

# Co-occurrence of Steatocystoma Multiplex, Eruptive Vellus Hair Cysts, and Trichofolliculomas



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## PRACTICE POINTS

- Steatocystoma multiplex (SCM) and eruptive vellus hair cysts (EVHCs) have similar clinical features but distinctive histologic features.
- Milia, pilar cyst, trichoepitheliomas, and trichoblastomas simultaneously developing in association with SCM or EVHC on the face are rare.
- This case supports the hypothesis that these benign follicular neoplasms are related but distinct nevoid malformations of the pilosebaceous unit within the same disease spectrum.

An association between steatocystoma multiplex (SCM) and eruptive vellus hair cysts (EVHCs) has been recognized. Steatocystoma multiplex and EVHC have similar clinical features but distinctive histologic features. Rare cases of co-occurrence of these conditions have been known to occur on the trunk and the forehead. We report a rare case of the simultaneous occurrence of SCM, EVHC, and trichofolliculomas localized to the forehead.

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An association between steatocystoma multiplex (SCM) and eruptive vellus hair cysts (EVHCs) has been recognized. They are related conditions representing nevoid malformations of the pilosebaceous junctions<sup>1-10</sup> that have similar clinical features but distinctive histologic features. Both conditions most commonly involve the anterior aspect of the chest. Six cases of a rare facial variant of SCM have been reported,<sup>11-16</sup> 3 involving lesions limited to the forehead.<sup>13-15</sup> Two patients with a rare facial variant of EVHC also have been reported.<sup>17</sup> The development of separate lesions of SCM and EVHC on the trunk can uncommonly occur.<sup>5,6,10</sup> One case of

SCM and EVHC on the forehead has been described.<sup>3</sup> Other types of benign follicular neoplasms simultaneously developing in association with SCM or EVHC also are rare. The simultaneous occurrence of multiple trichoblastomas, trichoepitheliomas, and SCM on the face and trunk has been reported in 1 case.<sup>18</sup> Milia, SCM, and EVHC on the face and trunk have been reported in 1 family.<sup>4</sup> A report of facial steatocystoma associated with a pilar cyst and bilateral preauricular sinus also has occurred in 1 patient.<sup>19</sup> Here, we report the simultaneous occurrence of SCM, EVHC, and trichofolliculomas localized to the forehead.

## Case Report

A 37-year-old man had an increasing number of flesh-colored to yellow papules on the forehead that had been present since puberty. Although the lesions were asymptomatic, some had recently become tender, which led him to seek medical care. There was no history of trauma, burns, irradiation, or application of topical agents to the area or use of eyeglasses or goggles. The patient's father had similar lesions limited to the forehead, which developed during adolescence.

On evaluation at our clinic, skin examination revealed 16 discrete, 0.3- to 1-cm, flesh-colored, yellow to blue, mobile, smooth papules, as well as flesh-colored papules with a central black punctum, on the forehead (Figure 1). Similar lesions were not present on the rest of the face; around the ears; or on the scalp, neck, chest, back, abdomen, genitalia, buttocks, palms, soles, axillae, arms, or legs. There were no nail abnormalities.

Multiple 3-, 4-, and 6-mm punch and excisional biopsies were performed to remove all 16 lesions on the forehead. Histologic examination revealed a collapsed cystic structure in the mid dermis in 10 lesions. The cysts were lined with a squamous epithelium without a granular

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layer but with an eosinophilic corrugated lining, and the cyst cavity contained scant homogeneous eosinophilic secretion. Mature sebaceous glands were adjacent to the outer portion of the cyst wall. These histologic findings were consistent with SCM (Figure 2).

In 3 lesions, histologic examination revealed a cystic structure lined by a few layers of stratified squamous epithelium in the mid dermis. The cyst cavity contained numerous small vellus hairs and laminated keratin. These histologic findings were consistent with EVHC (Figure 3).

