Presentation of massive orbital hidrocystoma at birth: case report and review of the literature

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Abstract

Background: Hidrocystoma, or sudoriferous cyst, is an eyelid tumor originating from apocrine or eccrine sweat glands. Its presence in the orbit is relatively rare.

Case presentation: A full-term female child with severe right eye extrusion was referred to our department two hours after birth. We performed cyst aspiration under ultrasonic guidance; 15 cc straw-colored fluid was obtained and proptosis resolved significantly. Orbital magnetic resonance imaging (MRI) showed a large unilocular retrobulbar mass with hypo-intensity in T1 and hyper-intensity in T2. The case underwent further daily ocular assessment until day 5; when proptosis began to worsen again. On day 6, under general anesthesia, we performed aspiration and then the cyst was completely removed with an intact wall through a trans-conjunctival incision. The diameter of the aspirated cyst was about 4 cm. In pathologic consultation, a cystic cavity lined by a layer of sweat duct like epithelium with apical snouts consistent with the diagnosis of apocrine hidrocystoma was reported.

Conclusion: To date, in the literature, six other cases of orbital hidrocystoma have been reported in childhood with protean clinical pictures; none of which presented at birth. Herein, we introduce the first case report at birth and also provide a review on the literature. Our report strongly argues against the well reputed theory of traumatic origin for orbital hidrocystoma; it has been postulated that this tumor may be the result of sweat gland cells implantation through the orbit. We thereby suggest the possible presence of choristomatous ectopic sweat gland cells in the orbit during embryogenesis.

Keywords: Orbital hidrocystoma, Pediatric orbital tumor, Sudoriferous cyst
afferent pupillary defect was detectable. Anterior/posterior segment examination of the left eye was unremarkable. Based on the opinion of the consulting neonatologist regarding the general conditions of the case, general anesthesia and imaging was postponed to at least 72 h later. We admitted the case in our ward and performed cyst aspiration under ultrasonic guidance; 15 cc straw-colored fluid was obtained and proptosis resolved significantly. Then, blepharorrhaphy was performed due to eyelids eversion and severe conjunctival chemosis. Appropriate topical drugs were administered and the patient referred again to the neonatology department for further stabilization and evaluation. Orbital magnetic resonance imaging (MRI) showed a large retroorbital mass being unilocular and extraconal with hypo-intensity in T1 and hyper-intensity in T2 (Fig. 2). The case underwent further daily ocular assessment until day 5; when proptosis began to worsen again (Fig. 3). On day 6, under general anesthesia, we aspirated the mass again and then, through a trans-conjunctival incision 3 mm beneath the tarsal plate, the mass was exposed. It was located in the inferior aspect of the orbit extending to its apex. Hemorrhagic vessels were cauterized and by blunt and sharp dissection, the mass was completely removed en-bloc. The wound was closed with vicryl 8.0. The diameter of the aspirated mass was about 4 cm (Fig. 4). In pathologic consultation, a cystic cavity lined by a layer of sweat duct like epithelium with apical snouts consistent with the diagnosis of apocrine hidrocystoma was reported (Fig. 5).
patient was followed for 3 months (Fig. 6) and had no significant complications.

**Discussion**

Presentation of hidrocystoma in the orbit is extremely rare to the extent that its presentation at birth, as a micro-cyst had only been reported once [2]. The point that makes our case very impressive was its massive size, orbital location and presentation at birth.

As mentioned earlier, there are 7 other known cases of orbital hidrocystoma in childhood [2–6] (Table 1). Of the 8 existing cases, 5 presented before the age of 1 while 3 cases presented at later ages. Tumor types were apocrine in 6 and eccrine in 2 cases. We did not have access to the full-text version of one of these cases [3]. Of 7 other known orbital locations, 3 were superficial and 4 were deep [2, 4–6]. Only one case had a history of significant orbital trauma [6]. Origin sites were medial (3 cases), superior (1 case), supra-temporal (1 case), inferior (1 case) and retro-bulbar (1 case). Computed tomography (CT) scan may demonstrate bone remodeling with no erosion. The MRI signal was reported as hypo- or iso-intense in T1 and hyper-intense in T2 [2–6].

From a pathological standpoint, hidrocystoma is a benign cyst originating from a sweat gland apocrine or eccrine in nature. The apical part of cellular cytoplasm is decapitated in the apocrine type (decapitation), while it remains intact in eccrine (exocytosis) glands [6]. This difference in secretion mechanisms gives the pathologic appearance of apical ‘snouts’ in the apocrine type.

