Giant eyelid eccrine hidrocystoma-induced progressive ptosis in childhood

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An upper lid eccrine hidrocystoma presenting as early childhood progressive ptosis is very rare. We present a 9-year-old female child with droopy right upper lid since birth and progressive increase in symptoms. She had right upper lid ptosis (marginal reflex distance 1 of −1 mm) with fair levator function (8 mm) and abnormal cystic change on the conjunctival side. Computerized tomography imaging delineated the well-defined cystic lesion with homogeneous cavity with no contrast enhancement. Following the cyst excision, a giant eccrine hidrocystoma measuring 25 mm × 15 mm was removed, the largest reported in pediatric eyes. The case demonstrates the possibility of giant lid eccrine hidrocystomas presenting as progressive ptosis at a pediatric age and the need for early surgical intervention to prevent amblyopia.

Key words: Childhood acquired ptosis, eccrine hidrocystoma, giant hidrocystoma, progressive ptosis

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Cystadenomas or hidrocystomas arising from eccrine glands are benign cystic or nodular lesion often seen in the periorbital skin.[1,2] It is rarely multiple and may be associated with Goltz syndrome or ectodermal dysplasia.[3] Commonly, the periocular or lid hidrocystomas are small to moderate in size, about 5 mm and rarely larger. Although giant "orbital" eccrine hidrocystoma is known to occur in early infancy, the "eyelid" is a very rare site in children.[1,2,4-7] We present a giant eccrine hidrocystoma of upper lid presenting in a child as progressive ptosis.

Case Report

A 9-year-old female child presented to our Oculoplasty Clinic with painless, progressive drooping right eye (OD). Her parents had noticed the drooping since birth, and there had been significant increase in the past 2 years. On ocular examination, her unaided visual acuity was 20/30 in OD and 20/20 in the left eye, respectively. Right upper lid ptosis was observed with marginal reflex distance (MRD 1) of −1 mm with significant reduction in vertical fissure height (5 mm) [Fig. 1]. The MRD in the fellow eye was 4 mm. Abnormal cystic change was detectable on the conjunctival side of the upper lid [Fig. 1]. However, no discrete mass was palpable. A fair levator muscle action (8 mm) in the right eye and good Bell's phenomenon was elicited. The extraocular movements were normal and the eyes were orthophoric. Her dilated fundus examination and intraocular pressure were within normal limits in both eyes. B-scan ultrasound examination of upper lid demonstrated fluid-filled cavity with no internal echoes [Fig. 2]. Computerized tomography showed well-defined cystic mass with four regular margins measuring 21 mm × 19 mm × 6 mm with no contrast enhancement [Fig. 2].

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Upper lid mass excision was performed [Fig. 3], and the histopathology confirmed and showed 25 mm × 15 mm cystic mass [Fig. 4] lined by double-walled (1 mm thick) cuboidal epithelium as eccrine hidrocystoma. The cyst cavity was filled with clear fluid. The postoperative period was uneventful with mild lid edema and conjunctival congestion that resolved spontaneously. There was significant improvement in the MRD (3 mm) after excision. At 1-month postoperative follow-up, the MRD was stable at 3 mm with no lid edema or recurrence at 6 months [Fig. 5].

