Case Report

By: *G Hamukali*¹,², K I M Muma¹,³

¹Department of Ophthalmology, School of Medicine and Clinical Sciences, Levy Mwanawasa Medical University, Lusaka, Zambia.
²Mwami Mission Hospital, Chipata, Zambia
³University Teaching Hospitals – Eye Hospital, Lusaka, Zambia

*EMAIL ADDRESS: Given Hamukali: hamukaligiven@yahoo.com*


ABSTRACT

This is a case report of a unilateral congenital proptosis that presented at Chipata Central Hospital (CCH) of a female neonate whose birth history was uneventful. At birth the neonate was given topical antibiotics and the parents were sure it was going to resolve. Upon noticing that the proptosis of the eye was not regressing, the parents took the neonate back to the health facility in Malawi where they were referred to CCH through Mwami Mission Hospital. After examination and investigations at CCH, a diagnosis of Lymphangioma was made for which the neonate was successfully managed and discharged.

INTRODUCTION

Proptosis is defined as the forward displacement of the eyeball [1]. This condition could be induced by inflammatory, vascular, infectious, cystic, neoplastic (benign and malignant), and traumatic factors [2].

Unilateral proptosis has several differential diagnoses, including microphthalmos with a cyst, congenital cystic eyeball, and unilateral congenital glaucoma, dermoid cyst of the orbit, neuroblastoma, neurofibroma, nasofrontal or sphenoidal meningocoele, orbital haematoma, haemangioma, and lymphangioma [3]. Congenital proptosis is uncommon but may potentially be underreported. Differential diagnosis of congenital proptosis includes congenital tumours (dermoid cyst, teratoma, retinoblastoma, rhabdomyosarcoma, neuroblastoma, neurofibroma, myofibroma), vascular malformations, lymphangiomas, cysts, encephalocele/meningomyelocele, bony abnormalities, abscesses, and thyrotoxicosis [4].

Lymphangiomas are relatively rare, benign, congenital malformations, which may affect the orbit, eyelids and conjunctiva [5, 6]. Lymphangiomas are commonly located behind the orbital septum and usually manifest with proptosis, as well as the pain caused by spontaneous intraliteral haemorrhage or upper respiratory infection [7].

CASE SUMMARY

A six day old female neonate from a rural village in Mchinji District of Malawi was presented to CCH by the mother who was concerned that the left eye protrusion was not regressing despite the treatment given and the assurance from medical personnel. The mother reported that the eye was swollen and gradually increasing in size and the neonate was unable to close the left eye. The neonate was born via spontaneous vaginal delivery with no complications. This neonate was a third child in the family and the siblings were reported to be healthy with normal eyes. General systemic evaluation was suggestive of respiratory distress and infected left eye. Visual interest was positive especially to her right eye which had normal anterior and posterior segment findings. Left eye findings included a mild proptosis, moderate chemosis, retracted eyelids, megalo-cornea, dilated irregular and fixed pupil. The patient was admitted in a paediatric ward pending laboratory and radiology investigations and consequent management. Ultrasound showed a small cystic orbital mass suggestive of lymphangioma and the C-reactive protein (CRP) was positive. Full Blood Count (RBC) confirmed sepsis and systemic antimicrobial therapy was commenced. Ultrasound and laboratory investigations were repeated to monitor the progress of the condition. During the 3 weeks of hospitalization, C-reactive protein became negative and the orbital mass had regressed. The patient was then discharged in good condition. The patient was reviewed a one month later the proptosis had regressed.

DISCUSSION

Lymphangiomas are relatively rare, benign, congenital malformations, which may affect the conjunctiva, lids and orbit of the eyes [5, 6]. Lymphangiomas are commonly located behind the orbital septum and usually manifest with proptosis, as well as the pain caused by spontaneous intraliteral haemorrhage or upper respiratory infection [7]. In this case the lymphangioma was found to be behind the septum in the orbit. In this article, we described a rare case of proptosis with sudden onset and complete regression, which led to no complications in a new-born referred to the neonatal ward of Chipata Central Hospital. Kasim and Gendeh (2013) reported that neonatal proptosis with post-orbital lymphangioma is a rare malformation, which has been associated with severe causes in previous studies. The rarity of such cases could be true because even at CCH such cases are rarely attended. Solarte et al., 2010, reported a case of acute proptosis in a 26-day infant caused by dural fistula, and the neonate was considered as the youngest case of acute proptosis to have been reported [9]. However, in this case report the neonate was born with a proptosis which was steadily increasing in size as was noted by the mother.

In another study, Erickson and Tse, (2014) presented a case of gross proptosis at birth as an uncommon manifestation of various lesions, which were likely to compromise vision and lead to deformity or death [2]. Furthermore, a study by Paragache et al., 2004, described the case of a one-month-old neonate with marked proptosis in the right eye. In this
In a case report, the neonate did not present with gross proptosis but enough to warrant intensive investigations to confirm the cause of proptosis.

In the study by Ghosh et al., acute basophilic leukaemia was reported as a rare diagnosis in a seven-month-old male neonate who presented with a 3-week history of bilateral proptosis. In a similar research, Salihu et al., described a 15-year-old male patient with orbital lymphangioma who presented with symptoms such as sudden pain, proptosis, visual loss, restricted eye movements, diplopia, decreased visual acuity, compressive optic neuropathy, and subconjunctival haemorrhage. The patient underwent surgical operation (orbital decompression) unlike the patient in this case.

Neonates, especially sick or preterm infants, are at risk of developing severe infections (such as bloodstream infections) during their stay on neonatal units. Infections are often difficult to diagnose early with certainty, and quick tests such as measuring the blood level of a protein that responds to infection (called CRP) are sometimes used to help make an earlier diagnosis. A low C-reactive protein level is better than a high one, because it indicates less inflammation in the body. No medical care has been proven effective for lymphangiomas however, treatment for sepsis and respiratory distress with antibiotics has seen some significant improvement. Lymphangioma is not responsive to radiation therapy or steroids. However, propranolol represents a potential option, which may be of benefit even for intractable diffuse lymphangiomatosis [10]. Antibiotics are given for secondary cellulitis as was the scenario in this case.

**CONCLUSION**

Lymphangiomas can present at birth and can be complicated with respiratory distress and sepsis which can effectively be treated with antibiotics without any surgical intervention.

**LIST OF REFERENCES**