WESTERN NIGERIA JOURNAL OF MEDICAL SCIENCES (WNJMS)

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PUBLISHED BY: UNIOSUN TEACHING HOSPITAL, OSOGBO, NIGERIA

PUBLICATION DATE: January-June, 2024

https://www.wnjms.com.ng

VOL. 7 (ISSUE 1) 2024

Western Nigeria Journal of Medical Sciences
Publication Date: June 30, 2024; Vol. 7, Issue 1

CASE REPORT

CASE REPORT OF CONGENITAL BILATERAL ECTROPION OF THE UPPER EYELIDS IN A 6 YEAR OLD BOY WITH DOWN SYNDROME

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ABSTRACT

Introduction: Congenital ectropion is a rare but usually benign condition in which the upper eyelid is turned outward. It may be associated with Down syndrome, congenital lid disorders such as blepharophimosis syndrome, congenital ichthyosis and birth trauma. We report this case to call attention to, and improve the understanding of various paediatric caregivers about this rare condition, and to encourage prompt referral to the specialists when it is diagnosed.

Case Report: We report a case of congenital bilateral ectropion in a 6 year old boy with Down Syndrome. His condition was noticed at birth and he was admitted for two weeks from birth because of the eyes; but was discharged home despite minimal improvement. His mother brought him to our facility because the problem persisted and the child also has tearing and recurrent purulent eye discharge. He was placed on topical antibiotics; the anxious mother was reassured and child was referred for surgery.

Conclusion: The condition is largely benign though complications may result from delayed and improper treatment.

Keywords: Congenital ectropion; Down syndrome; eversion; eyelids.

INTRODUCTION

Congenital eyelid ectropion is an uncommon disorder characterized by a complete out-turning of the eyelids in infants, which is generally associated with conjunctival prolapse and chemosis. Congenital eversión or ectropion of the upper eyelids is uncommon. The eversion of the lid margin is usually bilateral, though may be unilateral.[1] This condition was first described as "double congenital ectropion" in 1896 by Adams and about 100 cases have since been reported.[2,3] Congenital ectropion is twice as common in males, and there is a racial preference for black infants.[4-6]

It can occur alone or in conjunction with other conditions such as infections, inflammation, birth canal trauma, or systemic diseases such as lamellar ichthyosis, collodion skin disease, and Down syndrome.[6-10] Goncalves *et al*. reported congenital ectropion of the eyelids in three Down syndrome patients.[11] The association with Down syndrome is thought to be due to faulty development of the lateral canthal ligament.[3,8,11]

According to Krishnappa *et al*, affected Down syndrome patients typically have a more complicated course. [8] This is likely due to eversion of the lid from vertical shortening of the anterior lamella or the vertical elongation of the posterior lamella with failure of the

Date Submitted: 08 December, 2023

Date Accepted: 18 April, 2024

orbital septum to fuse with the levator aponeurosis, in addition to the eyelid laxity.[8] Congenital ectroprion may lead to overflow of tears (epiphora) and subsequent maceration of the skin of the lid, inflammation of the exposed conjunctiva, or superficial exposure keratopathy.[7]

The condition is largely benign though complication may result from delayed and improper treatment.[2] Congenital ectropion though rare, may also resolve completely within a few weeks of life if well treated.[2,3,5,6] Unfavorable outcome can arise from failure to treat this condition at birth.[3] The management may be conservative using topical lubrication, antibiotics, and hypertonic saline or surgical correction.[6,12,13] Protection of the cornea is essential since complications such as corneal abrasions, impaired vision and permanent blindness can arise in poorly treated cases.[4] We report this case to call attention to, and improve the understanding of various paediatric caregivers about this rare condition, and to encourage prompt referral to the specialists when it is diagnosed.

