Orbital apocrine hidrocystoma with ptosis

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ABSTRACT

Purpose: To describe a patient with orbital apocrine hidrocystoma presenting with ptosis and subsequent management.

Observations: A 43-year-old woman presented to the oculoplastic surgery clinic with a left-sided ptosis and enlarging but painless mass in the sulcus of the left upper eyelid. Magnetic resonance imaging demonstrated a large, circumscribed T2 bright cystic lesion in the extraconal space. Surgical excision and histopathology confirmed a diagnosis of apocrine hidrocystoma.

Conclusions: Although uncommon, ptosis may be a presenting symptom of an orbital apocrine hidrocystoma, which should be considered in the differential diagnosis for an extraconal cystic lesion. Apocrine hidrocystomas are benign tumors and are cured with surgical excision with rare recurrence.

1. Introduction

Apocrine hidrocystomas are benign tumors arising from apocrine sweat glands that are associated with hair follicles. Apocrine sweat glands are the predominant sweat glands of the head and neck region. In the eyelids, they are modified sweat glands located at the eyelid margin and are referred to as glands of Moll. The exact process leading to their growth is unknown. In the head and neck region, apocrine hidrocystomas most commonly occur intradermally at the eyelid margin or within the external auditory canal. Intraorbital apocrine hidrocystomas are especially rare in adults and of the few reported cases, may be associated with facial trauma. We report the case of a previously healthy woman with an orbital apocrine hidrocystoma in the setting of unilateral ptosis and enlarging orbital mass.

2. Case report

A 43-year-old woman presented with left-sided ptosis that had progressed over the prior two years. She noticed a gradually enlarging, nontender mass centrally located in the sulcus of the left upper eyelid. She denied any visual changes, discharge, or bleeding. She also denied prior orbital trauma or surgeries. Distance visual acuity with her current eyeglasses was 20/20 in the right eye and 20/40 in the left. Ocular motility was full, and pupils were equal, round, and reactive to light. External examination was notable for a firm, mobile, subcutaneous mass in the center of her left supratarsal fold with a few overlying feeder vessels but without erythema, warmth, scaling, or ulceration (Fig. 1). No lesions were observed on eyelid eversion. Margin-reflex distance (MRD1) on the left was 2.5 mm with good levator function. The remainder of her examination was normal. Magnetic resonance imaging (MRI) demonstrated an 18 × 8 mm circumscribed T2/Short Tau Inversion Recovery (STIR) bright lesion with thin internal septations and trace delayed enhancement without bony erosion (Fig. 2).

An incision through the eyelid crease revealed a multilobed cystic-appearing lesion partly embedded within the superior tarsal muscle and involving the anterior orbital portion of the lacrimal gland. The cyst was first drained and then filled with fluorescein-stained fibrin glue. The glue was allowed to harden, and the cyst was completely excised as previously described. It was then submitted to pathology. Histopathology revealed an arborizing, cystic space lined by a double epithelium exhibiting apical snouting, consistent with an apocrine hidrocystoma (Fig. 3).

On follow up examination at 1 month, the patient’s MRD1 improved with no recurrence of the lesion (Fig. 4). The patient was reassured and advised to follow up for recurrence or new concerns.

3. Discussion

Apocrine hidrocystomas are benign cystic tumors that grow from apocrine sweat glands. Unlike eccrine hidrocystomas, apocrine hidrocystomas rarely occur as multiple lesions in the general adult population, usually presenting instead as a solitary lesion. However, multiple
Apocrine hidrocystomas have been associated with inherited conditions, including Schopf-Schulz-Passarge syndrome and Goltz-Gorlin syndrome. Since they arise from apocrine glands, extension beyond the eyelid margin is rare. It is hypothesized that trauma may introduce epithelial cells into the orbit, seeding their growth, but no history of trauma was present in our patient. Apocrine hidrocystomas usually range from 3 mm to 15 mm in diameter and remain asymptomatic. Larger lesions may cause ocular discomfort and pressure sensation. Mukherjee et al. reported a 3 cm hidrocystoma causing proptosis and mechanical ptosis secondary to mass effect. Ferraz et al. reported cases of ptosis resulting from occult hidrocystomas discovered only at the time of surgery arising within the levator muscle and adherent to the levator aponeurosis. There are many potential mechanisms for acquired ptosis. While our patient is approaching the appropriate age range for the development of aponeurotic ptosis, this would not be expected to resolve with only resection of the cyst and without additional manipulation of the levator or superior tarsal muscle. Similarly, neurogenic and myogenic causes of ptosis would not be expected to resolve without additional intervention. With regard to mechanical ptosis, our patient’s mild degree of ptosis would be consistent with direct impingement of the superior tarsal muscle supported by intraoperative visualization of superior tarsal muscle infiltration with additional contribution from mass effect and weight of the lesion. Additional imaging may be used to further characterize orbital masses and inform surgical approach. MRI of an apocrine hidrocystoma usually demonstrates a circumscribed, cystic mass with variable T1 signal and high T2 signal. STIR is a fat suppression technique used to enhance sensitivity to tissue fluid and may highlight the cystic contents. Unfortunately, an isointense T1 and hyperintense T2/STIR signal can also be seen in vascular lesions, such as a cavernous hemangioma, which was the original radiologic interpretation in this patient. Because the clinical presentation of an apocrine hidrocystoma may resemble that of other orbital lesions, such as dermoid cysts, mucoceles, or hemangiomas, definitive diagnosis is made by histopathology. Apocrine hidrocystomas may be unilocular or multilocular cystic spaces and characteristically have a bilaminar epithelium composed of an inner cuboidal to columnar cell layer with eosinophilic cytoplasm and prominent apical snouts and an outer layer of more flattened cells. Chronic inflammation may also be seen in the interstitium. Small, asymptomatic apocrine hidrocystomas may be observed, and
treatment of symptomatic lesions is surgical excision, which almost always leads to complete resolution. Recurrence has been reported with incomplete excision of the cyst wall. Excision of the intact cyst may be complicated by cyst rupture, so fibrin glue mixed with fluorescein was utilized in this patient’s surgery to fill the cyst cavity and facilitate delineation of margins for complete excision. This method has been previously described using fibrin glue mixed with other dyes, including indocyanine green, trypan blue, and methylene blue. Postoperative scarring may occur, so less invasive methods are currently under investigation. Del Pozo et al. described the use of carbon dioxide laser ablation to treat multiple small scattered hidrocystomas. Osaki et al. reported successful treatment of small hidrocystomas with hypertonic glucose sclerotherapy. For lesions as large as 15 mm, chemical ablation to treat multiple small scattered hidrocystomas.

4. Conclusions

Although apocrine hidrocystomas are benign, they should remain on the differential for an extracanal cystic lesion, and concerning lesions should undergo surgical excision and biopsy to rule out other malignancies. Histopathology leads to definitive diagnosis, which demonstrates a characteristic apical snouting, and complete excision is the standard treatment.

Patient consent

Written informed general consent was obtained from the patient.

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Authorship

All authors attest that they meet ICMJE criteria for authorship.

Declaration of competing interest

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