Unilateral congenital eyelid eversion: An unusual presentation

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Congenital eyelid eversion is a rare condition of unknown aetiology. It has a higher prevalence in males and African populations, particularly in west African countries. Eyelid eversion is thought to be caused by birth trauma and/or intrauterine factors in the presence of certain predisposing factors. These factors include a vertically short anterior lid lamellae or vertical elongation of the posterior lamellar, generalised tissue hyperelasticity, congenital elevator disinsertion, absence of levator muscle function and/or some other intrinsic lid abnormality. Once everted, irritation of the eye causes orbicularis muscle spasm. This acts as a sphincter resulting in conjunctival strangulation with venous congestion and stasis. Increasing venous congestion causes conjunctival oedema that worsens the eyelid eversion. Here, we present one of the first known cases of congenital eyelid eversion that was successfully treated with conservative measures in a South African setting.

S Afr J Child Health 2020;14(3):161-163. https://doi.org/10.7196/SAJCH.2020.v14.i3.1741

The baby boy was born at 41 weeks via emergency caesarean section (CS) with a birth weight of 3 680 g. The indication for CS was for fetal distress. The CS was done under general anesthesia with no opioids given.

The mother was a 24-year-old gravida 2 para 0 with one previous miscarriage. Her booking blood results were normal (rapid plasma reagin negative, HIV negative and Rhesus positive). Her pregnancy was complicated by pre-eclampsia that required treatment with magnesium sulphate. She had no history of substance abuse, nor a family history of genetic or connective tissue disorders. Both parents originated from Burundi with no family history of congenital eversion of the eyelid(s).

The baby was born through meconium stained liquor grade 3. No signs of chorioamnionitis were noted. At delivery, the baby appeared floppy but responded well to suctioning and stimulation, scoring APGARS of 9/10 at one minute, 9/10 at 5 minutes and 10/10 at minutes, losing 1 point for acrocyanosis. Mild respiratory distress was evident (nasal flaring and mild subcostal recessions) and the baby was transferred to the neonatal high care unit and put on nasal prong oxygen.

On initial examination, the baby had soft dysmorphic features (a 3rd fontanelle, flat nasal bridge, wide spaced nipples and a sandal gap), a café-au-lait spot (CALS) on the back and prolapse of the right superior conjunctival fornix secondary to chemosis resulting in mild eversion of the upper eyelid. Mild periorbital and eyelid oedema of the left eye was also present without eversion of the eyelid (Fig. 1). No obvious abnormalities of the eye were observed.

A preliminary diagnosis of congenital eversion of the right eyelid and transient tachypnoea of the newborn was made.

Chloramphenicol ointment was applied to both eyes every 8 hours to prevent bacterial infections. Moist 5% hypertonic saline dressings were applied every 6 hours to both eyes to prevent desiccation of the exposed conjunctiva. Slight pressure patching was also applied over the saline dressing.

Fig. 1. Picture showing prolapse of the right superior conjunctival fornix secondary to chemosis resulting in mild eversion of the upper eyelid at day 1 of life. Mild periorbital and eyelid oedema of the left eye without eversion of the eyelid

The Department of Ophthalmology was consulted, and they advised us to continue the management together with light eyelid pressure with a cotton earbud over the everted eyelid every 8 hours. If there was no reversion by day 5 of life, Ophthalmology was to be consulted again.

While in the neonatal high care unit, the baby had three episodes of bloody vomitus on day 1 and 2 of life. He was subsequently kept nil per os, started on omeprazole with a neonatolyte infusion for maintenance and a full blood count (FBC) and C-reactive protein

(CRP) were drawn for analysis. Both were within the normal limits (Table 1). No further episodes of bloody vomitus were reported on day 3 of life. Feeds were gradually increased successfully and trauma secondary to suctioning at birth was considered to be the probable cause of bloody vomitus.

Pustular melanosis was evident on day 2 of life. Additionally, concern was raised when no urine had been passed despite a normal abdominal and urological examination. A urinary catheter was passed, and concentrated urine was drained at a 3 cm insertion depth. Dipstix showed no abnormalities. An informal kidney, ureter and bladder (KUB) ultrasound showed normal kidneys and urine in the bladder. On further urine output monitoring, normal urinary output was recorded, and the catheter was removed on day 3.

Post KUB ultrasound, an unexpected blue discolouration was noted on the anterior abdominal wall with an irregular and undefined border. On day 3 of life, the umbilical stump detached on handling and a compression dressing was applied to the umbilical area to prevent blood loss. Blood was drawn after concerns of sepsis or a bleeding disorder were raised. All the analysed parameters were within normal limits (Table 1). This was an unusual case of congenital eyelid eversion given the additional soft dysmorphism, bloody vomitus, easy bruising and early detachment of the umbilicus.

