Congenital Sudoriferous Cyst within the Orbit Followed by Esotropia

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***Purpose*:** To report a case of congenital sudoriferous cyst of the orbit with esotropia.
***Methods*:** A 20-day-old male, born prematurely presented with a palpable lump on left upper lid. Orbital ultrasonography including color doppler image and orbital magnetic resonance image were performed to evaluate the lid lesion. The mass was excised and histologically examined. Complete ocular examination including visual acuity, duction, version, and the presence of strabismus were performed.
***Results*:** A well circumscribed round cystic mass, measuring 1.4×1.3 cm was noted at medial superior aspect of the left orbit. It compressed and displaced the left globe to inferior posterior position with intact optic nerve. Histopathologic examination showed the lesion to be a solitary sudoriferous cyst lined by two layers of cuboidal epithelial cells with eosinophilic cytoplasm. After the excision of the mass, limitations of extraocular muscle movements, esotropia, and amblyopia were noted.
***Conclusions*:** If an orbital cyst affects the globe or extraocular muscles, it should be excised as soon as possible to prevent strabismus and amblyopia especially in infant.
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Key Words: Amblyopia, Esotropia, Sudoriferous cyst

Sudoriferous cysts are fairly uncommon in the eyelid, where a cyst drives from the apocrine glands of Moll.[1] Shields and Shields have described that this type of cyst typically occurs at the lid margin, but rarely appears within the orbit.[1] Other previous literatures reported similar sites of lesion such as beneath the lids, canthal area, inferior orbital rim and floor.[2-8] This tumor is rare in adults and even more rare for children. A sudoriferous cyst that appears as a congenital tumor is certainly most unusual.[2]

Although there are rare case reports of sudoriferous cysts in the orbit of children, this case report is the first to describe a congenital sudoriferous cyst of the orbit followed by esotropia.

Case report

A 20-day-old male, born prematurely, with a palpable round lump on the upper left eyelid was presented to us for consultation by the department of pediatrics. The upper eyelid was closed due to the mass and could not be opened nor everted. There was no change in the color or the size when the patient's head was placed in a dependent position or when the infant cried (Fig. 1A, B). His mother was in good general health and had not taken any medications during pregnancy. Examination of the infant showed no other congenital malformations.

Ultrasonography (US) revealed a 1.1×1.3 cm round anechogenic cyst in the medial portion of the left orbit. It displaced the left eyeball to the posterior lateral portion. (Fig. 1C). Magnetic resonance images (MRI) of the orbits revealed a 1.4×1.3 cm round cystic mass with a low signal intensity in T1WI and a high signal intensity in T2WI. The left eyeball was compressed and displaced to the inferior-posterior portion of the eye socket by the mass. Although the mass was bound by the intracranial and nasal cavities, it was observed that the mild stimulation of the left optic nerve caused slight inferior displacement of the medial rectus muscle (Fig. 1D).

Since there was visual axis encroachment in conjunction with eyeball displacement, surgical intervention was scheduled. The mass was excised via an approach through the skin. It was found to be attached with two stalks to the upper eyelid where it was carefully dissected and removed (Fig. 2A). The specimen was a grayish polypoid mass with fibrous band-like stalks and filled with a clear fluid. Histopathologically, the specimen consisted of a solitary cyst with a very thin cystic wall (Fig. 2B). Microscopically, it was lined by a double layer of non-keratinizing cuboidal and/or

