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Comprehensive Study of Management of Upper Lid Coloboma

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Abstract: ***22 years old girl having isolated congenital upper eyelid coloboma was treated with tenzel flap and closing coloboma defect after freshening of margins of the defect. Eyelid coloboma is a rare congenital condition where there is an absence of the development of eyelid tissue. Eyelid coloboma, in addition to being a cosmetic disfigurement, affects the cornea and vision and, if associated with other systemic abnormalities, can cause severe morbidity. Usually, the upper eyelid is commonly affected, and the most common site is the junction between the medial and middle third of the upper eyelid. This article reviews the evaluation and treatment of eyelid coloboma.***

Keywords: Eyelid Coloboma, Congenital Defect, Tenzel Flap, Ophthalmic Reconstruction, Corneal Protection

1. Introduction

Coloboma of the eyelid has a different embryological origin from a coloboma of the globe. Eyelid colobomas are typically located at the medial part of the upper eyelid. Lid coloboma arises from the defective eyelid development; either during fusion, occurring during the third and fourth months of embryological development, or during re-separation, which occurs in the sixth or seventh months.[1,2] Although the sequence of eyelid development itself is induced by the developing globe, there is no clear evidence that eyelid coloboma results from globe abnormalities[.3].

Environmental or mechanical events during pregnancy with or without a genetic predisposition may contribute to the development of an eyelid coloboma. Isolated eyelid colobomaare mostly sporadic, with cases having de novo mutations. Moreover, cases of isolated eyelid coloboma have been described in association with radiation exposure.[4]

Isolated lidcoloboma can be graded based on the severity of the CorneoPalpabral Adhesions (CPA), as follows [4]:

Grade 1 - Coloboma without cryptophthalmos
Grade 2 - Coloboma with abortive cryptophthalmos
Grade 3 - Coloboma with complete cryptophthalmos
Grade 4 - Classic cryptophthalmos (absence of all eyelid structures and complete coverage of eye by skin)
Grade 5 - Severe cryptophthalmos (with severe deformity of the nose and ectropion of the upper lip)

2. Case Report

22 year old female patient presented to eye OPD with eyelid defect in Right upper eyelid since birth. She was full term normal delivery and post-natal period was uneventful.

There was 11mm horizontal x3mm vertical eye lid defect in upper eyelid at middle and medial one third of eyelid. Bells phenomenon was good. Upper eyelid central horizontal full thickness defect sizing 40% of lid involving lid margin, not involving medial or lateral canthus.

Elasticity of lid was good. Levator function was normal. Her right eye vision was 6/6 with -1.0DS. and 6/6 unaided in left eye. She had clear cornea, and no sign of exposure keratitis. Other anterior and posterior segment examination was normal.

Her general and systemic examination was normal As per CPA severity grading, she had Grade 1.

She came for cosmetic correction of eyelid defect and was operated for the same.

Patient was operated under general anesthesia. Coloboma edges were freshened up.

Incision was taken 10 mm lateral to lateral canthus in skin and subcutaneous tissue and dissection was done to mobilize tenzel flap[5]. Lateral cantholysis was done. Lid defect was approximated. Gray line and posterior lid margin sutured with 6/0 Vicryl sutures. Vertical cut edges of tarsal plate was sutured in partial thickness with 6/0 Vicryl sutures. Orbicularis oculi muscle and skin sutured with 6/0 Vicryl. Canthopexy done with 6/0 Vicryl suture, Subcut 6/0 Vicryl, Skin 6/0 silk suture.
Post operative defect was well corrected with acceptable cosmetic results.

3. Images

Figure 1: Clinical photograph of the patient showing 11mm horizontal x 3mm vertical eye lid defect in upper eye lid at middle and medial one third of right superior eye lid

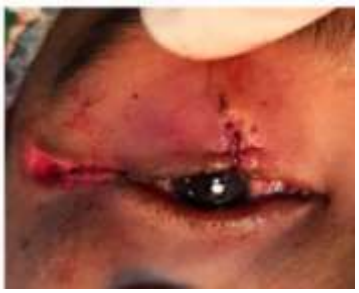

Figure 2: Intra op. photograph showing Tenzal flap and closure of lid defect

Figure 3: Post operative photograph

4. Discussion

Goals of lid reconstruction are to restore physiological function of lids (Vision, Lid closure, Mobility,Tear drainage),to Re-establish anatomic integrity and provide best cosmetic appearance.

Eyelid coloboma are mostly central lid defects, that cannot be closed directly. Superiorly/inferiority based semicircular flap has to be transposed from the lateral canthalarea which is called Tenzel flap technique.[5]

Defect is repaired with meticulous layer by layer closure.There should be no overlapping of wound edges and ensure eversion of wound edges at margin to prevent notching.

Advantages of this technique are it is single stage procedure and gives excellent lid stability.

5. Conclusion

Large eye lid coloboma can be repaired with good functional and cosmetic outcome with Tenzel flap and meticulous layer wise closure in a single stage procedure.

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