

Case report

Congenital giant orbital eccrine hidrocystoma

Raghad Al Rasheed (MBBS)^a, Mazen AlSamnan (MD, FRCS)^{b,*}^a Division of Ophthalmology, Department of Surgery, National Guard Health Affairs, Riyadh, Saudi Arabia^b Ophthalmology Department, Security Forces Hospital, Riyadh, Saudi Arabia

ARTICLE INFO

Keywords:

Orbital cystic mass
Eccrine
Hidrocystoma

ABSTRACT

Purpose: To describe the clinical presentation, management, and histopathological results of a congenital eccrine hidrocystoma.**Observations:** A 4-year-old healthy boy presented with left upper eyelid ptosis and superonasal painless mass since birth. Computed tomography showed a hypodense cystic lesion located in the extraconal orbital compartment medially. Following complete excision, histopathology revealed an eccrine hidrocystoma.**Conclusions and Importance:** Orbital eccrine hidrocystomas should be considered in the differential diagnosis of orbital cystic masses in the pediatric age group.

1. Introduction

Hidrocystomas are benign cutaneous cystic lesions that arise from eccrine or apocrine sweat glands that are located in the periocular area.¹ They account for 5% of all eyelid tumors in adults and 1% in children, and it rarely arises in the orbital cavity.^{2,3} As for eccrine hidrocystomas, they are usually located in the head/neck region and can manifest as a single or multiple small cyst. However, giant lesions situated in the orbital area are rarely reported, and their presentation is predominant in young infants.^{1,2,4} This report describes the clinical presentation, management, and histopathological results of a congenital giant orbital eccrine hidrocystoma. Despite its scarce presentation, orbital eccrine hidrocystoma should be in the differential diagnosis of orbital cysts in all age groups. Early management in the pediatric group is of significant concern to prevent amblyopia.

2. Case report

A 4-year-old boy presented with left upper eyelid ptosis and superonasal mass since birth. He is a healthy full-term boy with normal delivery and no significant past medical history. The parents denied any complaints of pain or change in the size of the mass. On examination, uncorrected visual acuity was 20/30 right eye, 20/70 left eye. Mechanical left upper eyelid ptosis was noted to be secondary to a

superonasal swelling (Fig. 1A). No skin changes were noted over the area. The mass was non-pulsatile, non-tender, soft, freely mobile with no attachment to adjacent tissue. No discharge or tear reflux was noted on digital palpation. Both pupils were reactive to light with no afferent pupillary defect. Extraocular motility was full at all gazes without any restrictions. Slit lamp and fundus examinations were normal. Autorefractometry was as follows: +0.75–1.00 × 173 for the right eye compared to a high astigmatic error in the left eye: –8.75–8.75 × 31. B-scan ultrasound demonstrated a fluid-filled cavity with no internal echoes (Fig. 2). Computed tomography (CT) revealed a well-defined, hypodense cystic lesion located in the extraconal orbital compartment medially. Also, there was a mild compression of the left orbit and minimal remodeling to the adjacent bone (Fig. 3). The patient underwent anterior orbitotomy through lid crease incision to excise the mass (Fig. 4). The cyst was excised intact and gross examination of the mass revealed a delicate smooth wall, filled with clear fluid. Histopathologic evaluation revealed a 1.5 × 2.2 × 1.9 cm cystic lesion with a clear cavity lined by a smooth surface of a double layer of cuboidal cells. No apical snouts were identified which is consistent with the diagnosis of eccrine hidrocystoma. (Fig. 5).

At 1-month follow up visit, the patient was asymptomatic with no signs of recurrence (Fig. 1B). Visual acuity improved to 20/50 with a noticeable improvement in astigmatic error in the left eye tested by autorefractometry: –4.25–2.25 × 10.

* Corresponding author. Department of Ophthalmology, Security Forces Hospital, P.O. Box 3543, Riyadh, 11480, Saudi Arabia.
E-mail address: mazensamnan@gmail.com (M. AlSamnan).