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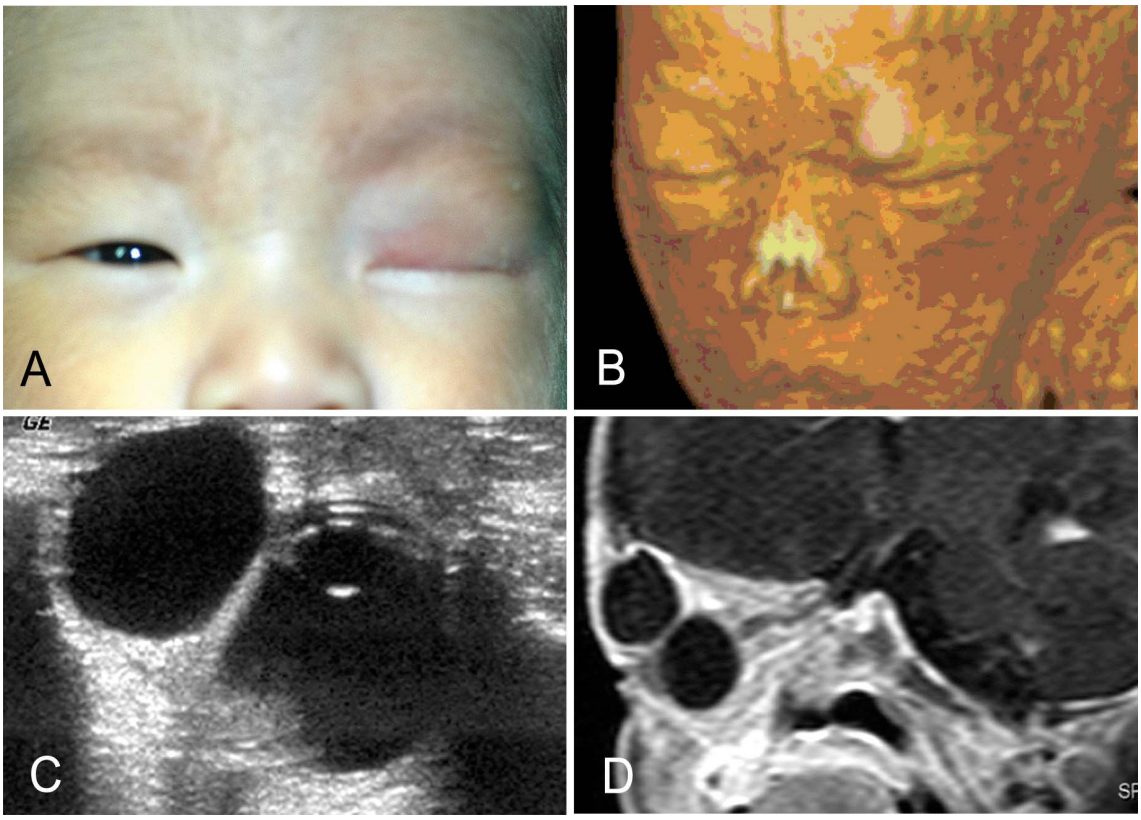

Fig. 1. (A) A round palpable pinkish lump on left upper lid with limitation of eye opening due to the mass. (B) Three dimensional reconstruction of orbit MRI. (C) An orbit ultrasonography shows well circumscribed, round, anechogenic cyst (left) and the left globe is displaced to posterior lateral portion and compressed by the mass (right). (D) Orbit MRI. A 1.4×1.3 ㎝ sized circumscribed, round, cystic mass in medial superior aspect in left orbit. Eye ball is displaced to the inferior posterior portion with intact optic nerve and the mass shows low signal intensity on Gd-DTPA enhancement sagittal T1WI.

flattened epithelium, with features identical to the glands of Moll. The innermost cells displayed a granular and distinctly eosinophilic cytoplasm with apical expansions, while the outer wall was composed of a fibrous tissue. The diagnosis of a sudoriferous cyst was thus confirmed (Fig. 2C).

The patient underwent a comprehensive ocular examination one week after the operation. Since there appeared to be little fixation or following efforts towards the mother's face or a hand at near distances with the left eye, the patient was prescribed 30 minutes of occlusion by patch per day. Esotropia was then able to be diagnosed. The angle of deviation was 30 prism diopters (PD) by the Krimsky method. Even with intensive occlusion therapy, examination at four months showed no interval change for fixation and an abduction limitation on left eye was found. Seven months after the operation, occlusion therapy was ceased because of a newly developed poor response to visual stimulation on the right eye, probably due to occlusion time in excess of the prescribed treatment. The angle of deviation was then found to be 50 PD by an alternative prism cover test at a near distance. US revealed no abnormal echogenecity in the left orbital area and no movement during scanning on the left eyeball which was deviated medially (Fig. 3A). Nine months after the operation, 30 PD esotropia was determined by the Krimsky method and an abduction limitation was found in both eyes (Fig. 3B). Computed tomography (CT) of the orbits revealed no eyeball deformity with normal extra-ocular muscle structure and normal peri-orbital tissue (Fig. 3C).