Discussion

Sweat glands can be eccrine (open in epidermis) or apocrine (open through follicle) in type. Hidrocystomas are benign cystic masses that emanate from an obstructed sweat gland duct with subsequent fluid retention.[1-10] Moreover, it is reported to constitute only 1% of childhood eyelid tumors.[4] Although giant orbital eccrine hidrocystomas have been observed in pediatric age, isolated giant eyelid eccrine hidrocystomas are less common. Orbital eccrine hidrocystoma in two pediatric cases has been reported by Malihi *et al*.[1] Of the two, one was an infant presenting 2 weeks after birth with orbital eccrine hidrocystoma. Chung *et al*. reported a 20-day-old child with dystopia and complete lid closure following a congenital superior orbital hidrocystoma.[7] Congenital hidrocystomas are often apocrine and present commonly as orbital mass and rarely as ptosis.[2] Eshrahi reported orbital hidrocystoma of apocrine type presenting at birth as proptosis.[10] Sheth and Raina in their report has shown ptosis in adult-onset giant eccrine hidrocystoma presenting along with epiphora.[6] Similarly, Rodallec *et al*. reported adult-onset hidrocystoma with progressive ptosis.[5] Mukherjee *et al*. reported giant apocrine hidrocystoma presenting with ptosis and dystopia in an adult.[8] Ferraz *et al*. showed isolated ptosis in adult-onset orbital hidrocystoma which presented as occult in two cases and as subcutaneous mass in one.[11] Eccrine hidrocystoma of the eyelid as studied by Singh *et al*. has been noted to have a median size of 1 mm and majority are adult-onset (mean age 59 years).[12] In our literature search (PubMed and Google search using keywords), we noted no clinical case report that showed progressive ptosis in a child as the presenting complaint in giant eyelid eccrine hidrocystoma. Although atypical presentations of hidrocystoma, where an apocrine hidrocystoma presented as a lacrimal gland mass with mechanical ptosis in adults, have been reported, those of an eccrine nature are very few.[8]

While favored management for an asymptomatic small hidrocystoma in children is only observation, surgical excision is advocated in cases with vision-threatening symptoms or unacceptable cosmetic appearance. Unlike orbital cysts, which are frequently symptomatic at the beginning, eyelid lesion may be asymptomatic until it grows in size to induce symptoms such as ptosis. Progressive ptosis has probably induced the drop of one line in the affected eye due to sensory deprivation and pressure. Although large or giant eccrine hidrocystomas are known to originate in orbit, it is rare to notice the giant eccrine mass on the eyelid, and as a matter of fact, this is the largest eccrine eyelid hidrocystoma (25 mm × 15 mm) removed from the pediatric age group.

Conclusion

Eccrine hidrocystoma can be considered one of the differential diagnoses in progressive ptosis in the pediatric age group and timely imaging can aid diagnosis. Early interventions in such symptomatic cases also prevent amblyopia due to mechanical ptosis.

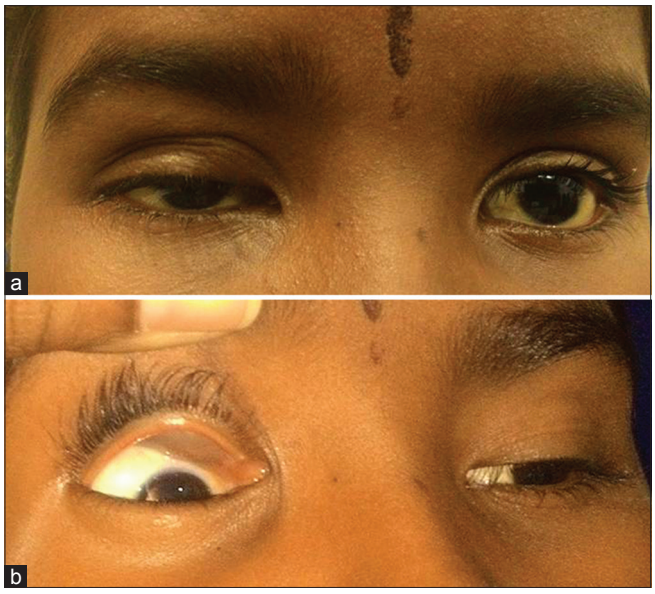

Figure 1: Preoperative clinical photograph showing the ptosis (a) and cystic conjunctival surface (b)

