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Early Reconstruction Surgery Resolving Visual Occlusion and Ocular Malformation

A Case Report

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Abstract: Congenital ocular malformation may lead to failure of the development of ocular regions and visual function in the pediatric population. Orbital bones are established within the first 2 months of embryogenesis. Any abnormalities may lead to failure in development of the ocular region.

In this case report, we present a newborn with congenital canthus malposition, a distorted tarsus, and mild enophthalmos caused by an embryogenesis fibroma, which although rare, can result in amblyopia.

Surgical resection of the fibroma followed by orbital reconstruction of the canthus, correction of malposition and tarsus extension were performed. Twelve months and 5 years follow-up showed no recurrence and the visual acuity was 20/20 in the left eye and 16/20 in the right. The deformity of the left orbit was corrected without complications and was symmetrical with the right side.

This is the first known case of children with embryonic fibroma. It is likely that early reconstruction surgery may avoid visual occlusion and ocular malformation.

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Abbreviations: CT = computed tomography, ICD = inner canthal distance, OD = oculus dexter, OS = oculus sinister.

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INTRODUCTION

The growth of the periocular region is most rapid in the first 12 to 24 months of life.[1] Any abnormalities may lead to failure in development of the ocular region. More importantly, amblyopia, which can be caused by ocular malformation, appears stable by 3 years of age.[2] Interventions should be performed to avoid irreversible cosmetic and functional deformity. Since the pediatric orbit can be highly resilient, management of the periocular region requires an understanding of the growing face and sensitivity to the long-term impact of intervention.[3] Fibroma as a benign neoplasm has never previously been described to occur in an infant, and result in severe congenital ocular malformation and a high risk of visual

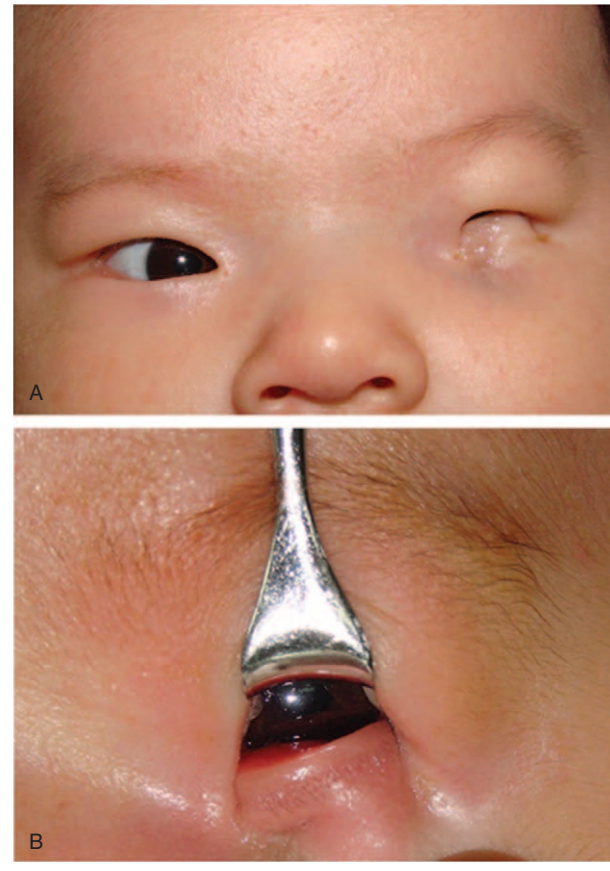

FIGURE 1. Ophthalmic examination showing a small left palpebral fissure in the lower orbit (A) and the cornea clear and of normal size (B).

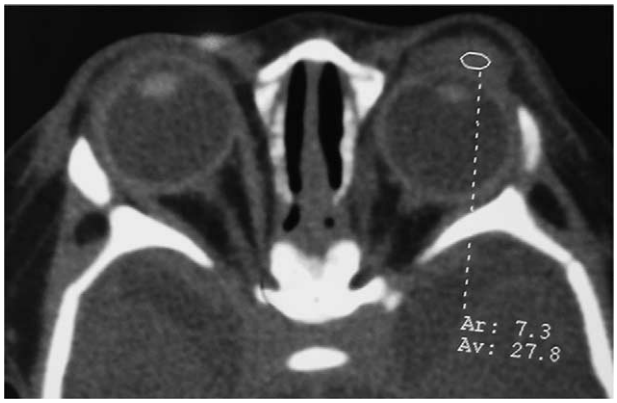

FIGURE 2. Axial CT showing a 10 mm × 7 mm neoplasm causing enophthalmos of the left eyeball.

dysfunction. We report a newborn who underwent surgical resection of an embryonic fibroma followed by successful orbital reconstruction for her congenital ocular malformation and visual occlusion.

CASE REPORT

An otherwise healthy newborn presented to the ophthalmology clinic for evaluation of a small left palpebral fissure and a palpable mass adherent to the rim of the inferior orbit since birth (Figure 1A).

