

Mature teratoma presenting as orbital cellulitis in a 5-month-old baby

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Ann Saudi Med 2013; 33(6): 623-626

DOI: 10.5144/0256-4947.2013.623

We have presented an interesting case of an unexpected orbital tumor in a 5-month-old female healthy baby who presented initially with a 1-week history of acute left proptosis, swollen eyelid, conjunctival chemosis and exposure keratopathy. She was treated with intravenous antibiotics without any improvement and was referred for the drainage of the left orbital abscess. The procedure was performed with partial resolution, however the proptosis persisted and an underlying orbital pathologic lesion was suspected. Further exploring the left orbital revealed a partially cystic tumor. The histopathologic features were consistent with a mature teratoma. The case is presented with a brief discussion on this entity to attract the attention of general ophthalmologists to such tumors in the orbit.

Extragenital teratomas are thought to arise from misplaced, pluripotential, primordial germ cells. Childhood teratomas occur primarily in extragenital sites, mainly the spine, head, and neck while most adult teratomas are gonadal. The head and neck location is the second most common site (after the sacrococcygeal region), accounting for 5% to 14% of cases in infancy.^{1,2} The orbit is a rare site either primarily or secondarily as an intracranial extension. Females are approximately doubly affected compared to the males, and there is a propensity of the tumor to occur more frequently in the left orbit.³

CASE

A 5-month-old female healthy baby referred as a case of left orbital cellulitis for the drainage of an orbital abscess following treatment with intravenous antibiotics with no improvement. She had a history of sudden onset proptosis of 1-week duration. Examination showed that she was able to fix and follow objects with her right eye while fixing with limitation of movements on the left. There was left eye proptosis, severe eyelid swelling, conjunctival chemosis, and exposure keratopathy (Figure 1a). The fundus view was hazy. She was otherwise afebrile and healthy apart from anemia and mild leukocytosis. The B-scan showed a normal left globe and large diffuse orbital lesion of low reflec-

tivity pushing the globe forward. Magnetic resonance imaging (MRI) showed multicompartamental cystic collection with fluid signal intensity, slightly higher signal intensity on T1-weighted images, and marginal rim of enhancement occupying the left orbital cavity (Figure 1b) in addition to sinusitis. The initial clinical and radiological diagnosis was orbital cellulitis complicated by abscess formation. She was admitted for intravenous gentamicin and cefazolin with drainage of intraconal and extraconal abscess under general anesthesia. The patient was discharged home following initial improvement. Underlying orbital pathologic lesion was suspected on her follow-up and was readmitted two months later. Her systemic evaluation revealed persistent anemia. Her eye examination showed less left eye proptosis with conjunctival prolapse and faint corneal scar (Figure 1c). Her repeated MRI revealed left orbital extraconal and inferonasal multicompartamental cystic lesion with a marginal contrast enhancement. The radiologic diagnosis confirmed a possible hematic cyst.

Orbital exploration showed a huge mass with cystic component, which was excised. The histopathologic findings showed incomplete irregular cystic cavities lined by focally ciliated pseudostratified columnar epithelium with goblet cells representing the respiratory type of epithelium (Figure 2a), adjacent epidermal component with skin including skin appendages

