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Unusual Case of Unilateral Neonatal Eyeball Luxation: A Case Report

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Abstract

The newborn infant presented with severe proptosis. Data on clinical history, presentation, photos, radiological imaging, and laboratory results were presented. A literature review was conducted for the case and relevant treatment modalities. The surgical technique and the outcome were also discussed. The mass was present since birth in a full term neonate, causing complete globe dislocation of the left eye. Imaging showed a large cystic retro-bulbar mass completely occupying the orbital cavity and causing anterior dislocation of the left globe. Surgical excision of the tumor was carried out with globe preservation and Histopathology examination confirmed the diagnosis of optic nerve glioma. This was an atypical presentation of an optic nerve glioma causing globe dislocation in a neonate. The resection of such a large orbital tumor made globe preservation possible and resulted in an excellent cosmetic outcome.

Keywords

Neonatal Proptosis, Optic Nerve Glioma, Globe Dislocation, Globe Preservation, Cosmetic Surgery

1. Introduction

Optic pathway glioma comprises about 5% of all intracranial tumors in pediatric patients [1]. It is commonly associated with neurofibromatosis type 1 (NF1) for whom the prognosis was found to be better than patients without features of NF1. Optic pathway glioma develops in around 20% of patients with NF1 [2].

Tumor mainly affects children about the age of 6 years and the most common presenting sign or symptom is squint [3]. Optic nerve gliomas are diagnosed based on clinical examination and imaging. Obtaining a tissue biopsy is not recommended for the diagnosis since it carries a high risk of visual loss [4]. In general, treatment for optic nerve gliomas is not required unless there is a noticeable mass effect causing symptoms such as pain, disfigurement, or severe visual impairment [5]. Radiation and chemotherapy can delay or stabilize visual deterioration and have also been found to improve visual outcomes in some cases [6]. In our case report, we are describing unusual presentation of optic nerve glioma causing huge proptosis and globe subluxation in newborn baby.

2. Case Presentation

A 5-days-old baby girl was referred from a Peripheral hospital to a tertiary specialized hospital, Riyadh, Saudi Arabia as a case of huge proptosis of the left eye noticed since birth. The baby was born by a typical vaginal birth and was full term. She is the first child of her parents. There is no history of maternal disease or similar diseases in the family, nor is there any consanguinity between the parents. Upon examination, she was vitally stable and had a pronounced left eye proptosis (the eye was completely outside the orbital cavity) (**Figure 1**). There was chemosis and severe exposure keratopathy with corneal haze and exposure related diffuse corneal infiltrate, the anterior chamber was deep with 1 mm whitish hypopyon, pupil was not reactive to light with no view posteriorly. Right eye examination was totally normal. The baby was medically free with normal general examination and no other malformations were noticed. The baby was admitted under multidisciplinary care involving ophthalmology, neonatology and oncology for further investigations and treatment. Corneal scraping was done but the result was negative so, Corneal infiltrate was treated with topical fortified broad-spectrum antibiotics (Cefazoline and Ceftazedime 50 mg/mL alternating around the clock) and the hyopyon resolved within few days. Measures were taken to treat exposure keratopathy and to prevent corneal perforation by using