Discussion

Sudoriferous cysts, also known as apocrine gland cysts and apocrine hydrocystomas are derived from the sweat glands of Moll. They probably result from the obstruction of excretory ducts with the retention of clear fluid.[1,3] They are divided into two types, apocrine and eccrine, and represent 4~5% of all eyelid tumors in adults and 1% in children, but are less common in the orbit itself. Symptoms appears as fluctuant swelling beneath the eyelid or in the canthal area.[1,3,4-7]

Shields and Shields[1] reported that it is probably not possible to differentiate a small cystic lesion without bone involvement from other orbit cysts even with a CT or an MRI. However, Rosen and Li[8] reported sudoriferous cysts in adults and associated bony remodeling of the inferior orbit

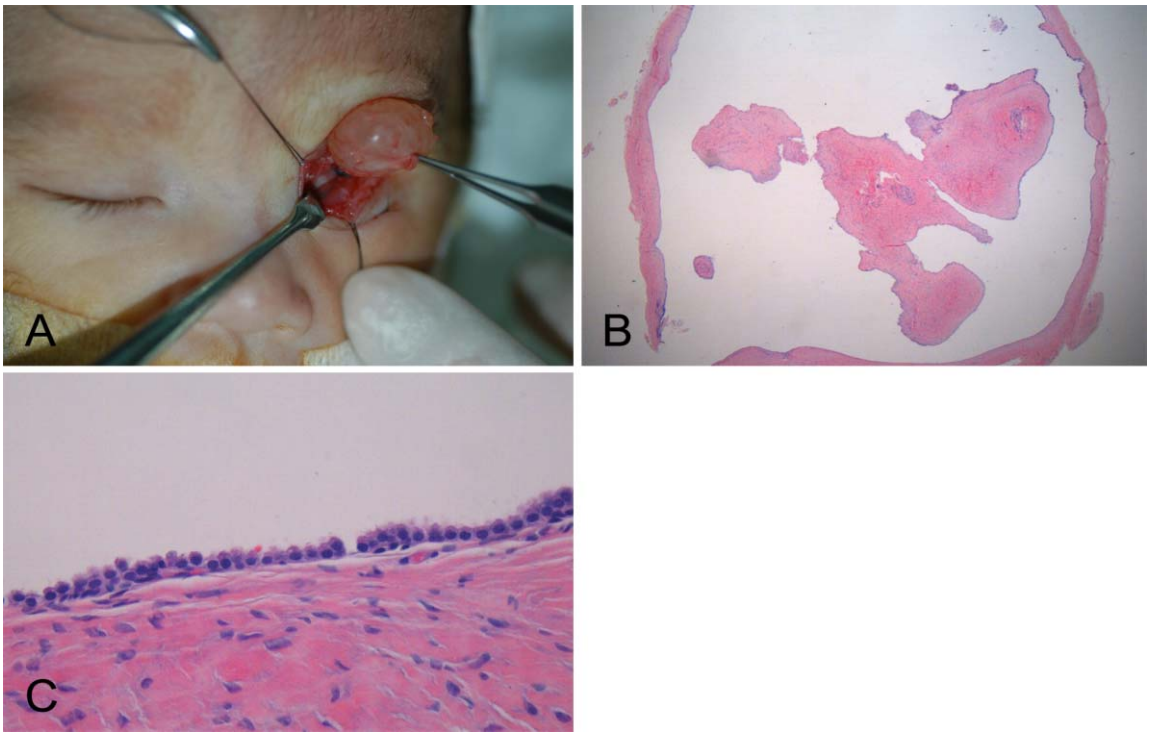

Fig. 2. (A) Intraoperative finding of cyst excision shows a gray white cystic mass is attached to upper palpebral conjunctiva with two fibrous pedicles. (B) A cross section of the cyst filled with cystic fluid shows an unilocular cystic mass which consists of a thin fibrous wall (×14). Hematoxylin and Eosin stain. (C) The luminal surface is lined by two layers of atrophic, non-keratinizing cuboidal or flattened cells; the innermost cells show apical expansions, granular, deeply eosinophilic cytoplasm and basally located round nucleus (×400).

