CLINICAL IMAGES

Bilateral Painless Eye Lesions

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A thin 65-year-old African American female with a past medical history of hypertension presented to her primary care physician with a chief complaint of bilateral "eye bumps". She stated that they were present for years and only decided to come to the office after her relatives insisted it be evaluated. The patient did not endorse any other ocular problems including decreased vision, photophobia, pain, irritation or tearing. She denied any change in size, color or shape of the bumps over the years. She had no past history of other ocular lesions, no known allergies or drug use. The patient did not present with any other notable craniofacial abnormalities including auricular appendages, fistulas, cleft palate or postural dysfunction. Review of systems was otherwise unremarkable.

Physical exam revealed the presence of bilateral, symmetric, 1cm in diameter, pinkish-yellow, circular masses superior-temporally to the lateral canthus of the eyes. Lifting of the eyelid superiorly further elucidated the size of the mass to be closer to 3cm in diameter. Upon palpation, the mass was soft, non-mobile and could not be indented with a cotton-tip applicator. The mass appeared to be lying on the lateral bulbar conjunctiva. Extraocular muscle motility was within normal limits. Pupils were round, equal, reactive, and with no afferent pupillary defect. A funduscopic examination revealed an unremarkable optic disc without papilledema or signs of neovascularization.

QUESTION:

What is the diagnosis?

A. Dermolipoma

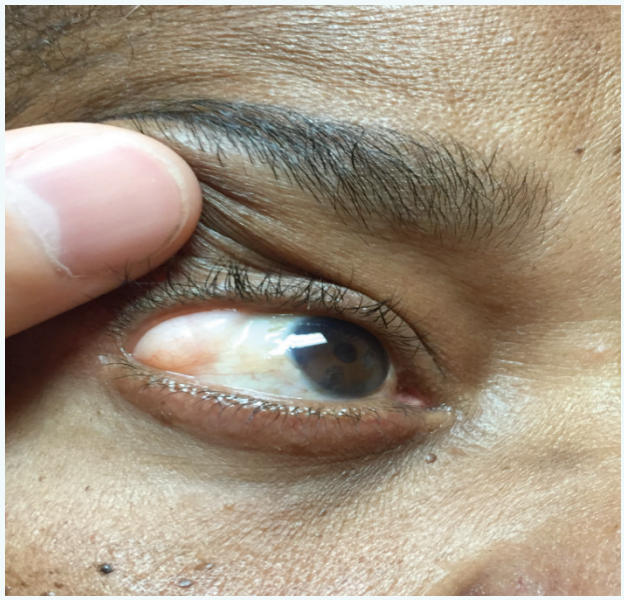
B. Orbital fat prolapse

C. Pinguecula

D. Pterygium

E. Squamous Cell Carcinoma

FIGURE 1:



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ANSWER

What is the diagnosis?

The correct answer is: A) Dermolipoma

DISCUSSION

An orbital dermolipoma is a benign congenital tumor of the bulbar conjunctiva. It can be more broadly described as a type of dermoid cyst, or choristoma which a group of normal cells in an abnormal location in the body.[1] Histologically composed of adipose tissue surrounded by an outer connective tissue covering, these typically small tumors often go undetected until later in life due to their asymptomatic nature. Their incidence is noted to be rare (less than one in 10,000 live births) and they have an overall female predominance.[2,3]

The classic description on physical exam, consistent with the patient noted on page 45, includes a well demarcated, pinkish-yellow, gelatinous appearing immobile mass in the superior temporal region of the bulbar conjunctiva.[4] These lesions are almost always found laterally and frequently unilaterally; however, case reports have noted rare medial presence of dermolipomas.[5]

Orbital fat prolapse is often noted to be the closest mimicker of dermolipomas given that both of these conditions typically present as soft yellowish masses in the superotemporal bulbar conjunctiva. However, a closer examination reveals key differentiating features between the two conditions. First, demographically, orbital fat prolapse has a propensity to occur in older (mean age 65-72) obese, males, unlike dermolipomas which classically is first noticed in thin younger(mean age 22.5) females.[2] Next, orbital fat prolapse's convex, freely mobile anterior margin can be easily reduced back into the orbit which sharply contrasts the concave, immobile, non-reducible anterior margin of dermolipomas. Finally, upon magnification of the margin's surface, superficial blood vessels are more often seen in the orbital fat prolapse while fine hairs can be appreciated on the outer layer of dermolipomas.[6]