On ophthalmic examination, the horizontal distance between the commissures was 22 mm in the right eye and 13 mm in the left, whereas the vertical distances between the palpebral fissures measured 7 mm in the right eye and 1 mm in the left. The inner canthal distance (ICD) was 32 mm. A 10 mm × 5 mm palpable, firm, pulseless mass was adherent to the inferior orbital rim. The palpebral fissure was so small that the eyeball could hardly be seen; thus, the left cornea was checked with the retractor and was found to be transparent and of normal size (Figure 1B). Both a direct and consensual light response could be seen with each pupil. However, the patient was too young to cooperate with visual acuity and eye movement exams. The review of the systems was unremarkable. Computed tomography (CT) scan revealed an oval-shaped, well-defined, medium density 10 mm × 7 mm mass adherent to the inferior orbit, causing compression, and enophthalmos of the left eyeball (Figure 2).

After extensive discussion of the risks and benefits of surgery, such as bleeding, infection, scar, recurrence, asymmetrical development of the periocular region, and visual occlusion, the parents of the patient agreed to proceed with surgical excision and orbital reconstruction at same time under general anesthesia. Approval was obtained from the institute's ethics committee, and the study was conducted in accordance with the guidelines of the Declaration of Helsinki and patient consent was obtained. A subcillary incision was made, followed by dissection between the mass and surrounding normal tissue to completely expose the lesion. A grey-white, well-demarcated area of fibrous tissue adhering to the tarsus and the inferior orbital rim was resected. The compression of the inferior orbital rim was released and the distorted lower tarsus was flattened (Figure 3A and B). A Bowman probe was passed through the canaliculus until a soft stop was felt at the nose, to ensure the lacrimal system was intact. A Z-plasty skin flap was used to correct the medial canthus malposition (Figure 3C). Since it was found to be oblique, the medial canthus tendon was then anchored horizontally to the medial orbital mucoperiosteum; therefore, making it symmetrical to the other side. A lateral canthopexy was also performed to extend the palpebral fissure. A 2 mm horizontal incision was made from the lateral canthus,

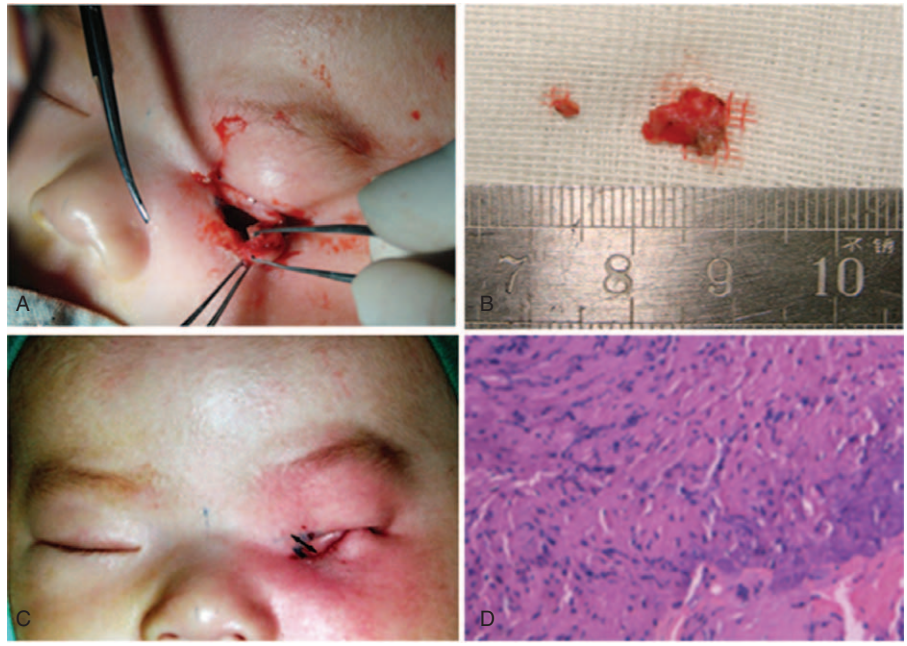

FIGURE 3. Intraoperatively, a grey-white, well-demarcated fibrous tissue adherent to the tarsus, and inferior orbital rim was resected (A and B). A Z-plasty flap was made to correct the medial canthus malposition and lateral canthopexy was performed (C). Histopathologic findings of a tarsus neoplasm, with dense fibroblasts and bundles of collagen spindles (H & E staining, 400×) (D).

