Translucent Periorbital Papules

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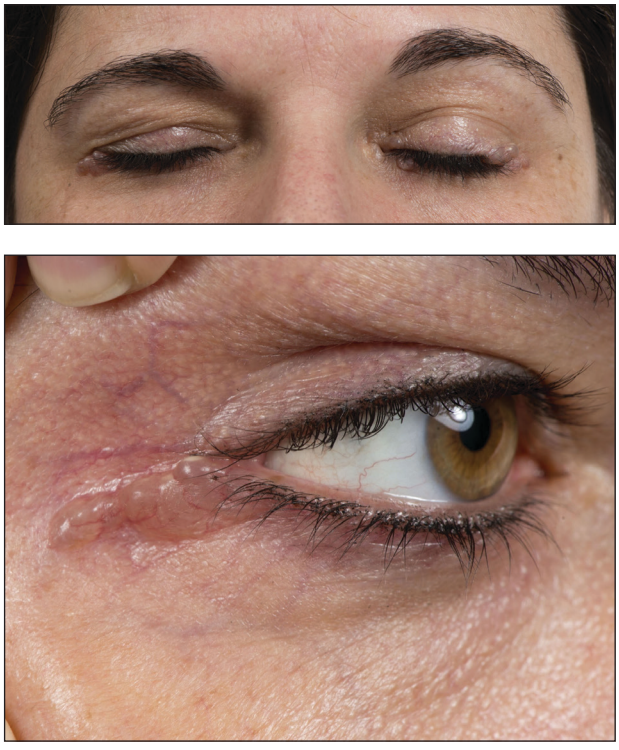

A 40-year-old woman was referred to dermatology for evaluation of occasionally pruritic periorbital papules that had gradually increased in size and number over the last 7 to 8 months (top). She had a similar solitary lesion on the left lower eyelid that was removed twice: 10 years and 10 months prior. She was taking an oral contraceptive (desogestrel) but otherwise had no notable medical history or drug allergies. Physical examination revealed individual and clustered translucent papules along the eyelid margins, left medial canthus, and both lateral canthi (bottom).

WHAT'S THE **DIAGNOSIS?**

a. apocrine hidrocystoma
b. lipoid proteinosis
c. lymphangioma circumscriptum
d. primary systemic amyloidosis
e. syringoma

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THE **DIAGNOSIS:** Apocrine Hidrocystoma

Histopathologic examination of one of the papules revealed cystic cavities located within the dermis (Figure 1) lined by a cuboidal epithelium demonstrating decapitation secretion (Figure 2), confirming the diagnosis of apocrine hidrocystomas. The presence of multiple lesions prompted further examination for an underlying genetic disorder; however, the patient's hair, nails, and teeth were normal. There also was no evidence of palmoplantar keratoderma or blaschkoid dermatosis.

Hidrocystomas are benign cysts of the sudoriferous apparatus that can be subdivided based on histogenesis (apocrine vs eccrine) or lesion count (single vs multiple).[1] Multiple lesions may be associated with disorders of ectodermal dysplasia, including Goltz syndrome and Schopf-Schulz-Passarge syndrome. Apocrine hidrocystomas tend to present as solitary, translucent, flesh-colored to bluish facial papules, and the occurrence of multiple lesions is rare in contrast to its eccrine counterpart.[2]Various extrafacial sites have been described including the trunk, axillae, umbilicus, genitalia, and digits.[3] Apocrine hidrocystomas do not demonstrate aggravation with exposure to heat, unlike their eccrine counterparts.[2]

A review of 107 patients with 215 histologically proven hidrocystomas demonstrated a preponderance for women in their mid 50s; 74.8% of patients had unilateral disease, and 69.8% of all lesions affected either the lower eyelid or lateral canthus. Recurrence following conventional surgical excision was observed in 2.3% of lesions.[1]

A review from Japan recounted an incidence of 5 cases per year from 1999 to 2003.[4] Patients ranged in age from 30 to 70 years, but there was no gender predilection. Individual apocrine hidrocystomas were mostly less than 2 cm and varied from flesh colored to light red, brown, blue, or purple; 61% of lesions arose periorbitally. Within their cohort, patients with multiple lesions were uncommon, with only 2 cases presenting with 2 lesions simultaneously.[4]

Apocrine hidrocystomas are thought to result from a cystic proliferation of the secretory component of apocrine sweat glands, though the exact pathogenesis still is unclear.[3] Histologic features include a unilocular or multilocular cystic cavity within the dermis lined by columnar cells demonstrating decapitation secretion, followed by a peripheral rim of flattened myoepithelial cells.

