

Case report

Orbital teratoma in neonates: a report of a rare congenital orbital mass

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Abstract:

The eye is an essential organ for vision. In newborns, there are various conditions that can cause an orbital mass, such as retinoblastoma, rhabdomyosarcoma, lymphoma, leukemia, and metastatic cancer, among others. In this report, we present a case of a male neonate who was diagnosed with a congenital orbital teratoma, which is a rare disease and can have a significant impact on the patient's quality of life in the future. Surgery is the primary treatment for this condition.

Keywords : ● Teratoma ● Orbital mass ● Neonate

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รายงานผู้ป่วย

เนื้องอกเทอร่าโตมาที่ตาในทารกแรกเกิด

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บทคัดย่อ

ตาเป็นอวัยวะสำคัญต่อการมองเห็น ภาวะก้อนที่ตาเกิดได้จากหลายสาเหตุที่พบโดยส่วนใหญ่ได้แก่ โรคมะเร็งจอตาในเด็ก มะเร็งกล้ามเนื้อลาย มะเร็งต่อมน้ำเหลือง มะเร็งเม็ดเลือดขาว มะเร็งระยะแพร่กระจาย เป็นต้น รายงานนี้เป็นการรายงานผู้ป่วยทารกแรกเกิดที่ได้รับการวินิจฉัยว่าเป็นเนื้องอกเทอร่าโตมาที่ตา พบได้ไม่บ่อยและมีผลต่อคุณภาพชีวิตของผู้ป่วยในอนาคต การผ่าตัดเป็นการรักษาหลักของผู้ป่วยโรคนี้

คำสำคัญ : ● เทอร่าโตมา ● โรคเนื้องอกของเบ้าตา ● ทารกแรกเกิด

วารสารโลหิตวิทยาและเวชศาสตร์บริการโลหิต. 2567;34:67-71.

Introduction

The eye is a vital organ responsible for vision. In children, various conditions can lead to the development of an orbital mass, with the most common being retinoblastoma, rhabdomyosarcoma, lymphoma, leukemia, and metastatic cancer. Teratomas, on the other hand, consists of all three germ cell layers, mostly found in the testes, ovaries, or retroperitoneum. Teratomas are rare in the orbital location and account for less than 1% of teratomas in children¹⁻³.

Orbital teratomas are generally benign in histology, and the prognosis of the disease depends on resectability, age, and histologic grading. If complete excision can be performed at a younger age, the prognosis is excellent. Clinical manifestations of orbital teratoma include a massive, rapidly growing, unilateral orbital mass that can cause proptosis, exposure keratopathy, and loss of vision. It is not associated with bony destruction or intracranial or sinus involvement¹⁻³.

Case Report

A male infant, weighing 3,150 grams, was born at 38 weeks' gestation via normal labor. His mother had gestational diabetes and did not adhere to dietary management or receive treatment during pregnancy. At birth, he presented with a large right orbital mass; the right eye was bulging and unable to close. No discharge or signs of infection were observed. The left eye showed no abnormalities. He was referred to King Chulalongkorn Memorial Hospital for further treatment at the age of 20 days. The right orbital mass rapidly grew, and the cornea appeared cloudy. A physical examination revealed a reddish, round, right orbital mass measuring ten by 8 centimeters, with marked proptosis and chemosis (Figure 1). Examination of the head and face showed normal contour and no deformities. There was no hepatosplenomegaly or lymphadenopathy.

Initial laboratory investigations were performed consisting of a complete blood count that revealed Hb 14.1 g/dL, Hct 42.7%, WBC 18,900 cell/mm³, Neutrophil 39%,

Lymphocyte 49%, Monocyte 9%, Eosinophil 1%, Basophil 2%, and Platelet 333,000 cell/mm³. The tumor markers, including Alpha-Fetoprotein (AFP), Beta-Human Chorionic Gonadotropin (B-HCG), and Lactate Dehydrogenase (LDH), were measured, and their levels were found to be 30,905 (0-93,454 in term neonate), 2.5, and 365 U/L, respectively. The serum NSE level was 31.8 ng/mL which was in normal limit. The patient's CBC and all tumor markers were normal according to age.

A CT scan of the brain and orbit revealed a heterogeneous enhancing mixed solid-cystic lesion measuring 3.0 by 2.5 by 3.0 centimeters located at the extracranial superior aspect of the right orbit, adjacent to the right superior rectus muscle. Additionally, CT scans of the chest and abdomen were performed, detecting no abnormalities.