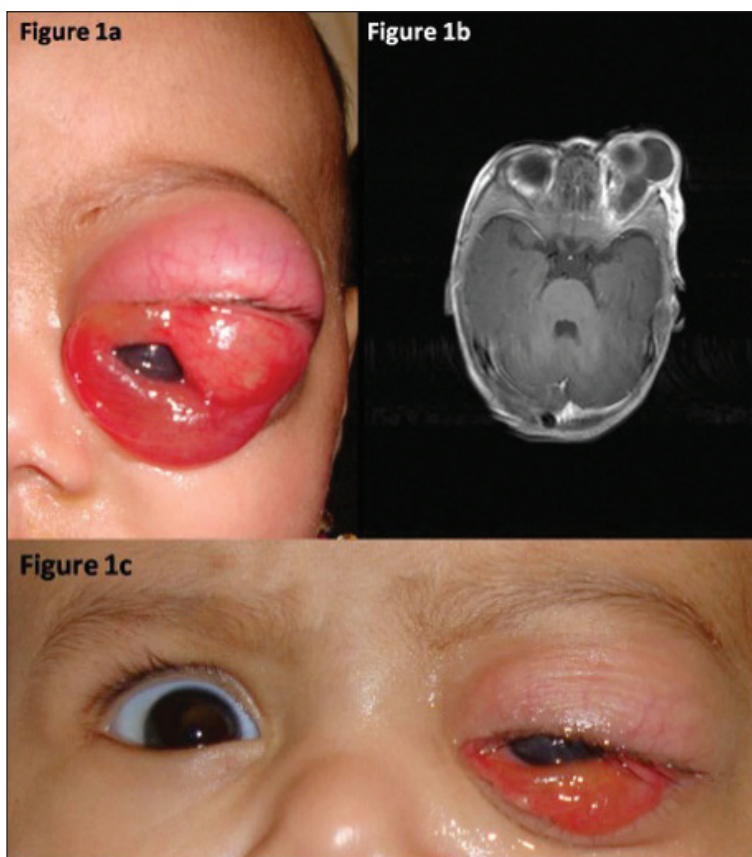


Figure 1. a) The clinical appearance of the marked left eyelid swelling, proptosis, and chemosis at initial presentation. b) Magnetic resonance image of the multicompartmental cystic mass within the left orbit showing ring of contrast enhancement. c) Partial improvement in the clinical appearance of the left eye following initial treatment for cellulitis and surgical drainage.

(Figure 2b), cartilage, adipose tissue (Figure 2c) and smooth muscle fibers. Several mucoserous, apocrine, and sweat glands were seen (Figure 2d).

The final diagnosis was a mature teratoma. The patient was followed up with no recurrence for a period of 16 months; however, her follow-up examination revealed left esotropia due to documented amblyopia in the left eye, which was managed by patching.

DISCUSSION

Extragonadal teratomas arise from pluripotential primordial germ cells that are thought to become misplaced, or have aberrant migration to several structures near the midline (such as in the head, mediastinum, or sacrococcygeal region). Another hypothesis suggests an origin from ordinary somatic cells with full genetic code. Theoretically, these cells can produce any other type of cell without being a "germ cell."¹ Holme reported the first case of orbital teratoma in 1862.⁴ Since then

several cases were reported.^{3,5-7} The review of published reports indicated the preponderance of left-sided orbital teratomas and a female-to-male ratio of 2:1.⁸ Our patient was a male with left orbital involvement.

Childhood teratomas occur primarily in extragonadal locations, and the majority of head and neck teratomas were identified during the first year of life or even at birth, particularly the ones affecting the orbit.^{1,2}

Orbital teratomas in contrast to the frequent orbital dermoid cysts are extremely rare.⁸ Orbital teratomas are usually benign and well differentiated; however, they may cause facial deformity due to rapid growth as well as destructive proptosis and exposure keratopathy.⁸ Proptosis at birth due to a congenital teratoma is unusual, but has been reported.^{7,9} Our patient presented with rapidly increasing proptosis over 1 week at the age of 5 months. An extension into the orbit from an adjacent space such as paranasal sinuses, cavernous sinus, or pterygopalatine fossa can also occur. In this case the presentation can be delayed and an associated airway obstruction may be encountered.¹⁰ Teratomas can extend into the periorbital sinus or through the superior orbital fissure, which can be further complicated by intracranial extension of the tumor.^{3,11} In addition, orbital teratomas can present with recurrent orbital cellulitis similar to our case, which might delay the diagnosis.¹²

Histopathologically, orbital teratomas are mostly benign and well differentiated. A total of 90% of teratomas contain all 3 embryonic layers: ectoderm, mesoderm, and neural tissue. Between 20% to 40% of childhood teratomas contain some immature tissues.^{1,2} Overall, neuroectodermal elements "both mature and immature" frequently dominate the childhood teratoma, particularly in the head and neck region compared with the sacrococcygeal teratomas. It was reported that up to 95% of childhood teratomas have some neuroectodermal elements.² Such teratomas may be confused with primitive neuroectodermal tumors (PNETs), since immature teratomas might include regions that resemble the developing neural tube or cells forming rosettes, similar to those seen in PNETs, neuroblastoma, and medulloblastoma.² Benign teratomas may contain mature tissues or smaller amounts (less than 50%) of immature elements.¹³ Immature neural tissue in a young-child teratoma does not reduce the prognosis; however, when they are found in an adult, they are usually associated with a malignant behavior and poor outcome.² Malignant extragonadal teratomas usually have carcinomatous features. They constitute a small number and tend to occur in older children. Few teratomas also show malignant degeneration over time.^{2,12}