Fig. 1. An external photograph showing left upper eyelid superonasal mass causing mechanical ptosis. (A); Post-operative appearance at 1-month follow-up visit. (B).

3. Discussion

Hydrocystoma can originate from eccrine or apocrine sweat glands. It accounts for 5% of all eyelid tumors in adults and 1% in children.⁵ Both genders can experience solitary hydrocystoma, whereas multiple lesions usually show a marked female propensity.⁶ Eccrine hydrocystomas are ductal retention cysts of obstructed sweat glands that cluster in the periocular area not involving the lid margin.⁵ On the other hand, the apocrine subtype develops from a cystic proliferation of the secretory glands of moll located in the eyelid margin.⁷ Epithelial cell sequestration at the embryonic stage could give rise to such congenital lesions.⁴ For acquired cases, theories suggest epithelial cell implantation into the orbit secondary to trauma or orbital surgeries.⁸

Due to similarities in the clinical picture, a histopathology study is vital in differentiating both subtypes. Eccrine hydrocystomas constitute of unilocular cysts lined with one or two layers of cuboidal cells.⁵ While the apocrine type characterized by papillary projections growing into

the cystic cavity with decapitation secretion from the apocrine cells.⁷ Both subtypes should be differentiated from other cystic lesions, including mucoid cysts, inclusion cysts, basal cell carcinomas, lymphangioma, and hemangioma.

With a large congenital nasal orbital mass, ruling out a meningoencephalocele is in consideration with this case, where bony defects are better visualized by CT. A major advantage of CT is its ability to image bone, soft tissue and blood vessels all at the same time. Ultrasound with doppler evaluation of the orbit obtains dynamic information, is readily available in many ophthalmic centers, and does not expose patients to radiation. For that, we elected to choose CT along with orbital ultrasound to determine the consistency of the mass, the extension, and to plan the surgical approach for our case. However, a histopathological biopsy remains the mainstay confirmatory test for diagnosis.

A thorough review of the reported cases showed a total of 10 cases of periocular eccrine hydrocystoma; only 5 were orbital in origin (Table 1). In addition to the present case, to the best of our knowledge, only two cases of congenital eccrine hydrocystoma have been reported so far^{1,9} (Table 1). Malihi et al.¹ operated on a 2-month-old boy with an orbital eccrine hydrocystoma presented with a bluish nodular non-tender subconjunctival swelling since birth. Also, they reported another case as an idiopathic orbital eccrine hydrocystoma in an 8-year-old boy presented with proptosis and limitation in abduction.¹ Eslami et al.¹⁰ reported a 14-year-old girl with a painless medial canthal cyst for 3 months which was diagnosed as idiopathic orbital eccrine hydrocystoma. A giant lesion has been previously reported in the superior part of the orbit causing inferior dystopia and posterior eyelid lamellar inversion.¹¹

Careful excision with complete removal of the cyst wall is advocated for large hydrocystomas to prevent the regrowth from remnant parts.^{12,13} On the other hand, small asymptomatic hydrocystomas can be observed though spontaneous resolution is rare.¹¹ For the pediatric-age group, prompt management is essential to prevent amblyopia. It can develop directly from occlusion of the visual axis, or indirectly from the development of astigmatism secondary to mass effect. Robb studied

