Case Report

Congenital orbital teratoma: A case report

Sana Gull*[1], Mahwish Faizan[1], Salma Sadia[2], Ahmad Raza[3]

1. Department of Pediatric Hematology-Oncology, University of Child Health Sciences Lahore. 2 Department of Obstetrics and Gynecology, Sharif Medical and Dental City Hospital, Lahore, Pakistan 3. Department of Pediatric Ophthalmology, University of Child Health Sciences Lahore, Pakistan

Correspondence*: Sana Gull, Department of Pediatric Hematology- Oncology, University of Child Health Sciences Lahore, Pakistan, Pakistan, E-mail: sanagull49@gmail.com.

 | Submitted on: 17-10-2024 | Accepted on: 20-02-2025

Conflict of interest: None | **Source of Support:** Nil

Cite as: Gull S, Faizan M, Sadia S, Raza A, Congenital orbital teratoma: A case report. *J Child Health Sci.* 2026; 1:56-58.

ABSTRACT

Background: Teratomas are germ cell tumors composed of multiple tissue types originating from one or more of the germ cell layers, i.e., ectoderm, endoderm, and mesoderm. Congenital teratomas of the orbit are rare.

Case Report: We report a case of a congenital orbital teratoma in a 24-day-old neonate who presented with a swollen eyeball at birth, which progressively increased in size, leading to proptosis of the right eye. Congenital orbital teratoma was suspected based on clinical findings since birth and radiological imaging. Complete tumor excision and exenteration were performed, and the final diagnosis of a mature congenital orbital teratoma was confirmed histologically.

Conclusion: Congenital orbital teratomas are rare but potentially curable tumors. They should be considered as a differential diagnosis in cases of neonatal proptosis.

Keywords: Neonate, Proptosis, Orbital, Teratoma, Exenteration.

INTRODUCTION

Teratomas are neoplasms composed of cells from the germ cell layers [1]. They account for 6–10% of childhood tumors, most commonly arising in the male gonads (testes), female gonads (ovaries), or retroperitoneum [2]. Orbital localization is very rare, with an incidence of only 0.8% [2].

Orbital teratomas predominantly affect the left eye and show a female predominance with a ratio of 2:1 [3, 4]. These neoplasms enlarge rapidly, leading to disfiguring proptosis and causing keratopathy due to environmental exposure within days [5].

Herein we report a case of orbital teratoma managed with excision successfully.

CASE REPORT

We present the case of a 24-day-old female neonate who was brought to our hospital with massive proptosis of the right eye, a condition present since birth (Figure 1). She was born full-term through an uneventful spontaneous vaginal delivery (SVD) at a private hospital. However, there had been no regular antenatal follow-ups, and anomaly scans were not performed. Her mother's antenatal history was unremarkable, with no known risk factors or complications.

On examination, the infant's right eye exhibited extreme proptosis due to a large, multilobulated mass that completely occupied the eyeball. The pupils were distorted, and no distinct ocular structures could be identified. The conjunctiva

was markedly reddened, and the cornea showed destructive changes due to prolonged exposure keratopathy. Both the anterior and posterior segments of the eye were obscured. A B-scan ultrasonography confirmed the presence of calcifications within the vitreous humor of the right eye. In contrast, the left eye appeared completely normal. General and systemic examinations also revealed no abnormalities.

Given the clinical findings, we considered several differential diagnoses, including retinoblastoma, rhabdomyosarcoma, and orbital teratoma. MRI of the brain and orbit revealed a 7.6 × 7.7 × 6.2 cm heterogeneously enhancing solid and cystic lesion in the right orbit. The mass extended through the orbital apex, causing its widening, and the normal orbital contents were not discernible. However, there was no evidence of intranasal or intracranial extension of the disease. Brain imaging appeared normal.

Laboratory investigations, including a complete blood count, liver function tests (LFTs), renal function tests (RFTs), serum electrolytes, and serum alpha-fetoprotein levels, were all within the normal range for age. Given the extent of the mass, we proceeded with complete tumor excision through a lid-sparing modified right eye exenteration. The surgery was uneventful, and the empty socket was packed with a sterile dressing.

Histopathological analysis of the excised mass revealed a well-formed 10 × 8 × 4 cm tumor containing stratified squamous epithelium with hair follicles, myxoid stroma, fat, primitive glandular structures, cartilage, and inflammatory cells. Additionally, nerve bundles, mucinous epithelium, bone trabeculae, a cartilaginous cap, mature glial tissue, and sebaceous tissue were identified, confirming the diagnosis of a mature teratoma.

Cosmetic rehabilitation is planned once the socket is adequately covered with granulation tissue. The patient was advised to return for regular follow-ups every three months, with continued monitoring through ophthalmology and oncology outpatient visits.

DISCUSSION

Teratomas are the most common germ cell neoplasms of the orbit, arising from primitive stem cells that are carried to the orbit via blood circulation. Congenital orbital teratomas are rare but can be potentially vision-threatening tumors. To date, approximately 70 cases have been reported worldwide, with only two cases documented from Pakistan [5,6]. Most reported cases are unilateral [7], with a predominance in the left eye; however, our index case involved a teratoma of the right orbit.

