IJN
Iranian Journal of Neonatology
http://ijn.mums.ac.ir

Open Access
Case Report

A Newborn with Right Eye Proptosis Secondary to Lymphangioma: A Case Report

Reza Sharafi, Afagh Hassanzadeh Rad*

Pediatrics Growth Disorders Research Center, School of Medicine, 17-Shahrivar Hospital, Guilan University of Medical Sciences, Rasht, Iran

ABSTRACT

***Background*:** Lymphangiomas are relatively rare, benign, congenital malformations, which may affect the conjunctiva, lids and orbit of the eyes. Lymphangiomas are commonly located behind the orbital septum and usually manifest with proptosis, as well as the pain caused by spontaneous intralesional hemorrhage or upper respiratory infection. Previous studies have proposed the major causes of neonatal proptosis. In this article, we present the case of a female neonate born to a mother with triplet pregnancy. On day 28 of birth, clinicians were concerned about the proptosis in the right eye of the second sibling, and the neonate was evaluated via repeated ultrasound. The infant had good general appearance, and after ophthalmological consultation, both neonates were discharged on day 30 of birth. According to clinicians, the right eye proptosis was possibly caused by a benign lesion, such as lymphangioma. Moreover, it was noted that the proptosis would be absorbed gradually, and no interventions were required for the neonate. As predicted, the proptosis was completely absorbed at two months of age. Therefore, it is recommended that clinicians consider benign causes for neonatal proptosis, such as lymphangioma originating from neonatal sepsis.

Keywords: Lymphangiomas, Neonate, Proptosis

Introduction

Multiple pregnancies are commonly associated with high risk of miscarriage and premature birth (1). Preterm neonates are at an increased risk of acute and chronic morbidities (2). According to the literature, multifetal pregnancy and preterm birth could lead to prolonged admission in the neonatal intensive care unit (NICU) (3).

Proptosis is defined as the forward displacement of the eyeball (4). This condition could be induced by inflammatory, vascular, infectious, cystic, neoplastic (benign and malignant), and traumatic factors (5).

In children, orbital cellulitis often leads to unilateral proptosis and neuroblastoma, while leukemia may cause bilateral proptosis. Other conditions that might rarely induce childhood proptosis include rhabdomyosarcoma, retinoblastoma, capillary hemangioma, dermoid cyst, optic nerve gliomas, granulocytic sarcoma, and metastatic disease (6).

In the literature, it has been noted that clinicians may consider congenital orbital teratoma as an extremely rare tumor of the orbit in newborns with unilateral proptosis.

Unilateral proptosis has several differential diagnoses, including microphthalmos with cyst, congenital cystic eyeball, unilateral congenital glaucoma, dermoid cyst of the orbit, neuroblastoma, neurofibroma, nasofrontal or sphenoidal meningocele, orbital haematoma, hemangioma, and lymphangioma (7).

Lymphangiomas are relatively rare, benign, congenital malformations, which may affect the conjunctiva, lids and orbit of the eyes (8, 9). Lymphangiomas are commonly located behind the orbital septum and usually manifest with proptosis, as well as the pain caused by spontaneous intralesional hemorrhage or upper respiratory infection (10).

Previous studies have proposed the major causes of neonatal proptosis. In this article, we present a rare case of neonatal right eye proptosis secondary to lymphangioma.

Case presentation

In this study, we aimed to describe the case of a female newborn presented with right eye proptosis secondary to lymphangioma on day 28 of birth. The neonate was born to a mother

* *Corresponding author*: Afagh Hassanzadeh Rad, Pediatrics Growth Disorders Research Center, School of Medicine, 17-Shahrivar Hospital, Guilan University of Medical Sciences, Rasht, Iran. Tel: 01333369002; Email: afaghrad@gums.ac.ir

with triplet premature pregnancy (30 weeks and two days). The first neonate died at birth, and two other siblings were hospitalized in the NICU of Al-Zahra Hospital in Rasht, Iran for 30 days.

Both alive neonates were female with Apgar scores of seven and eight, and birth weight of these infants was calculated at 1,250 and 970 grams, respectively. The mother was 30 years old with intermediate thalassemia.

Sepsis and respiratory distress syndrome workup was performed on both neonates. Moreover, the infants received antibiotic treatment with ampicillin and amikacin. Also, neonatal brain ultrasound was normal during hospitalization, C-reactive protein was negative, and laboratory results were normal for both siblings.

On day 28 of birth, the second sibling manifested symptoms of proptosis in the right eye.

Results

The neonate with right eye proptosis was evaluated via repeated ultrasound. She had good general appearance, and after ophthalmological consultation, both neonates were discharged on day 30 of birth.

On day 37 after birth, ocular CT-scan was performed on the second sibling, and the results revealed a post-orbital mass. Moreover, a large low-density mass was detected in the right side of the retrobulbar space leading to the expansion of the globe, severe exophthalmos, obliterated and encased optic nerve, and extraocular muscle displacement. Neonatal evaluations were indicative of no bone destruction and intracranial extension. As presumed by the clinicians, the right eye proptosis was possibly caused by a benign lesion, such as lymphangioma (Figure 1). In addition, the clinicians stated that the proptosis would be absorbed gradually, and no