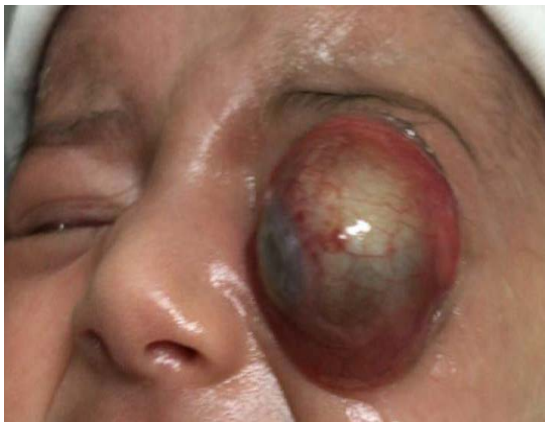
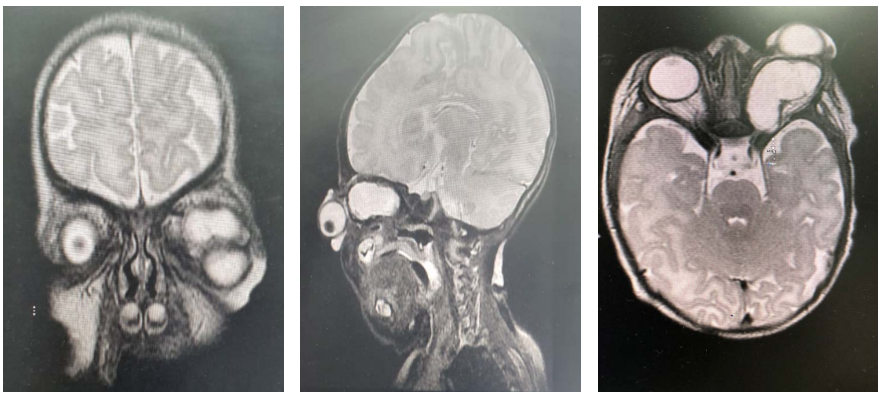


Figure 1. External photo of the patient showing huge proptosis and subluxation of the left eye.

moist chamber and heavy lubrication (Figure 2). Our top differential diagnosis in the beginning includes Teratoma, Neuroblastoma, Retinoblastoma and orbital lymphangioma. Patient underwent serial laboratory tests which were normal including alpha fetoprotein and urine catecholamine. A diagnostic ultrasound B scan was performed. It showed vitreous condensation, a flat retina and there was retro-Bulbar mass pushing the globe forward. Magnetic Resonance Imaging with contrast (MRI) of the brain and orbit was performed (Figure 3). It showed a large intraconal cystic mass non-enhancing with gadolinium on the left orbit; which measures 30 mm Antero posteriorly and 16 mm transverse dimensions, extending from the orbital apex until the scleral margin and completely occupying the orbital cavity. It was reported also a thin internal septation. The extraocular muscles were separable from the mass but the optic nerve which appeared incorporated in the mass, indicating a cystic tumor involving the optic nerve. There was an irregular enhancing tissue extending from the inferolateral wall of the left globe and extending to the retrolental region highly suggestive of persistent fetal vasculature. The brain was normal. MRI of the abdomen and pelvis was performed to rule out metastatic neuroblastoma, which showed normal findings. CT of the brain and orbits with contrast was also performed. It showed

Figure 2. Moist chamber to keep the eye wet and to protect the cornea from injuries.



(A) (B) (C)

Figure 3. MRI with contrast showing retro-bulbar mass in different views ((A) Axial, (B) Sagittal, (C) Coronal).

mural calcifications of the lesion, there were no intraocular calcifications, and the orbital bones were intact. Histopathology report (Figure 4) showed that the tumor mass was grossly tubular fusiform in shape, and the optic nerve was completely replaced by a spindle cell tumor which was S100, Glial Fibrillary Acidic Protein (GFAP) and Neuron Specific Enolase (NSE) positive. The presence of meningeal cells positive for Epithelial Membrane Antigen (EMA) and Progesterone Receptors (PR) immuno-stains at the periphery of the tumor supports optic nerve origin. Mitosis is noted and KI-67 (proliferation marker) was as high as 10% which might reflect a more aggressive behavior. So, the diagnosis of Optic nerve glioma was established. The surgery was then planned and discussed with the parents and they agreed to proceed. An anterior orbitotomy was carried out through a conjunctival incision, the lateral rectus was disinserted to gain more room and accessibility to the lesion; once the anterior part of the tumor was exposed and gently dissected from the surrounding tissues including a careful apical dissection since the tumor aroused from the optic nerve. The tumor was then completely removed along with the optic nerve. The lateral rectus muscle was then reattached to its insertion on the globe and the conjunctival wound was sutured, as such, the globe was preserved with good cosmetic result.

3. Outcome

Although there was no visual potential in the left eye since the presentation, the globe preservation ended up with excellent cosmetic outcomes and family satisfaction (Figure 5). The baby is doing well, with no tumor recurrence observed for a one-year follow-up period. Sensory Esotropia was developed in the blind eye for which she is following up with pediatric ophthalmology for possible intervention later in life.