Figure 1: Massive proptosis

The differential diagnoses of orbital teratomas include rhabdomyosarcoma, retinoblastoma, lymphangioma, epidermal inclusion cyst, hemangioma, dermoid cyst, and encephalocele [6].

Mizuo [8] and Duke-Elder [9] classified orbital teratomas into five categories:

1. A whole fetus inserted into the orbit (parasitic orbit).
2. A second fetus partially inserted into the orbit.
3. A neoplasm containing components of all three germ layers—ectoderm, endoderm, and mesoderm.
4. A neoplasm containing components from any two of the three germ layers.
5. A neoplasm composed of structures derived from a single germ layer.

According to this classification, our index case falls into Group 3, as the tumor contained elements from all three germ layers.

The primary goals of management are early diagnosis to preserve vision and the globe whenever possible, along with complete tumor excision. However, management can be challenging due to the tumor's rapid growth, making vision and globe preservation difficult. In our case, salvage

of the globe and vision was not feasible due to extensive tumor involvement of orbital structures, as confirmed by MRI, along with exposure keratopathy leading to complete destruction of the eyeball. Nevertheless, early surgical intervention was performed to prevent further tumor progression, which could have led to damage to adjacent structures and necessitated more radical procedures like total exenteration.

Immature teratomas are extremely rare and life-threatening, with a poor prognosis—only 7% of affected infants survive beyond the first year of life [6]. In cases of mature teratomas, serum alpha-fetoprotein (AFP) levels have limited diagnostic value, as newborns naturally exhibit elevated levels of this tumor marker. However, regular post-surgical oncology follow-ups, including ophthalmological examinations and periodic serum AFP measurements, are essential. A rising AFP level postoperatively may indicate tumor recurrence or metastasis [10].

Despite their rarity, congenital orbital teratomas require early recognition and appropriate management. It is crucial for general pediatricians, oncologists, and ophthalmologists to be aware of the importance of early diagnosis, timely treatment, and regular follow-ups to ensure the best possible outcome for the patient.

CONCLUSION

Congenital orbital teratomas are rare but potentially curable tumors. They should be considered in the differential diagnosis of neonatal proptosis. The primary goal of treatment is complete surgical removal of the tumor as early as possible, with an emphasis on preserving the globe whenever feasible.

Consent to Publication: Author(s) declared taking informed written consent for the publication of clinical photographs/material (if any used), from the legal guardian of the patient with an understanding that every effort will be made to conceal the identity of the patient, however it cannot be guaranteed.

Authors Contribution: The authors confirm contribution to the paper as follows: **SG**: Conception and design, drafting and writing of manuscript, **MF**: Direct Supervision, critical revision, final approval, **SS:** Primary attending gynecologist who observed the congenital anomaly and referred for further management. Literature review and drafting of manuscript,**AR**: Ophthalmologist who operated the patient. Provided surgery details, literature review and manuscript drafting.

Acknowledgments: Histopathology and Radiology department of The Children's hospital, University of child health sciences, Lahore.

REFERENCES

1. Prakash MVS, Indira R, Radhakrishnan M, et al. Malignant orbital teratoma in a neonate: A clinicopathological case report. *J Postgrad Med*. 2017;63(3):203-5.
2. Tsoutsanis PA, Charonis GC. Congenital orbital teratoma: a case report with preservation of the globe and 18 years of follow-up. *BMC ophthalmology*. 2021;21:1-7.
3. Reinhold A, Meyer P, Bruder E, et al. Congenital orbital teratoma: A clinicopathologic case report. *Am J Ophthalmic Clin Cases*. 2022;26:101420.
4. Pellerano F, Guillermo E, Garrido G, et al. Congenital orbital teratoma. Ocul Oncol Pathol. 2017;3(1):11-6.
5. Raza A, Mushtaq A, Qayyum S, et al. Congenital orbital teratoma: A case report. *J Neonatal Surg*. 2020;9:9.
6. Chaudhary R, Qamar NM, Riaz A. Mature orbital teratoma. *J Coll Physicians Surg Pak*. 2021;31(5):596-8.
7. Gemolotto G, Gaipa M. Quadro bilaterale di teratoma epibullbare associate a malformazioni palpebrali. *Arch Ottalmol*. 1956;60:161-70.
8. Mizuo G, Eine S. Foetus in orbita: Orbitopagus parasiticus. *Arch Augenheilkd*. 1910;65:365-83.
9. Duke-Elder S. System of ophthalmology. Vol. 3, Normal and abnormal development, Part 2. London: Henry Kimpton; 1964. p. 966-75.
10. Brandão V, Neves LM, Rangel LG, Fiúza SA, Sobreira T, Régis Pacheco LF, et al. Immature orbital teratoma: A rare case diagnosed in utero – A clinicopathological case report. *Pan Am J Ophthalmol*. 2023;5:5..