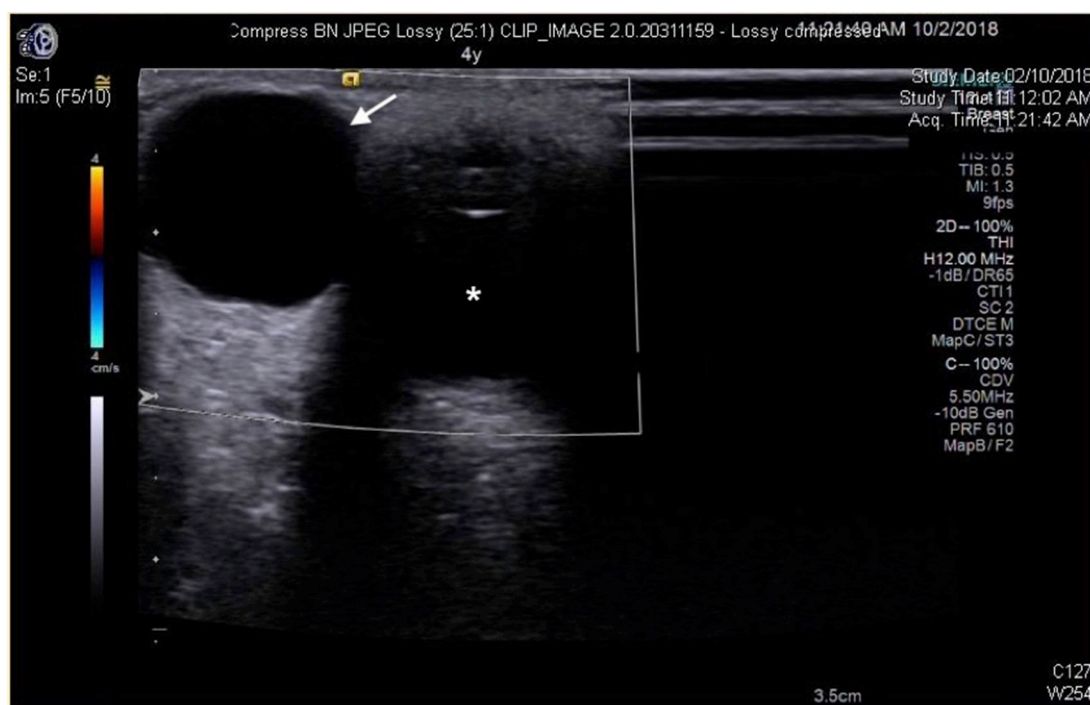


Fig. 2. B-scan ultrasound demonstrate: anechogenic fluid-filled cavity (arrow), normal composition of left globe (asterisk).



Fig. 3. A computed tomography scan of the orbit showing a well-defined, hypodense cystic lesion located in the medial wall of the left orbit.

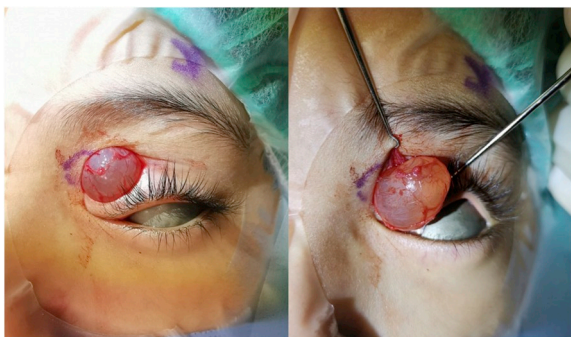


Fig. 4. An intraoperative photograph demonstrating surgical excision of the cystic mass through anterior orbitotomy through an upper eyelid crease incision.

the refractive errors associated with hemangioma of the eyelids, found cylinders of 1.5–7 D in 44% of patients.¹⁴ Stigmar et al.¹⁵ reported similar errors in 20% of his studied sample. They both stated that plus cylinder axes correspond to hemangioma site.^{14,15} Our findings support their statement.

4. Conclusion

Ecchymotic hidrocystomas are benign ductal retention cysts of obstructed sweat glands that can rarely develop in the orbital cavity. The lesion presents as a painless mass with various symptoms according to location and size. It is managed by a complete excision and histopathological confirmation. Prompt management in the pediatric age group is essential to prevent amblyopia. This entity should be in the differential diagnosis of orbital cysts in all age groups.