In the other 3 lesions, histologic examination revealed a dilated central cystic cavity filled with laminated keratin in

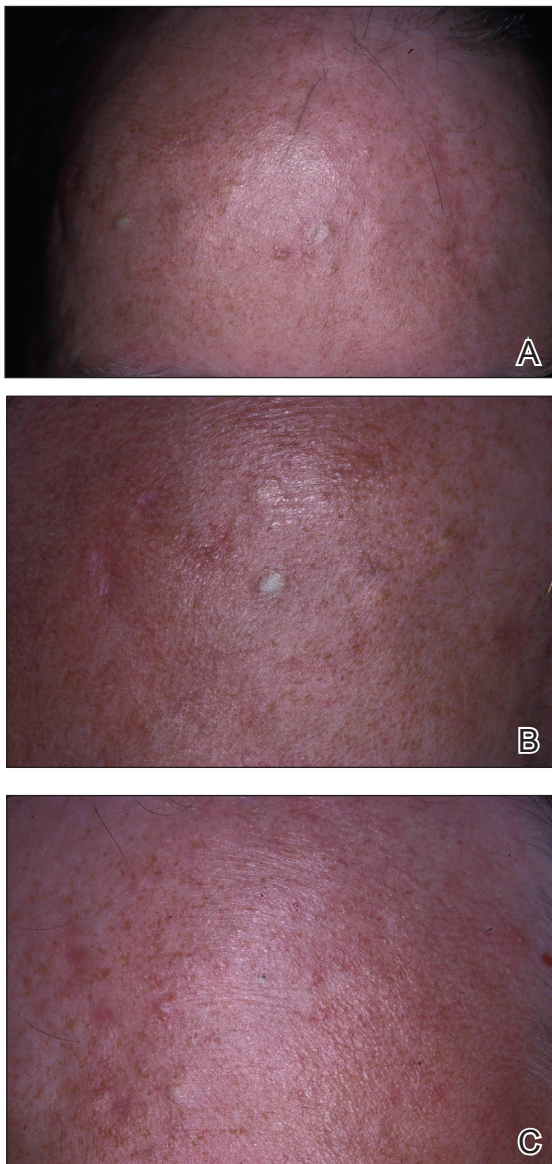
the mid dermis. Multiple small follicles arose from the cysts and showed differentiation toward germinative epithelium. The surrounding stroma was fibrotic and contained a patchy lymphocytic infiltrate. These histologic findings were consistent with trichofolliculomas (Figure 4).

### Comment

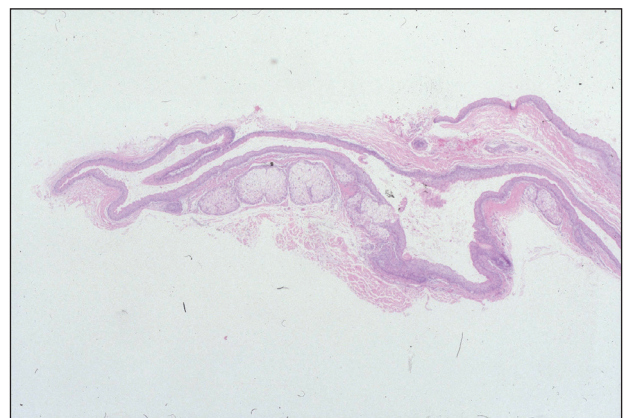
**Characteristics of SCM**—Steatocystoma multiplex is an uncommon condition characterized by the formation of asymptomatic, 0.2- to 2-cm, yellow to flesh-colored, soft, mobile papules or nodules on the trunk, extremities, axillae, genitalia, and/or chest. The lesions contain a clear or opaque, oily, milky or yellow, odorless fluid and most commonly are located on the anterior aspect of the chest. The face is not a commonly involved site in this condition. Six cases of a rare facial variant of SCM have been reported,<sup>11-16</sup> with lesions limited to the forehead in 3 cases.<sup>13-15</sup>

In 1937, Mount<sup>20</sup> credited Bozellini for describing the first case, though 3 cases reported in the late 1800s probably were SCM.<sup>21</sup> In 1899, Pringle<sup>22</sup> coined the term *steatocystoma multiplex* for this condition. It can be sporadic or have an autosomal-dominant inheritance pattern. Steatocystoma multiplex can occur at any age, though lesions develop most frequently in adolescence or young adulthood. There is no sex predilection.