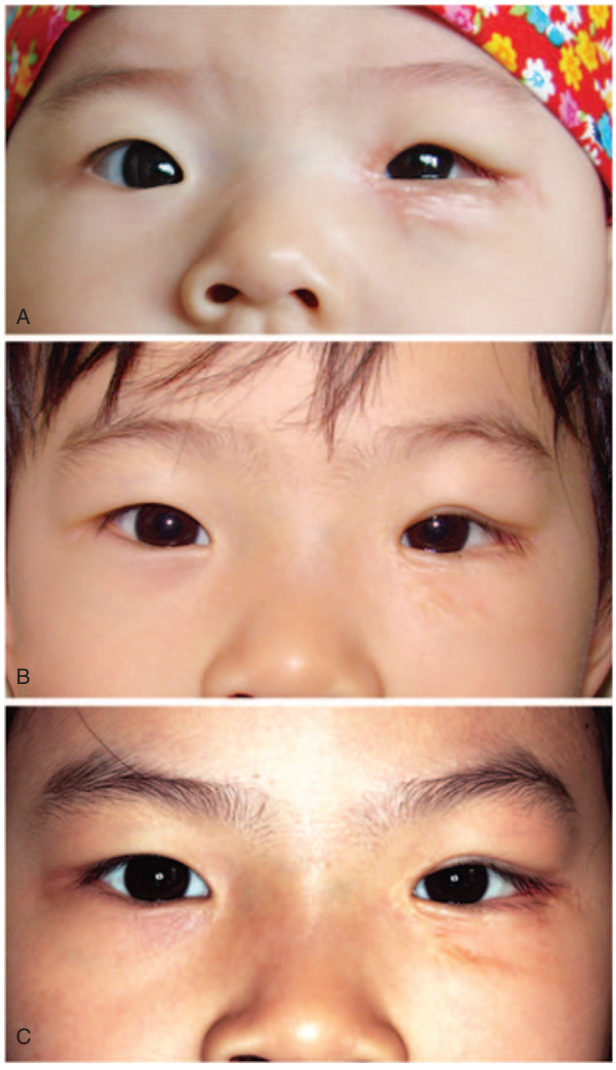

FIGURE 4. Cosmetic outcomes 12 months (A), 2 years (B), and 5 years (C) postoperatively with intact visual acuity.

followed by anchoring the conjunctival flap to the lateral canthal ligament. The skin was closed with 5-0 silk sutures. On pathology, fibroblastic proliferation with infiltration of collagen spindle bundles and partial mucinous degeneration (hematoxylin and eosin, original magnification 400×, Figure 3D) was seen, confirming the diagnosis of orbital fibroma.

The patient was followed up at 12 months and 5 years. The palpebral fissure measured 22 mm oculus dexter (OD) and 21 mm oculus sinister (OS) in width, whereas the length was 8 mm OD and 6 mm OS at 12 months (Figure 4A). At the 5-year follow-up point, there was no recurrence and the visual acuity was 20/20 OD and 16/20 OS. The deformity of the left orbit was corrected without complications and was symmetrical with the right side (Figure 4B and C).

DISCUSSION

In this case report, we described for the first time a newborn diagnosed with a left periocular deformity secondary to a fibroma, treated successfully with surgical resection and orbital reconstruction. To our knowledge, congenital orbital deformity due to a growing fibroma during the embryonic stage has not yet been reported.

The anatomy and development of the orbit and periocular region is complex. According to Berger and Kahn[4] the orbital bones are established within the first 2 months of embryogenesis, while eyelid differentiation begins at week 6 of embryogenesis.[5] Congenital deformity of the periocular region can result from any abnormality of the orbit or surrounding tissue during the process of embryonic development.

Fibroma is a rare, benign neoplasm generated from the connective tissue of any organ and characterized as a well-demarcated, fibrous appearing, grey-white, non-tender mass. Microscopically, fibroblasts widely separated by abundant broad bundles of collagen with positive vimentin staining are seen. According to our literature review, it has only been diagnosed pathologically in 7 middle-aged and older adults, in the periorbital region; since the orbits of these adults were completely developed, no deformity was found.[6–12]

Therefore, it is paramount to approach the evaluation with a thorough differential diagnosis that includes infantile hemangioma,[13] dermoid cyst, cellulitis, lymphangioma, and neuroblastoma. In this case, the clinical features and radiologic examinations enabled us to rule out other etiologies; the diagnosis of fibroma was confirmed by biopsy and on pathology. The orbit completes half of its postnatal development by age 2 years; it is not until age 7 that adult dimensions are attained.[4] In general, the fibroma continues to grow postnatally, throughout the development of the orbit and differentiation of the lower tarsus, therefore leading to a disability in tarsus extension and malposition of the canthus. More importantly, deformity of the periocular region may result in amblyopia, which is the leading cause of early childhood unilateral reduced vision secondary to deprivation.[14] Therefore, fibroma excision and reconstruction of the periorbital region appears to be of necessity, not only to improve patient appearance but also to relieve the delay in ocular development, canthus malformation, and visual axis occlusion.

CONCLUSION

This case report adds fibroma to the differential diagnosis of congenital periocular malformation, which also represents a clinical problem never before reported in the literature. Children are likely to benefit from early surgical intervention, with fibroma resection, and orbital reconstruction, to avoid deprivation amblyopia and asymmetric periocular development.

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