Based on the results of the physical examination and initial laboratory and radiologic studies, the possible diagnoses were congenital orbital teratoma, congenital neuroblastoma, retinoblastoma, and optic nerve sheath tumor. However, the vision prognosis in this patient on the affected eye was unlikely to be preserved.

Subsequently, the patient underwent evisceration with total tumor removal surgery on the right eye. During the procedure, it was observed that the patient had a severe corneal ulcer and chemosis in the right eye with clear vitreous content. Through a lateral canthotomy incision and cantholysis, the tumor, along with its capsule, was separated from the orbit and successfully removed. The right optic nerve was intact (Figure 2).

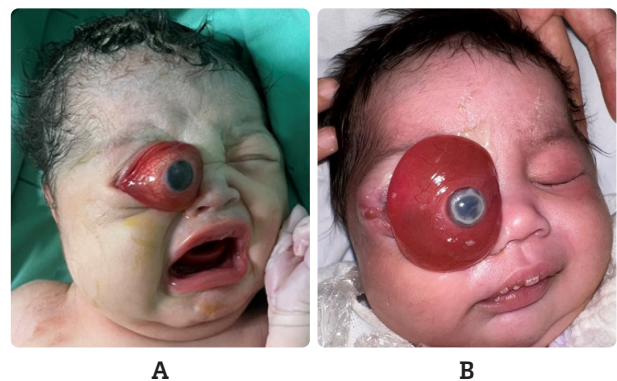


Figure 1 Orbital mass at birth (A) and at age 20-day-old (B)



Figure 2 Total tumor removal with evisceration of the right eye

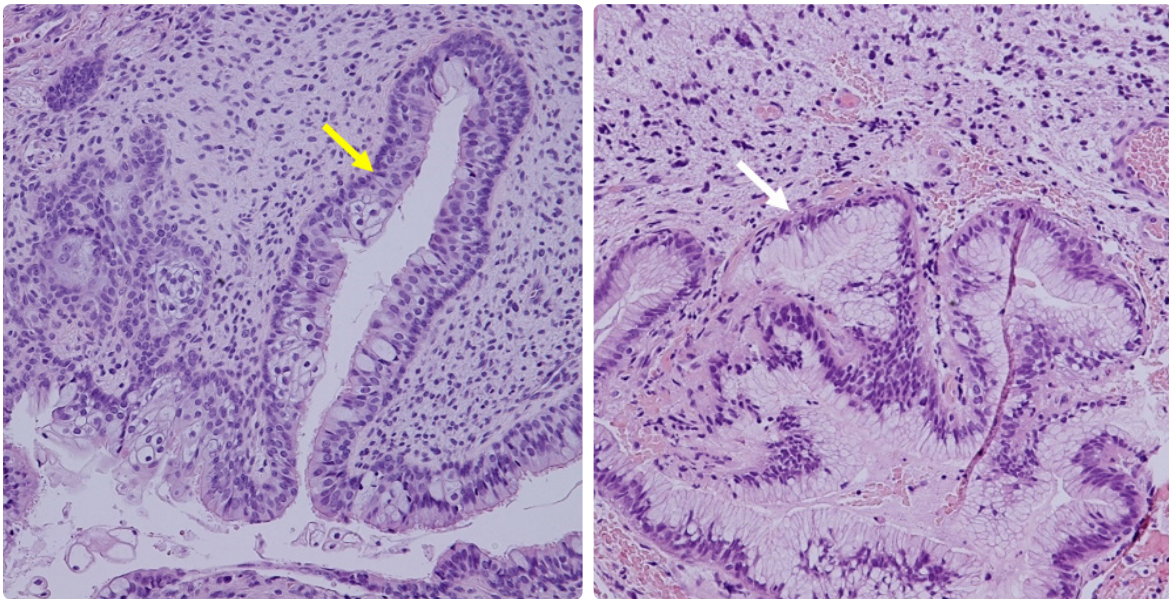


Figure 3 Endoderm is identified: Respiratory epithelium (yellow arrow), Foveolar epithelium of stomach (white arrow)