Treatment of apocrine hidrocystomas includes topical anticholinergics, surgical excision, electrodesiccation, 1450-nm diode or CO_2 lasers, and trichloroacetic acid.[2] The novel use of cryotherapy,[5] botulinum toxin,[2] and intralesional injections of 50% glucose (as a sclerosant)[6] also have been reported. Caution should be exercised when managing digital lesions, as digital papillary carcinoma has been described as a clinical and histopathologic mimicker.[7]

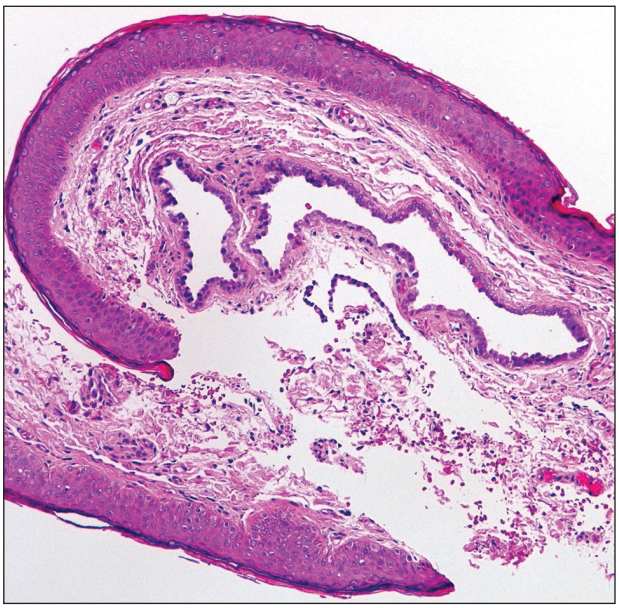

FIGURE 1. Cystic spaces within the dermis (H&E, original magnification ×10).

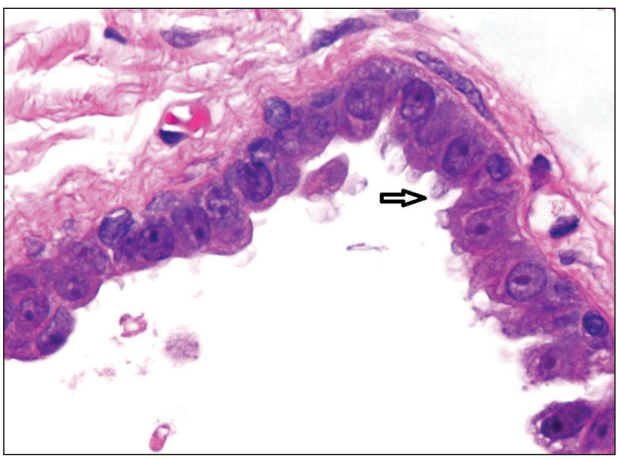

FIGURE 2. The cyst was lined by epithelial cells demonstrating decapitation secretion (arrow)(H&E, original magnification ×40).

Lipoid proteinosis is a rare autosomal-recessive disorder. Cutaneous lesions manifest in 2 overlapping stages, typically within the first 2 years of life. The first stage consists of vesicles and hemorrhagic crusts on the face and extremities and intraorally, which may heal with scarring. In the second stage, the skin becomes diffusely thickened and waxy, with the appearance of papules, nodules, or plaques along the eyelid margins (moniliform

blepharosis), face, axillae, or scrotum. Verrucous lesions also may develop on the knee or elbow extensors.[8]

Lymphangioma circumscriptum represents microcystic lymphatic malformations that can arise anywhere on the skin or oral mucosa. They present as clusters of clear or hemorrhagic vesicles of variable size and number favoring the proximal extremities and chest. Histologically, dilated lymphatic channels are seen in the upper dermis.[8]

Syringomas are common benign tumors of the sweat ducts characterized histologically by superficial dermal proliferations of small comma-shaped ducts set in a fibrotic stroma. Clinically, syringomas appear as small, firm, flesh-colored papules with a predilection for the periorbital area. An eruptive onset may be observed, most commonly affecting the trunk. Syringomas may be associated with Down syndrome, while the clear cell variant may be associated with diabetes mellitus.[8]

Primary systemic amyloidosis may present with a variety of systemic manifestations. Skin involvement can present as waxy, translucent, or purpuric papulonodules or plaques characteristically affecting the periorbital region. Other mucocutaneous signs include macroglossia with or without translucent to hemorrhagic papulovesicles; bruising, especially on the eyelids, neck, axillae, or anogenital area; vesiculobullous skin lesions; or diffuse cutaneous infiltration imparting a sclerodermoid appearance.[8]

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