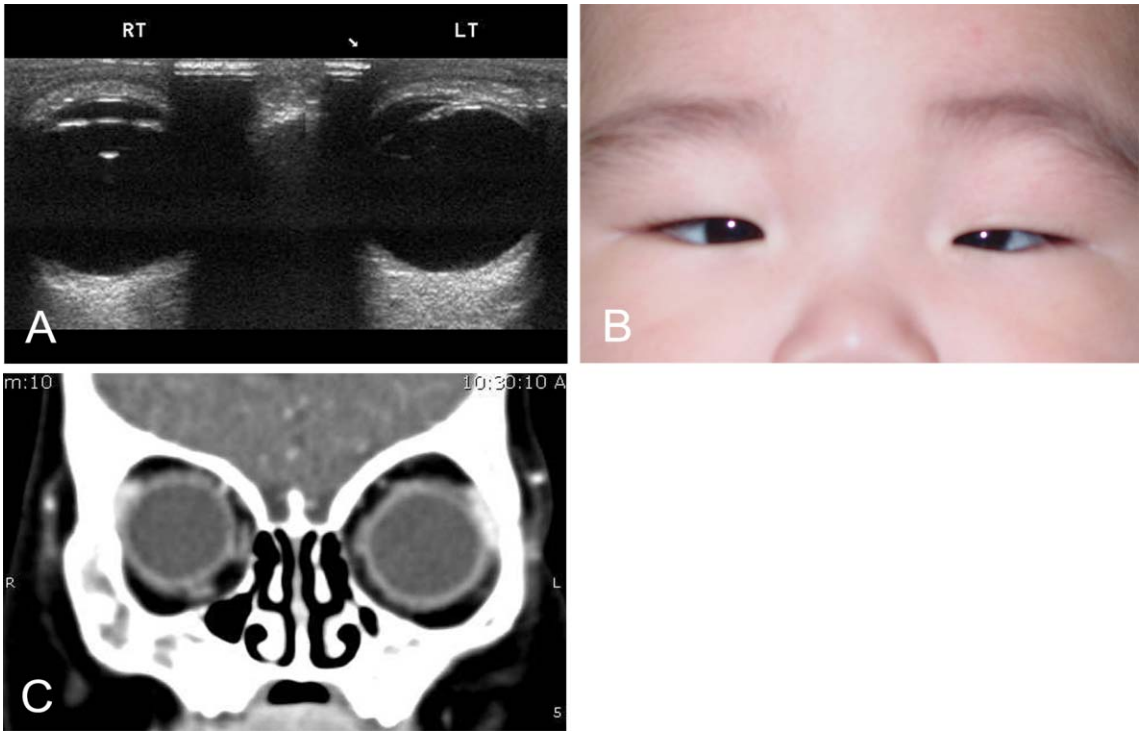

Fig. 3. (A) There is no focal abnormal echogenicity in the left eye, which is rotated medially without movement during examination. (B) Post-operative photograph showing esodeviation. 30 PD esotropia was determined by the Krimsky method and an abduction limitation was found in both eyes. (C) Computed tomography scan reveal no eyeball deformity and normal extraocular muscles.

rim and floor by use of CT and MRI. In this case, the CT and the MRI showed that the cyst compressed the eyeball and displaced the globe, and allowed for the rectus muscle to be found. We believe that the CT and the MRI were helpful to evaluate the status between the cyst mass and the healthy surrounding tissues and aided in making decisions about the time to operate although neither the CT nor the MRI was helpful for the differential diagnosis.

The management of apocrine gland cysts is usually a simple observation if the lesion is small and asymptomatic, but occasionally requires complete local excision if the lesion is symptomatic or cosmetically unacceptable.[1] In past reports, Saunders[4] had excised a congenital sudoriferous cyst from an 18 months old and Mims et al[3] have operated on a 24 month old. In these previous reported cases, there were no complications, such as encroachment upon the visual axis, an increase in the size, or inflammation. On the other hand, Rosen and Li[8] had excised the mass and reconstructed the orbital rim with a calvarial bone graft in a 45-year-old woman who presented with a painless lump in her left lower eyelid.

The histopathology of this report lesion had features typical of a sudoriferous cyst. Grossly, it had a thin wall and contained clear fluid. Microscopically, the lesion was a solitary cyst lined by two layers of non-keratinizing cuboidal epithelial cells with apical snouts on the luminal side of the cell. The cyst was periodic acid-Schiff (PAS)-positive, with diastase-resistant granules on their apical surface, similar to what has been previous reported.[8,9]

In this case, the radiologic appearance, the clinical features, and the histopathologic results of the cyst were compatible with the diagnosis of a sudoriferous cyst. We recommend that if a congenital orbital cyst affects the globe or the extra-ocular muscles, one should excise the mass as early as possible and regularly follow up and evaluate for strabismus and amblyopia.

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