Orbital teratomas are heterogenous complex le-

sions containing adipose tissue and occasional bone formation. Calcification can be dystrophic, but may also occur as mineralization of osteoid matrix (true bone), cartilage or well-formed teeth.^{12,13} Therefore, preoperative cross-sectional imaging is crucial, and computed tomography (CT) is required to define bony anatomy. Teratomas appear as multiloculated, cystic masses and solid areas within a well-delineated mass showing the areas of low attenuation and high signal intensity (representing lipid) on CT and MRI. This was observed in our case with slightly higher signal intensity on T1-weighted images.^{7,12,14} The differential diagnosis for an orbital mass with low attenuation includes vascular lesions, microphthalmia with cyst, coloboma and congenital cystic eye, epidermoid cyst, dermoid cyst, and meningoencephalocele.¹⁵

The clinical management of these lesions is unclear, due in part to their low incidence. Early surgery is now recommended aiming at complete surgical excision, thus saving the eye if possible. This is essential to avoid the risks of rapid growth resulting in necrosis, infection, hemorrhage, or spontaneous rupture.^{7,10,13,16} Orbital teratomas may rarely contain nongerm cell malignancies, such as neuroblastoma that has been reported, and in such cases, decision regarding adjunctive chemotherapy or radiotherapy should be based on the pathologic findings for the individual case.¹⁷ Teratomas with a malignant change are resistant to chemotherapy, and this explains the lack of initial response to chemotherapy and radiotherapy.¹³

The prognosis of teratoma is related to several factors including the age of the patient site, immaturity of the tumor, and extent of the teratoma.¹⁰ The prognosis is usually good; however, a complete surgical removal might be difficult and postoperative complications such as orbital asymmetry, ocular injury, and loss of vision can occur.^{7,13} Some reports have related the early surgical intervention to preservation of visual

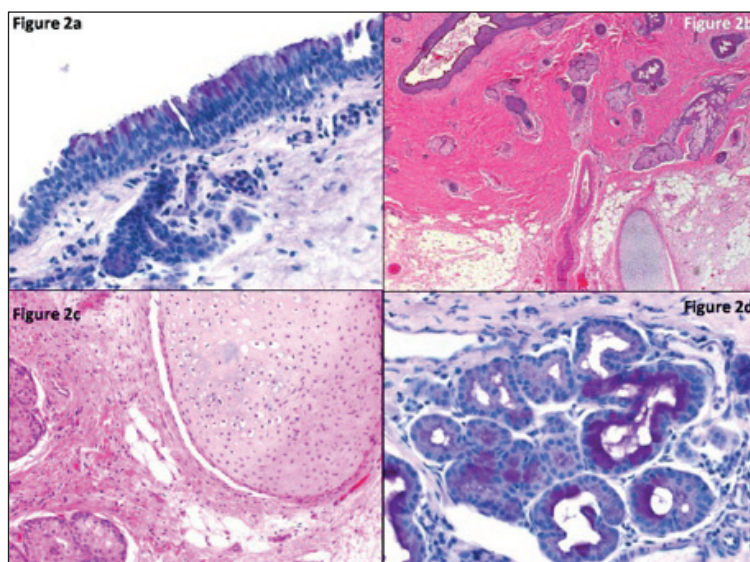


Figure 2. a) The lining of the cyst by respiratory type of epithelium (original magnification $\times 400$ periodic acid-Schiff). b) The epidermal component with adjacent fat and cartilage (original magnification $\times 50$ hematoxylin and eosin). c) Higher power appearance of the fat and cartilage within the teratoma (original magnification $\times 200$ hematoxylin and eosin). d) Glandular elements within the teratoma (original magnification $\times 400$ periodic acid-Schiff).

acuity.^{5,18} The eye-conserving approach might necessitate decompressing the tumor before dissecting it from the periorbital tissues if the tumor has a major cystic component.¹⁹

In conclusion, ophthalmologists should be aware of the occurrence of orbital teratomas that have variable heterogenous histologic characteristics by their nature. Both the pathologic and radiologic diagnosis of a teratoma may be suspected when a complex lesion contains fatty regions and calcifications. Early diagnosis is helpful to aim at a complete surgical excision of the lesion and to avoid the disadvantages of partial resection such as regrowth of the tumor and the potential for a malignant change at a later date.^{10,20}

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