, Cutler-Beard technique, or Mustarde rotational flap.

Other techniques have been described such as the lamellar-based technique, proposed in patients with a discordant defect between the anterior and the posterior lamellae of the eyelid . Others such as tarsomarginal grafts allow the defects involving more than 50% of the lid margin to be closed and all eyelid structures to be replaced .

Conclusions

Congenital upper eyelid coloboma is an uncommon defect of unknown etiology, which might be associated with other ocular and systemic pathologies, and therefore it requires a multidisciplinary approach in a large number of cases.

Eyelid colobomas are a potential threat to vision at an early age, requiring close monitoring of the visual development of patients.

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