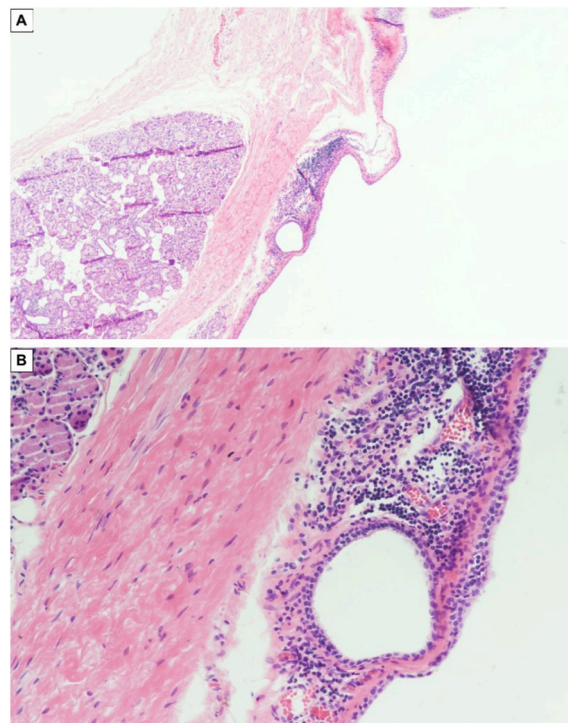


Fig. 5. Histopathologic cross-section of the mass stained with hematoxylin and eosin stain. Low magnification: unilocular cystic space lined by 2 layers of cuboidal cells. (A); High magnification: the lining epithelium is double layered cuboidal to flattened cells slightly eosinophilic cytoplasm and basally located round to oval nucleus. (B).

Table 1
The reported cases of eccrine hidrocystoma.

| No. | Author & Year | Age ^a /Gender | Presenting symptoms | Cyst size | Origin | Radiologic features |
|----------------|--------------------------------------|--------------------------|--|--------------------|------------|---|
| Eyelid | | | | | | |
| 1 | Feijó et al. ¹⁶ | 73/F | Bilateral multiple tumors on both eyelids. | NA | Acquired | NA |
| 2 | Hirata 2013 ⁹ | 2/M | Upper eyelid tumor with no pain or inflammation. The lesion was in direct contact with the tarsal plate. Ptosis and oblique astigmatism developed two years later. | 1.2 × 1.2 × 0.8 cm | Congenital | T2-enhanced MRI: a well-demarcated lesion from the surrounding tissue. |
| 3 | Kumar et al. 2017 ⁶ | 9/F | A painless, progressive drooping right eyelid. | 2.5 × 1.5 cm | Acquired | B-scan: a fluid-filled cavity with no internal echoes. CT: cyst without bone erosion. |
| 4 | Sarabi and Khachemoune ¹⁷ | 69/M | A small, clear, cystic fluid-filled papule in the left upper eyelid. | 0.4 × 0.3 cm | Acquired | NA |
| 5 | Sheth et al. 2008 ¹⁸ | 55/F | Mass, discharge, tearing and ectropion. | 2 cm | Acquired | NA |
| Orbital | | | | | | |
| 6 | Eslami et al. 2007 ¹⁰ | 14/F | Upper eyelid mass for 3 months without any pain or any other symptoms. | 1 × 1 × 2.5 cm | Acquired | CT: an extraconal mass above the medial rectus muscle, mild enhancement. No bone erosion. |
| 7 | Malih et al. 2015 ¹ | 2 months/M | Medial canthal superficial mass, no other symptoms. | Small | Congenital | NA |
| 8 | Malih et al. 2015 ¹ | 8/M | Supra-temporal deep orbital mass with inferior globe displacement and 2 mm proptosis, no diplopia or visual loss. | Large | Acquired | CT: an erosion of the lateral orbital wall. MRI: isointense in T1 and hyperintense in T2. |
| 9 | Marangoz et al. 2016 ¹⁹ | 29/F | A painless, progressive mass. | 1.5 × 1.5 × 0.6 cm | Acquired | MRI: mildly hyperintense on T1 and hyperintense on T2, and showed no enhancement pattern. |
| 10 | Palama et al. 2015 ¹¹ | 57/F | Upper eyelid swelling and pain. Entropion, large central corneal epithelial erosion causing irritation and epiphora. Inferior globe displacement, restricted downgaze. | 2.5 × 2 × 2 cm | Acquired | MRI: extraconal large cyst with an anterior extension towards the eyelid. |
| 11 | The present case | 4/M | A superonasal mass with ptosis. | 1.5 × 2.2 × 1.9 cm | Congenital | B-scan: anechogenic fluid-filled cavity. CT: a hypodense cystic lesion located in the extraconal orbital compartment medially. |

^a Expressed in years unless otherwise specified. CT: Computed Tomography; F: Female; M: Male; MRI: Magnetic Resonance Imaging; NA: Not Available.