In a review by Shield and Shield, the authors believe that differentiating between the main causes of orbital...
orbital cysts. Presentation of a large deep orbital hidrocystoma at birth in our case suggests the possible presence of choristomatous ectopic sweat gland cells in the orbit during embryogenesis. It is argued against the well-reputed traumatic origin for orbital hidrocystoma by which it was postulated that this tumor may be the result of sweat gland cells implantation through the orbit [6]. Presentation of a large deep orbital hidrocystoma at birth in our case suggests the possible presence of choristomatous ectopic sweat gland cells in the orbit during embryogenesis.

Taken together, orbital hidrocystoma should be considered as a differential diagnosis of small/large; superficial/deep; congenital/post-traumatic; and, childhood/adult orbital cysts.

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Authors’ contributions
BE, MAA and SHA conceived the idea for the case study and followed the patient. MAA, SHA, SAS, ZK wrote the article and SAS and ZK revised it critically for important intellectual content. All authors read and approved the final manuscript.

Competing interests
The authors declare that they have no competing interests.

Consent for publication
Written informed consents were obtained from the parents for publication of this Case Report and any accompanying images. A copy of the written consent is available for review by the editor of this journal.

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References
1. Sarabi K, Khachemoune A. Hidrocystomas—a brief review. Med Gen Med. 2006;8:57.
2. Saunders JF. Congenital sudoriferous cyst of the orbit. Arch Ophthalmol. 1973;89(3):205–6.
3. Mims J, Rodrigues M, Calhoun J. Sudoriferous cyst of the orbit. Can J Ophthalmol. 1977;12:155–6.
4. Haider E, Saigal G, Gill D, Brown E, Daniel S. Congenital orbital sudoriferous cyst: radiological findings. Pediatr Radiol. 2005;35:1142–4.
5. Chung I, Lee S, Kang SK, Park SH. Congenital sudoriferous cyst within the orbit followed by esotropia. Korean J Ophthalmol. 2007;21:120–3.
6. Malihi M, Turbin RE, Mirani N, Langer PD. Giant orbital hidrocystoma in children: case series and review of the literature. Orbit. 2015;34:292–6.
7. Shields JA, Shields CL. Orbital cysts of childhood—classification, clinical features, and management. Surv Ophthalmol. 2004;49:281–99.

Table 1 Reported cases of orbital hidrocystoma in childhood

| Case number | Author(s) Year | Patient Age; Sex | Presentation symptoms | Cyst size | Subtype | Radiologic features |
|-------------|----------------|-----------------|-----------------------|----------|---------|---------------------|
| 1 | Saunders [2] 1973 | At birth; Male | Medial superficial orbital mass, without globe displacement no visual axis involvement | 2 mm | Apocrine | CT: cyst without bone erosion |
| 2 | Mims et al. [3] 1977 | NA | NA | NA | Apocrine | NA |
| 3 | Haider et al. [4] 2005 | 4 months; Male | Inferior deep orbital mass with superior globe displacement | Large | Apocrine | CT: slight bone remodeling |
| 4 | Chung et al. [5] 2007 | 20 days; Male | Medial deep orbital mass with lateral globe displacement | About 1.3 cm | Apocrine | MRI: hypointense in T1 and hyperintense in T2 |
| 5 | Malihi et al. [6] 2015 | 8 y/o; Male | Supra-temporal deep orbital mass with inferior globe displacement and 2 mm proptosis, no diplopia or visual loss | Large | Eccrine | CT: erosion of lateral orbital wall |
| 6 | Malihi et al. [6] 2015 | 13 y/o; Female | Superior superficial orbital mass without globe displacement, no diplopia, proptosis or visual loss, history of significant blunt trauma | Large | Apocrine | MRI: isointense in T1 and hyperintense in T2 |
| 7 | Malihi et al. [6] 2015 | 2 months; Male | Medial canthal superficial mass, no other symptoms | Small | Eccrine | Not performed |
| 8 | Present case 2016 | At birth; Female | Deep retro-orbital mass, globe extrusion | Huge | Apocrine | MRI: hypointense in T1 and hyperintense in T2 |

NA = not available; y/o = years old; CT = Computed Tomography; MRI = Magnetic Resonance Imaging