CASE REPORT

The Patient was a 6-year old boy who was brought to the eye clinic of University of Medical Sciences Teaching Hospital, Akure, Nigeria by the mother. The complaint was inability to close the eyes, tearing and purulent eye discharge noticed since birth. He was delivered by spontaneous vaginal delivery (SVD) in a hospital and was on admission for two weeks after birth because of the eyes, though there was minimal improvement at discharge. The mother admitted to being distressed and frustrated as she had to constantly assist the child to close the eyes whenever he falls asleep and has had to buy several over the counter medications to treat the recurrent eye discharges and tearing which have persisted. The child has also mastered the act of manipulating the eye lids to reverse the eversion whenever it occurs when he is awake. He was hypotonic at infancy and gross development was delayed.

On examination, he was conscious and alert, not pale, was anicteric and had a normal temperature. He had low-set ears with single palmar crease, flat occiput and nasal bridge. He had short sturdy fingers and a short neck. He was playful and had friendly disposition. He was microcephalic and short for age. He had a normal weight and the heart sounds were normal. The visual acuity was 3/36 in both eyes using Snellen chart at 3 metres. He had upper eyelids ectropion on closing the eyes with lateral lower eye laxity and ectropion of the lateral third of the upper eyelids. There was epiphora and purulent eye discharges. Both corneas and lenses were clear, the anterior chambers were normal and pupils were reactive to light. Figures 1-3 show the different appearances of the eyelids.

Picture 1: Patient opening his upper eyelids with eversion of the upper lids

Picture 2: Patient closing his upper eyelids with eversion of the upper lids

Picture 3: Temporary resolution after manual reduction by the child.

A diagnosis of bilateral ectropion of the upper lids in a patient with Down syndrome was made. The mother was then counseled on the child's diagnosis and modalities of management. He was initially managed conservatively with the use of topical antibiotics, ointment and lubricants while being followed up in the out-patient eye clinic. There was significant reduction in the occurrence of purulent eye discharges, but ectropion did not resolve. He was subsequently referred to another hospital for surgery.

DISCUSSION

Congenital ectropion is said to be a rare condition with about 100 reported cases in the literature since it was first described by Adams in1896.[2] Our report is the first from our institution and state, although several cases have been reported from other states in Nigeria.[4,14,15] There are also reports from many other parts of the world.[1,5] In fact, Miller *et al* reported the involvement of the four eyelids in a Down syndrome patient.[1] Except for being male and a Down syndrome patient, our patient had no other correlations such as difficult labor, collodion skin disease, lamella ichthyosis, grandmultiparity, or post-maturity.

The occurrence of ectropion in older age group in Down syndrome patient may be because of the suspected pathophysiology in them which makes resolution with conservative management unlikely. Only the upper eyelids were affected in our patient, though Sethapathy reported the involvement of the upper and lower eyelids in a 12 year old down syndrome patient.[17] The ectropion which was noticed in our patient at birth was managed at the delivery hospital with no resolution, and the child was discharged home without referral to specialists even though there was no resolution of the symptoms. This brings to the fore the need for early and prompt referral to the specialist so that appropriate treatment could be effected as soon as possible to limit complication and the discomfort the patient can experience with delayed intervention.

Complications detected in the child were epiphora and recurrent purulent eye discharge, but there was no exposure keratitis. Delay in treatment of ectropion could result in these complications.[4] This trend can be disrupted in moderate cases, when diagnosed early and treated swiftly with topical anti-inflammatory and antibacterial lubricants, moist dressings, eyelid tape, manual lid inversion, and pressure patching.[4,8,12] The administration of antibiotic ointments serve to reduce infection. The management of patients with severe ectropion require medial and lateral canthoplasties, full-thickness pentagonal resection of the upper eyelids and placement of skin grafts.[18] Ectropion surgery is generally very successful with 95% of patients corrected with one surgery.[18] The patient has not been seen in our clinic since he was referred for surgery.

CONCLUSION

Congenital ectropion is a rare entity which is more common in blacks. It may be associated with Down syndrome. This case is reported to create awareness, improve recognition and the understanding of this rare condition among health care professionals especially those who are in the units of obstetrics and neonatal care. Early involvement of eye specialists (ophthalmologists) in the care is essential to allow appropriate interventions to be made.

Conflict of Interests: Authors declare no conflict of interest.

Informed consent: Written informed consent was obtained from the patient's parents for the publication of this case report and accompanying images.

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