Patient consent

Written informed general consent was obtained from the parents, which includes using patient's anonymous information.

Authorship

All authors attest that they meet the current ICMJE criteria for authorship.

Funding

No funding or grant support.

Declaration of competing interest

The authors declared that there is no conflict of interest.

Acknowledgements

None.

References

- Malihi M, Turbin RE, Mirani N, Langer PD. Giant orbital hydrocystoma in children: case series and review of the literature. *Orbit*. 2015;34(5):292–296.
- Mims J, Rodrigues M, Calhoun J. Sudoriferous cyst of the orbit. *Can J Ophthalmol*. 1977;12(2):155–156.
- Singh AD, McCloskey L, Parsons MA, Slater DN. Eccrine hidrocystoma of the eyelid. *Eye (Lond)*. 2005;19(1):77–79.
- Shields JA, Shields CL. Orbital cysts of childhood—classification, clinical features, and management. *Surv Ophthalmol*. 2004;49(3):281–299.
- Alfadley A, Al Aboud K, Tulba A, Mourad MM. Multiple eccrine hidrocystomas of the face. *Int J Dermatol*. 2001;40(2):125–129.
- Kumar DA, Agarwal A. Giant eyelid eccrine hidrocystoma-induced progressive ptosis in childhood. *Indian J Ophthalmol*. 2017;65(9):884–886.
- Alagheband M, Maida MF. Asymptomatic periorbital, bluish cystic papule. *Cortl Forum*. 2004 Jan;17(1):36–41.
- Mehta A, Rao A, Khanna A. Sudoriferous cyst of the orbit of adult origin after trauma. *Indian J Ophthalmol*. 2008;56(3):235–237.
- Hirata A. Eccrine hidrocystoma arising at the tarsal plate during childhood. *Case Rep Ophthalmol*. 2013;4(2):61–63.
- Eslami F, Fazel F, Ghasemibasir H, Beni AN. Giant eccrine hidrocystoma of orbit. *JRMS*. 2007;12:150–152.
- Palamar M, Yaman B, Akalin T, Yagci A. Giant hidrocystoma of the orbit presenting with inversion and ptosis of the upper eyelid. *Turk J Ophthalmol*. 2017;47(2):113–114.
- Valenzuela AA, Heathcote JG. Apocrine hidrocystoma of the orbit. *Orbit*. 2011;30(6):316–317.
- Ssi-Yan-Kai IC, Pearson AR. Recurrent giant orbital apocrine hidrocystoma. *Eye (Lond)*. 2012;26(6):895–896.
- Robb RM. Refractive errors associated with hemangiomas of the eyelids and orbit in infancy. *Am J Ophthalmol*. 1977;83(1):52–58.
- Stigmar G, Crawford JS, Ward CM, Thomson HG. Ophthalmic sequelae of infantile hemangiomas of the eyelids and orbit. *Am J Ophthalmol*. 1978;85(6):806–813.
- Feijó MJF, Sant'Ana HWO, Dias FODM, Salgado Filho IV, Barros ACA. Eyelid polyposis caused by eccrine hidrocystoma. *Rev Bras Cir Plást*. 2013;28(1):165–167.
- Sarabi K, Khachemoune A. Hidrocystomas—a brief review. *MedGenMed*. 2006;8(3):57.
- Sheth HG, Raina J. Giant eccrine hidrocystoma presenting with unilateral ptosis and epiphora. *Int Ophthalmol*. 2008;28(6):429–431.
- Marangoz D, Dogan Ekici I, Ciftci F. Orbital eccrine hidrocystoma. *Turk J Ophthalmol*. 2016;46(5):251–254.