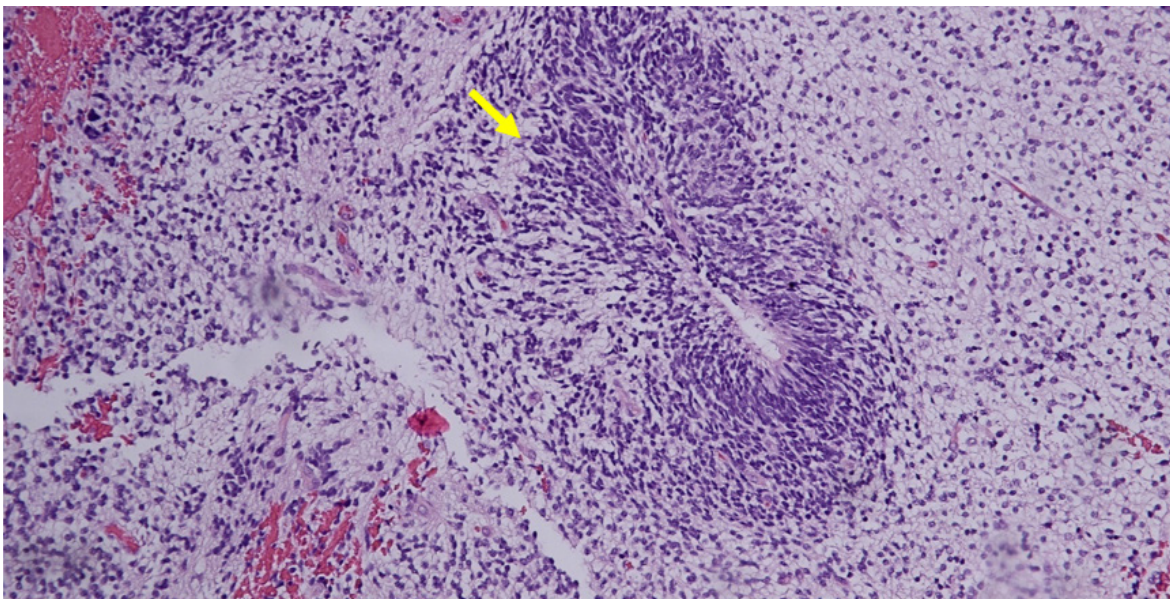


Figure 4 Immature neuroepithelium (yellow arrow)

The surgically resected specimen composed of a heterogeneous grayish-white mass with focal dark brown areas and soft to rubbery consistency, measuring 3.0x2.4x1.6 cm. The microscopic histological features were consistent with immature teratoma, characterized by the presence of multiple germ layers, including ectoderm (squamous epithelium), neuroectoderm (disorganized brain tissue), and endoderm (respiratory epithelium and mucus cells). Mesodermal components including vessels, smooth muscle, and skeletal muscle were also evident. The immature tissues, such as neuroectodermal tubules and rosettes forming primitive neural tubules, were demonstrated with high mitotic activity. No malignant elements or other germ cell tumors were identified.

Post-surgery, the patient had a good recovery without any complications. Currently, two years post-operation, he exhibits normal development and shows no signs of tumor recurrence based on both physical and radiological examinations.

Discussion

Congenital orbital teratoma is a rare condition characterized by a noncancerous tumor that contains tissues and structures normally found in other body parts. It is usually benign, can cause significant proptosis upon presentation. However, in most cases, the tumor tends to encase or adhere to the optic nerve, leading to irreversible ischemic damage². The prognosis of congenital orbital teratoma can vary depending on various factors, such as the size of the tumor, the extent of involvement of surrounding structures, and the age of the patient at diagnosis. Complete surgical excision is the pre-

ferred treatment, and adjuvant therapy is generally not necessary after the surgery. However, complications can arise from the surgical removal of the tumor, such as damage to surrounding structures like the eye or optic nerve, which can impact vision and other functions.

Early surgical intervention is crucial to save the vision of the affected eye, especially if the treatment is initiated soon after birth. The prognosis is usually favorable if complete surgical excision is achieved. Consistent follow-up with a multidisciplinary team of specialists, including ophthalmologists, oncologists, and surgeons, is important to monitor for any signs of recurrence or complications. The long-term prognosis for a child with congenital orbital teratoma can be promising with appropriate treatment and monitoring. In addition, regular follow-ups are essential to monitor for any recurrence or malignant transformation, which can occur despite successful surgery. It is important to address any recurrence promptly to ensure the best possible outcome.

In conclusion, we report a rare case of a neonate with a congenital orbital teratoma that was successfully treated with postnatal surgery.

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