Numerous Flesh-Colored Nodules on the Trunk

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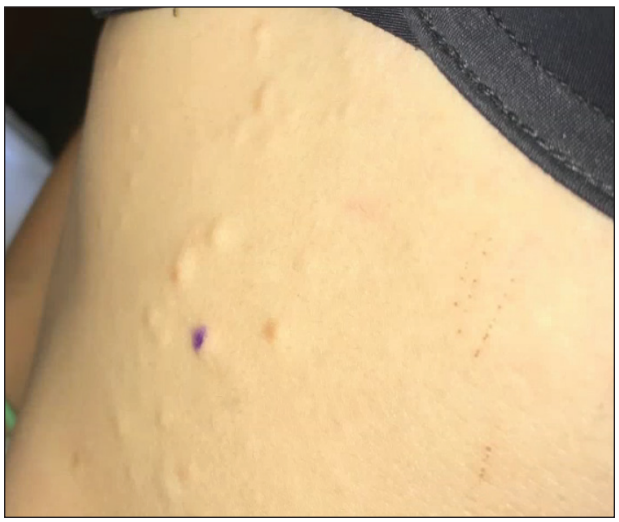

A 33-year-old woman presented with numerous firm, noncompressible, flesh-colored nodules that measured 3 to 4 mm and were distributed across the abdomen, chest, back, and neck. The lesions had been present for approximately 10 years. The patient denied any lesion-associated pain, itching, or bleeding, and there was no family history of similar lesions. A punch biopsy of a lesion on the central abdomen was obtained.

WHAT'S THE **DIAGNOSIS?**

a. eruptive vellus hair cysts
b. Gardner syndrome
c. lipomas
d. Muir-Torre syndrome
e. steatocystoma multiplex

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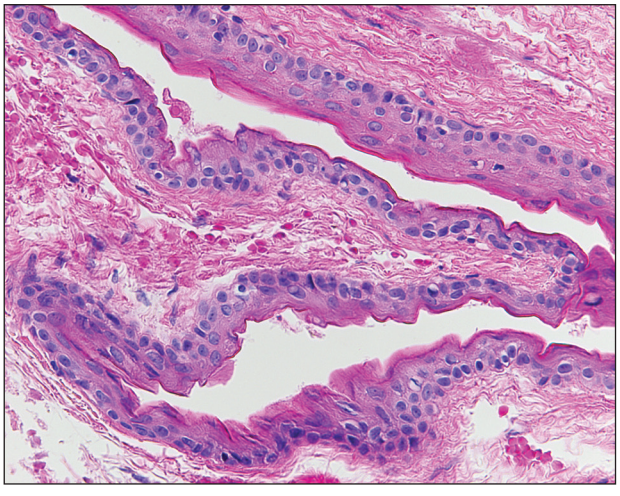
THE **DIAGNOSIS:** Steatocystoma Multiplex

The punch biopsy of an abdominal lesion demonstrated a folded cyst wall with a wavy eosinophilic cuticle (Figure), characteristics consistent with steatocystoma multiplex (SM).

Also known as eruptive steatocystoma, SM consists of numerous flesh-colored, dome-shaped papules and nodules that most commonly arise during adolescence, with a median age of onset of 26 years.[1] These hamartomatous nevoid malformations arise in areas with well-developed pilosebaceous units, such as the upper extremities, neck, axillae, and trunk.[1,2] They occur less commonly on the scalp, face, and acral surfaces.[2-5] The lesions range in size from 2 to 30 mm[6] and usually are asymptomatic.[1] Occasionally, steatocystomas become tender or can rupture.[7]

Steatocystoma multiplex may arise sporadically or may be inherited in an autosomal-dominant fashion. Mutations in exon 1 of the keratin 17 gene, *KRT17*, have been identified in autosomal-dominant SM.[6,8] *KRT17* mutations also are responsible for pachyonychia congenita type 2, which is associated with SM.[9] Some patients with pachyonychia congenita type 2 who have prominent SM and mild nail findings may be misdiagnosed as having pure SM.[2]

The histopathologic features of SM were described in a study by Cho and colleagues[1] of 64 patients. Steatocystomas have cyst walls that may be either intricately folded or round/oval, comprised of an average of 4.9 epithelial cell layers. In most cases, the cyst wall contains sebaceous lobules. In all cases, an acellular eosinophilic cuticle was present, and no granular layer was seen. Few vellus hairs may be observed in the cystic cavity.[1]



Punch biopsy demonstrated a folded cyst wall with a wavy eosinophilic cuticle characteristic of steatocystoma (H&E, original magnification ×200).

The differential diagnosis of SM includes eruptive vellus hair cysts, lipomas, Muir-Torre syndrome, and Gardner syndrome. Some have suggested that eruptive vellus hair cysts and SM exist on a disease spectrum because of their similar clinical presentation.[10] In contrast to SM, however, eruptive vellus hair cysts originate in the infundibulum of the hair shaft rather than the sebaceous duct, and more numerous vellus hair shafts are seen on histopathology.[1]

Various treatment modalities have been described, including isotretinoin for inflamed lesions,[11] cryotherapy for noninflamed lesions,[11] aspiration of lesions smaller than 1 cm,[12] and electrocautery combined with topical retinoids.[13] Laser treatment has been described, with a 1450-nm diode laser used to target the abnormal sebaceous glands and a 1550-nm fractionated erbium-doped fiber laser used to target the dermal cysts.[14] Carbon dioxide lasers also may be used to open the cyst for drainage.[15] Surgical excision or mini-incision also may be performed.[16,17]

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