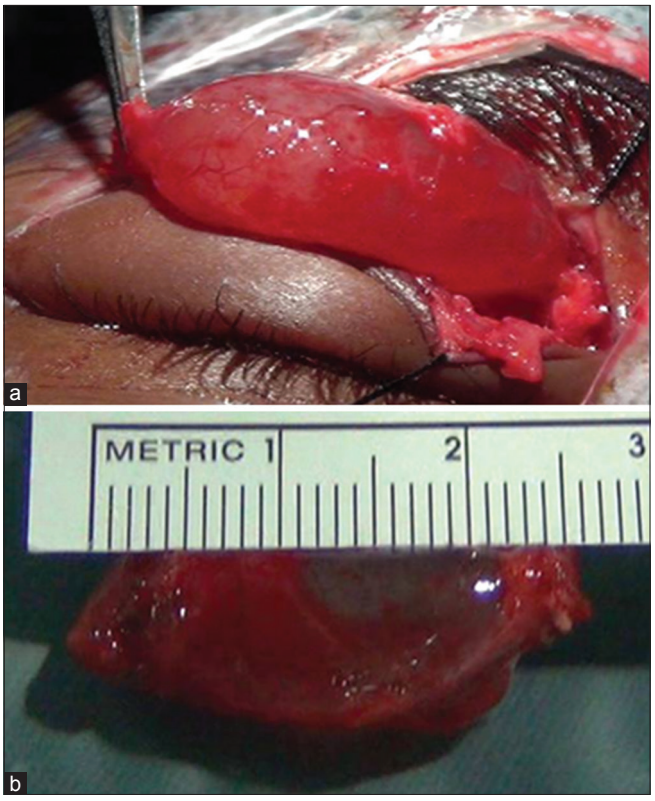

Figure 2: Intraoperative picture of the giant eccrine cystic mass (a and b) measuring 25 mm × 15 mm

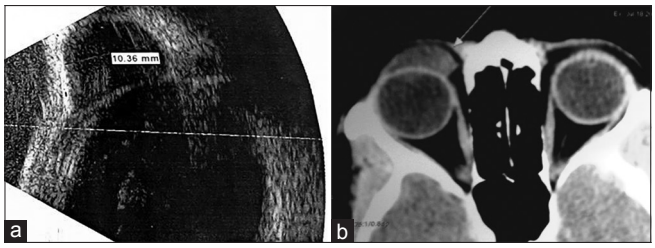

Figure 3: Ultrasound B-scan (a) and contrast-enhanced computerized tomography scan (b) showing cystic lesion of the upper lid

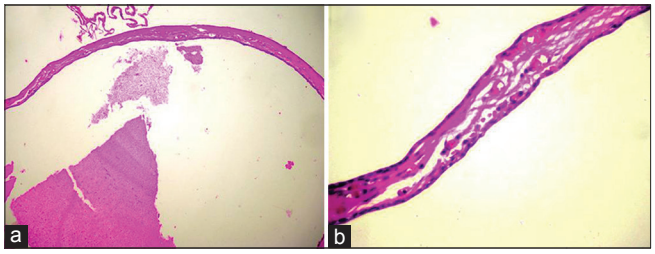

Figure 4: Histopathology showing the double-walled cyst lined by cuboidal epithelium. (a) (H and E, ×40) (b) (H and E, ×400)

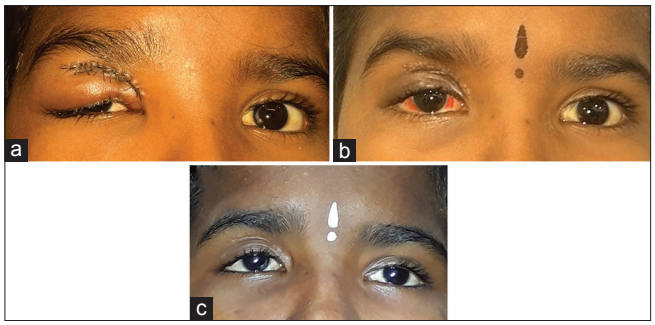

Figure 5: Clinical photograph showing immediate day 1 (a), 1-month (b), and 6 months (c) postoperative period

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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