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Rare orbital cystic lesions in children

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ABSTRACT

Purpose: To analyze the clinical features, imaging findings and surgical management of rare orbital cystic lesions in children.
Materials and methods: Clinical records of 5 Chinese children with rare orbital cystic lesions including cystic teratoma, congenital cystic eye, optic nerve sheath cyst, parasitic cyst, and meningoencephalocele were reviewed. Their clinical history, symptoms and signs, ultrasonography or computed tomography/magnetic resonance imaging (CT/MRI), surgical management were presented in detail.
Results: Among the 5 patients, 2 were male and 3 female. The right orbit was involved in 2 patients and the left orbit in 3 patients. Ages ranged from 2 months to 11 years (mean, 4.2 years). Cystic teratoma showed a well-outlined cystic mass with a focal bone-like structure (or calcific densities) in their lumens as revealed with CT scan. Congenital cystic eye was a rare ocular malformation that existed at birth and showed a cystic lesion with no definite ocular structures in the orbit. With MRI examination, the optic nerve sheath cyst demonstrated a clear figure of the central optic nerve and the surrounding sheath cyst. Patients with parasitic cysts usually had frequent animal contact and high levels of blood eosinophils. Meningoencephalocele, the herniation into the orbit of brain tissue, had the typical CT/MRI features, with bone defect and soft tissue mass in the orbit and a homogenous appearance that was isodense with brain.
Conclusions: Besides common dermoid/epidermoid cyst, rare orbital cystic lesions such as cystic teratoma, congenital cystic eye, optic nerve sheath cyst, parasitic cyst, and meningoencephalocele should be considered in children.

1. Introduction

Cystic lesions of the orbit are common orbital diseases, with an incidence of 8.3–12.1% (Henderson et al., 1994; Rootman, 2003). Usually, cystic lesions are grouped as those of epithelial versus nonepithelial origins (Rootman, 2003). Common epithelial cysts include dermoid and epidermoid cysts, mucocele, and implantation cysts, whereas typical nonepithelial cysts include hematoma, neurogenic cysts, and infectious cysts. In most instances, it is easy to differentiate a cystic lesion from other orbital masses based on both clinical features and imaging (Shields and Shields, 2004). However, in children, some cystic conditions of the orbit can be quite rare and are often misdiagnosed, leading to improper management. The purpose of this article is to report 5 cases of Chinese patients with rare orbital cystic lesions in childhood and to highlight their clinical, radiological, and pathological findings as well as the results of their surgical outcomes.

2. Materials and methods

A retrospective review was performed on cases of Chinese patients with a diagnosis of orbital cystic lesions, who were treated at the Zhongshan Ophthalmic Center, Sun Yat-sen University, Guangzhou, China between January 1, 2000, and December 31, 2009. The ethics committee of the Zhongshan Ophthalmic Center approved this retrospective study, and this study was conducted according to the principles expressed in the Declaration of Helsinki. The committee specifically waived the need for consent. The subjects involved (or their legal guardians) reviewed this article in manuscript form, along with the figures, and provided written consent for publication. Clinical, operative, and pathological records were reviewed. Among the inclusion criteria were that patients were <14 years of age, were treated by a single surgeon (J.H.Y.), underwent a systemic evaluation by a pediatric oncologist,

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Table 1
Clinical Data on five Patients with rare orbital cystic lesions in children.

Case no	Age/sex/eye/ diagnosis	Symptoms and signs	Imaging findings	Surgical management	Histologic examination	Final outcomes
1	4 months/F/R/ orbital teratoma	4 mm proptosis and tearing; entropion of lower eyelid; downward and inward displacement of the eye.	Ultrasonography and CT: a circumscribed, 15.4 × 26.6 mm cystic mass with several patch-like bone structures in right superior-temporal orbit.	A well-defined 25 × 15 × 10 mm cyst that merged with bone mass and four teeth, were totally excised through anterior orbitotomy.	An admixture of bone, epithelioid cells, fibrotic tissues and neural tissue. The tissues were mature without mitotic activity.	Cure, no recurrence 1 year postoperatively.
2	6 months/F/L/ congenital cystic eye	Ptosis and protrusion of left upper eyelid. A large well-defined soft mass (3.5 × 2.0 cm) was palpable through upper eyelid.	Ultrasonography and MRI: a well-delineated, cystic mass (3.45 × 3.14 cm) in left orbit, without any eyeball structures.	An anterior orbitotomy was performed. A cyst (3.0 × 2.0 cm) was removed.	A cystic lesion lined by a layer of connective tissue externally and a layer of neuroglial tissue internally.	Ptosis remained at 2 years after surgery.
3	9 years/M/L/ meningocele of the optic sheath	Visual acuity in left eye was no light perception. The optic disk of left eye was pale.	Ultrasonography and MRI: a well-delineated, hypoechoic cystic mass (1.89 × 1.54 cm) surrounding the optic nerve.	A cystic mass was identified originating from the optic nerve. The cyst and optic nerve were removed by lateral orbitotomy	A reactive proliferation of meningothelial cells and atrophy of the optic nerve.	No radiographic abnormality at 1-year follow-up.
4	11 years/F/L/ intraorbital hydatid cyst	A lump with slight pain at the orbit for 1 year. An immobile, tender, poorly-defined mass (1.0 × 1.0 cm) was palpable.	Ultrasound and MRI: an ill-defined cyst (1.89 × 1.54 cm) at the left superior-medial orbit, with a high internal echo within the cyst.	A poorly-defined cystic mass and a white live parasite (3.0 × 0.3 × 0.1 cm) were removed through anterior orbitotomy.	Confirmed the diagnosis of a primary intraorbital hydatid cyst.	No recur at 2 years after surgery.
5	2 months/M/R/ ancephalocele and ectopic brain tissue of the right side	Proptosis and a smooth, moderate consistency, unmovable mass (10 × 10 mm) in superior-medial orbit since birth.	Ultrasound and CT: a well-demarcated, homogenous “mass” which communicated with brain tissue. There was a bony defect between right orbit and cranial structures.	No	No	Referred to the department of neurosurgery for treatment.