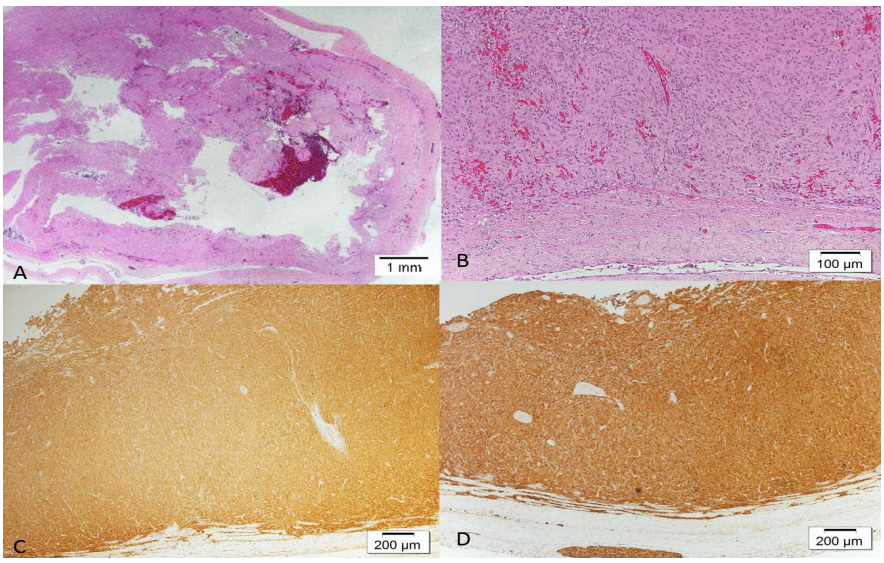


Figure 4. (A) Low power showing the tubular structure with proliferative tumor. H & E ×12.5. (B) High power showing proliferative spindle cell tumor that stains positive with neuron specific enolase (C) and S100 (D). (B) (C) & (D) ×40.

Figure 5. External photo 5 Months after tumor excision showing excellent cosmetic outcome.

4. Discussion

In reference to the study conducted by Varan and his group, they studied 101 cases of optic pathway glioma and discovered that the median age of presentation was about 6 years, and the disease was more common in patients with NF1 [3]. In our reported case, the disease presented since birth as a huge unilateral proptosis and globe dislocation, explaining the intrauterine growth of the tumor. Patients older than 10 years and those with NF1 are known to have better prognosis and less severity of the disease, which explains the aggressive behavior in our case, in which the disease developed early in life and without features of NF1. In their systemic review, Erickson and Tse reported three challenging cases of neonatal proptosis caused by neuroblastoma, infantile fibrosarcoma, and simple epithelial cyst. They described the possibility of globe preservation in 2 out of 3 cases, which ended up with an excellent cosmetic outcome [7]. Progressive proptosis in a 14-day-old neonate due to Hemangiopericytoma was found in literature, and the diagnosis was confirmed by histopathological evaluation [8]. Other rare causes of neonatal proptosis, like teratoma and trauma by Kielland's forceps (forceps delivery), were reported in literature [9] [10].

The treatment of Optic pathway gliomas is still controversial, but it depends on the age, location of the tumor and visual potential. So, if there is no visual potential like in our case and there is disfiguring proptosis, surgical intervention is recommended [11]. No peculiar MRI findings can help in the differential diagnosis [12]. Aggressive surgical treatment of optic pathway gliomas decreases both morbidity and mortality [13], as exemplified in our case where complete tumor resection led to an excellent prognosis with no tumor recurrence during a 2-year follow-up period.

5. Conclusion

This case report reports an unusual presentation of optic nerve glioma causing severe globe dislocation at birth in a full-term newborn, and complete tumor resection was carried out with globe preservation and excellent cosmetic outcome.

Informed Consent

Written consent was obtained from the patient's parents to disclose and publish

her medical information. Ethical approval was obtained from the Institutional Review Board (IRB: 1910-CR PI, KKESH).

Statement

All authors participated in diagnosis or treatment of the case, or the writing of this case report.

Conflicts of Interest

The authors declare no conflicts of interest regarding the publication of this paper.

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