Squamous cell carcinoma can also appear as a gelatinous unilateral conjunctival mass but is typically more central in location, near the limbus of the eye, with pronounced red blood vessels visible on a pinkish-white base. It can grow very fast, over period of months, and is usually seen in older men.[3] Pingueculas are yellow, well demarcated nodules that are very common in adults. These benign, immobile growths are usually only slightly raised, clearly distinct from the peripheral eye borders and more often located nasally—unlike dermolipomas.[3] A pterygium is a fleshy appearing, triangularly shaped, conjunctival growth that advances nasally toward the cornea, also seen in older men.[3]

The diagnosis of a dermolipoma is made clinically but if uncertain, imaging studies such as CT or MRI can help differentiate the mass based on fat extension and location.[4] Very rarely does a biopsy need to be performed to confirm the diagnosis. Most commonly no treatment is needed and conservative therapy such as observation is elected. Consideration of surgery should be limited but may be appropriate if there is mechanical compromise of lid function, cosmetic concerns, or persistent irritating foreign body sensation.[7] However, patients must be counseled on the many potential negative post-operative complications (xerophthalmia, diplopia, ptosis) due to close proximity to vital structures of the eye (lacrimal gland, lateral rectus muscle, conjunctiva) that often outweigh the benefits.[7]

While these lesions are benign, they can be associated with a constellation of other symptoms portending an overall worse prognosis. One of the most well-known examples of this is Goldenhar-Gorlin syndrome, a rare syndrome, estimated to be seen in 1 out every 3,500 to 5,600 live births, characterized by ocular anomalies, auricular appendages, and vertebral anomalies.[8,9] Additional associated abnormalities in this disorder may also include renal or facial hypoplasia, as well as cardiac defects(example ventral septal defects) which occur in 5 to 58 percent of all cases.[10]

Ophthalmologic anomalies occur in about 50 percent of Goldenhar-Gorlin syndrome cases, one of the most common of which is a dermolipoma.[8] The dermolipomas seen in this condition however, often are quite pronounced, located in the infratemporal quadrant and are most often unilateral.[5] The collection of symptoms required for diagnosis are noticeable at birth but because there is such a wide range of overlapping anomalies the diagnosis may be missed.[9] When Goldenhar-Gorlin syndrome is suspected, an echocardiogram and CT imaging can be used to confirm or detect the additional internal defects noted above that can be associated with the condition. The treatment of this disease varies with age and systemic associations but typically involves cosmetic reconstruction based on clinical presentation after the age of five.[10]

This patient was completely asymptomatic, with no other associated findings and satisfied with the reassurance of the lesions benign nature. She will continue to monitor the dermolipoma as she had for the past 60 years.

REFERENCES:

1. Mansour AM, Barber JC, Reinecke RD, et al. Ocular Choristomas. Surv Ophthalmol. 1989 Mar-Apr; 33(5):339-58.
2. McNab AA, Wright JE, Caswell AG. Clinical features and surgical management of dermolipomas. Aust N Z J Ophthalmol. 1990 May; 18(2):159–62.
3. Riordan-Eva P, Cunningham E, Jr: Vaughan & Asbury's General Ophthalmology. New York, Lange Medical /McGraw-Hill Medical Pub. Division, 2011, ed 18, pp 107-112.
4. Kim E, KimHJ, KimYD, et al. Subconjunctival Fat Prolapse and Dermolipoma of the Orbit: Differentiation on CT and MR Imaging. AJNR Am J Neuroradiol. 2011 Mar; 32(3): 465-467
5. Rajabi MT, Ramezani K. Lipodermoid cyst: A report of a rare caruncular case. MEAJO 2015; 22(4): 528-530.
6. Kim YD, Goldberg RA. Orbital fat prolapse and dermolipoma: two distinct entities. Korean J Ophthalmol 1994; 8:42-43.
7. Fries P, Kazim M. 2002. Benign Pediatric Orbital Tumors. Katowitz J, Editor. New York (NY): Springer. Chapter 13, Pediatric Oculoplastic Surgery; p. 442-44.
8. Mansour AM, Wang F, Henkind P, et al. Ocular findings in the facioauriculovertebral sequence (Goldenhar-Gorlin syndrome). Am J Ophthalmol 1985 Oct; 100(4): 555-9.
9. Kumaresan R, Srinivasan B, Narayanan M, et al. Craniofacial abnormalities in goldenhar syndrome: a case report with review of the literature. Plast Aesthetic Res. 2014;1(3):108
10. Sharma JK, Pippal SK, Raghuvanshi SK, Shitij A. Goldenhar-Gorlin's syndrome: A case report. Indian J Otolaryngol Head Neck Surg. 2006; 58(1):97-101.