Figure 1. Proptosis in Right Eye

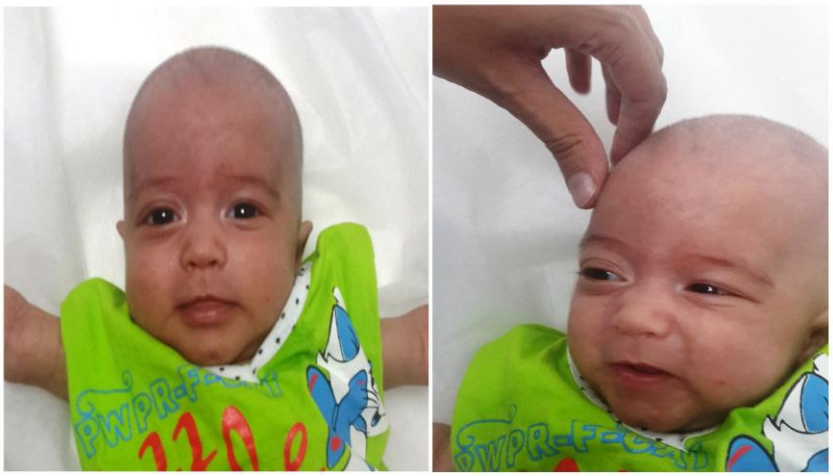

Figure 2. Absorbed Proptosis

Interventions were required for the neonate. As predicted, the proptosis was completely absorbed at two months of age (Figure 2).

At two months of age, both infants received vaccination against diphtheria, pertussis, tetanus, poliomyelitis, and hepatitis B. However, the third sibling was referred to the hospital after the vaccination due to high fever and convulsion, and unfortunately, she died. Consequently, the clinicians recommended hospitalization for the second sibling for further monitoring. After complete control of the second neonate, she was discharged, and after nine months of follow-up, she was reported to have normal growth and development.

Discussion

In this article, we described a rare case of proptosis with sudden onset and complete absorbance, which led to no complications in a newborn referred to the neonatal ward of Al-Zahra Hospital in Rasht, Iran. To the best of our knowledge, neonatal proptosis with post-orbital lymphangioma is a rare malformation, which has been associated with severe causes in previous studies. For instance, Solarte 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 (11).

In another study, Erickson et al. 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 (12). Furthermore, a study by Paragache described the case of a one-month-old neonate with marked proptosis in the right eye.

In the study by Ghosh et al., acute basophilic leukemia was reported as a rare diagnosis in a seven-month-old male neonate 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 presented with symptoms such as sudden pain, proptosis, visual loss, restricted eye movements, diplopia, decreased visual acuity, compressive optic neuropathy, and subconjunctival hemorrhage. The patient underwent surgical operation (orbital decompression) (9). In the present study, no interventions were required.

In another study, Murthy et al. described 12 cases of acute myeloid leukemia with proptosis. Moreover, orbital mass, visual symptoms, and subconjunctival hemorrhage were reported in five (41.7%), two (16.7%) and one (8.3%) of the patients (6). On the other hand, the results obtained by Porto et al. indicated that most children with granulocytic sarcoma, which is a rare presentation of acute myeloid leukemia, were referred with proptosis (14).

According to the results of the present study, it is recommended that clinicians consider benign causes for neonatal proptosis, including lymphangioma originating from sepsis.

Acknowledgements

Hereby, we extend our gratitude to all the colleagues who assisted us in this research project.

References

1. Papageorghiou AT, Avgidou K, Bakoulas V, Sebire NJ, Nicolaides KH. Risks of miscarriage and early preterm birth in trichorionic triplet pregnancies with embryo reduction versus expectant management: new data and systematic review. Hum Reprod. 2006; 21(7):1912-7.
2. Sengupta S, Carrion V, Shelton J, Wynn RJ, Ryan RM, Singhal K, et al. Adverse neonatal outcomes associated with early-term birth. JAMA Pediatr. 2013; 167(11):1053–9.
3. Afrasiabi N, Mohagheghi P, Kalani M, Mohades G, Farahani Z. The effect of high risk pregnancy on duration of neonatal stay in neonatal intensive care unit. Iran J Pediatr. 2014; 24(4):423-8.
4. Giugni AS, Mani S, Kannan S, Hatipoglu B. Exophthalmos: a forgotten clinical sign of cushing's syndrome. Case Rep Endocrinol. 2013; 2013:1-3.
5. Mercandetti M. Exophthalmos fallow-up. Medscape. Available at: URL: http://emedicine.medscape.com /article/1218575-followup; 2012.
6. Murthy R, Vemuganti GK, Honavar SG, Naik M, Reddy V. Extramedullary leukemia in children presenting with proptosis. J Hematol Oncol. 2009; 2:4-11.
7. Paragache G, Panda NK, Joshi K. Unilateral progressive proptosis in neonate. Indian J Otolaryngol Head Neck Surg. 2004;.56(1):35-6.
8. Al-Salem AH. Lymphangiomas in infancy and childhood. Saudi Med J. 2004; 25(4):466-9.
9. Salihu N, Sylaj A. Orbital lymphangioma. Int J Case Rep Imag. 2014; 5(1):28–31.
10. Kasim KS, Gendeh BS. Case report: infantile orbital lymphangioma with a rare ENT presentation. J Rhinol Otol. 2013; 1(1):17-19.
11. Solarte CE, Levin AV, Armstrong D. Acute proptosis in a newborn infant: a presentation of infantile dural fistula. J AAPOS. 2010; 14(1):88-9.

12. Erickson BP, Tse DT. Management of neonatal proptosis: a systematic review. Surv Ophthalmol. 2014; 59(4):378-92.
13. Ghosh I, Bakhshi S, Gupta R. Acute basophilic leukemia in an infant with proptosis. Indian J Pathol Microbiol. 2011; 54(1):210-1.
14. Porto L, Kieslich M, Schwabe D, Zanella FE, Lanfermann H. Granulocytic sarcoma in children. Neuroradiology. 2004; 46(5):374–7.