We observed significant daily improvement of eyelid swelling bilaterally with the conservative management approach. There was significant reduction in conjunctival oedema with mild eversion of the right eyelid still being present on day 2 of life. There was also decreased oedema over the left eyelid with no eyelid eversion. (Fig. 2). There was complete resolution of the everted eyelid with no eversion noted during crying on day 3 of life (Fig. 3).

With the unusual presentation described above, the baby was kept for observation in the neonatal high care unit. No new complications occurred except for some concern of nasal flaring on day 6 of life. To exclude the development of sepsis, blood was drawn, and analysis revealed normal CRP levels (Table 1). The baby was discharged at day 9 of life.

The baby was followed-up 2 weeks later at the high-risk neonatology clinic and then at the genetics clinic at 4 months of age. He was feeding and growing well, and no concerns were raised by the parents. No dysmorphic features were noted. However, two (CALS), irregular hyperpigmentation of the right upper arm and a small left accessory nipple were observed. The mother was also observed to have 2 significant (CALS) with no obvious features of neurofibromas or freckling. She also had a similar hyperpigmentation lesion on her arm. The baby will be followed-up annually to monitor the skin lesions.

Discussion

We achieved complete and successful resolution of congenital upper eyelid eversion by day 3 of life using conservative methods. This is in line with the literature that has demonstrated the effectiveness of conservative management strategies in congenital eyelid eversion.[1-4] Surgical interventions such compression sutures, temporary tarsorrhaphy, subconjunctival injection of hyaluronic acid, fornix sutures and even full thickness skin grafts should only be considered if more rapid resolution is preferred[5] or conservative measures fail because of inherent lid abnormalities. The time it takes to completely resolve eyelid eversion using

Fig. 2. Picture showing significant reduction in conjunctival oedema with mild eversion of the right eyelid still present on day 2 of life. Mild oedema over the left eyelid with no eyelid eversion.

Fig. 3. Picture showing complete resolution of eyelid eversion on day 3 of life. Minimal oedema was seen bilaterally.

Table 1. Analysis of blood results at days 1, 2 and 6 of life

	WCC ($\times 10^9$/L)	Hb (g/dL)	Plt ($\times 10^9$/L)	CRP (mg/L)	INR	PTT (s)	Urine MC & S (/mL)
Normal	9 - 30	15 - 25	120 - 450	<10		30 - 40	
Day 1	16.18	17.5	213	<1			
Day 2	9.86	16.4	199	4	1.13	37.9	Leucocytes – 1 200 Erythrocytes = 0 Bacteria – observed Crystals – uric acid
Day 6				2			

WCC = white cell count; Hb = haemoglobin; Plt = platelets; CRP = C-reactive protein; INR = international normalised ratio; PTT = activated partial thromboplastin time; MC & S = microscopy, culture and sensitivity.

conservative measures varies from a few days to weeks in the literature.[1-4] The rapid response in this case was probably due to a milder form of congenital eyelid eversion.

The goal of management is to prevent desiccation of the exposed conjunctiva and cornea, allowing for spontaneous resolution of the eyelid eversion.[4] This is achieved by breaking the vicious cycle of eyelid eversion, orbicularis muscle spasm, venous congestion and worsening conjunctival oedema. The rationale behind using 5% hypertonic saline dressings is that they create an osmotic gradient that results in the movement of fluid from the oedematous tissue through the semipermeable conjunctival membrane.[4] Pressure patching and eyelid manipulation with gentle cotton earbud pressure to the chemotic conjunctiva is thought to help push the fluid out of the conjunctiva and decrease the amount of oedema. However, lid massage should be performed with caution under cardiac monitoring as the oculorespiratory reflex can result in respiratory arrest as previously described in one case.[6] Lastly, the antibiotic ointments help to prevent the development of secondary bacterial infections and offer an added benefit of acting as a lubricant to prevent desiccation.

The initial unusual presentation in this case was thought to be incidental in nature or explained by the presence of a non-significant connective tissue and/or genetic abnormality. A plexiform neurofibroma of the lacrimal gland in a child diagnosed with neurofibromatosis Type 1 has been described as a cause of congenital eyelid eversion.[7] Neurofibromatosis was an unlikely cause in this case given the rapid resolution of eyelid eversion and the non-convincing clinical history and examination of the child during follow-up. However, it must still be considered during follow-up visits.

Conclusion

Although this is a rare condition, it is important to create awareness of its existence. Congenital eyelid eversion can be particularly alarming if encountered for the first time. Fortunately, it is easy to diagnose and treat with an excellent prognosis using simple conservative measures.

Declaration. Ethical clearance was obtained from the George Hospital Ethics Committee and signed consent to publish images of the newborn was obtained from both parents.

Acknowledgements. None.

Author contributions. SA and IE designed and conceptualised the study, acquired the data, interpreted the data and wrote the manuscript. PN played a role in the acquisition of data and revised the manuscript. FR revised the manuscript. All the authors approved the final manuscript for publication.

Funding. None.

Conflicts of interest. None.

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Accepted 26 May 2020.