Steatocystoma multiplex with pachyonychia congenita has been reported in a familial case.<sup>23</sup> Other findings reported in patients with SCM include ichthyosis, koilonychia, acrokeratosis verruciformis of Hopf and hypertrophic lichen planus, hidradenitis suppurativa, hypotrichosis, multiple keratoacanthomas, and rheumatoid arthritis.<sup>12,24-26</sup>

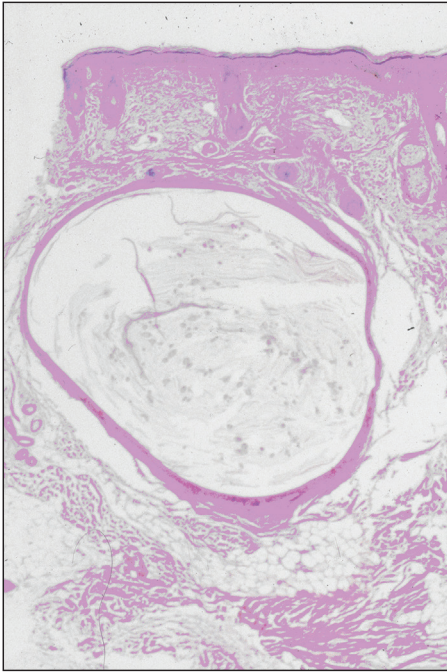


**FIGURE 1.** Multiple 0.3- to 1-cm, flesh-colored, yellow to blue, smooth, mobile papules on the forehead (A and B). A 3-mm, flesh-colored papule with a central black punctum on the center of the forehead (C).

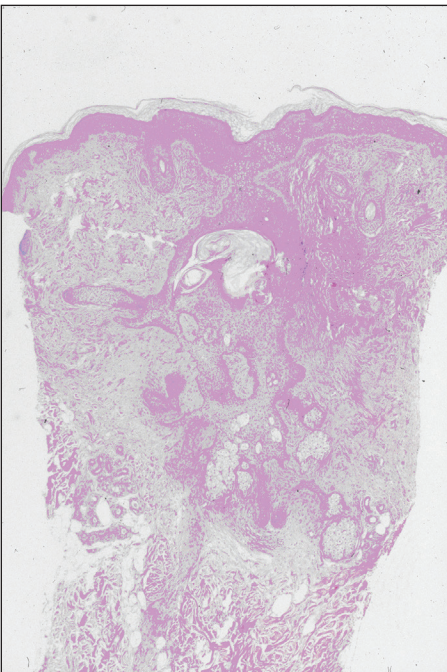


**FIGURE 2.** Photomicrograph of a steatocystoma multiplex lesion demonstrated a collapsed cystic space with parallel infoldings of the cyst wall. The cyst wall was composed of a squamous epithelium without a granular layer but with an eosinophilic corrugated lining. The cyst cavity contained scant homogeneous eosinophilic secretion. Mature sebaceous lobules emanated from the cyst wall (H&E, original magnification  $\times 10$ ).





**FIGURE 3.** Photomicrograph of eruptive vellus hair cysts demonstrated a small cystic structure lined by squamous epithelium with a granular layer in the mid dermis. The cyst cavity contained numerous small vellus hairs and laminated keratin (H&E, original magnification  $\times 10$ ).



**FIGURE 4.** Photomicrograph of a trichofolliculoma demonstrated a central dilated primary follicle that was contiguous with the surface epidermis and contained laminated keratin. Multiple small secondary follicles arose from the primary follicle. These secondary follicles showed differentiation toward germinative epithelium and hair bulb and papilla formation. The stroma was fibrotic (H&E, original magnification  $\times 10$ ).

Steatocystoma multiplex is a cyst lined by stratified squamous epithelium without a granular layer but with a thick eosinophilic cuticle. Mature sebaceous lobules are closely associated with the cyst wall. Steatocystoma multiplex arises from the sebaceous duct because the lining of the lumen is composed of undulating eosinophilic cuticle.

**Characteristics of EVHCs**—Eruptive vellus hair cysts, which were first described by Esterly et al,<sup>27</sup> can occur at any age but develop most frequently in adolescents or young adults. Sometimes the lesions are congenital or appear in childhood. There is no sex predilection. They can be sporadic or have an autosomal-dominant inheritance pattern.

Eruptive vellus hair cysts are asymptomatic, 1- to 2-mm, smooth, crusted, or umbilicated papules on the chest or arms and legs. Eruptive vellus hair cysts most commonly involve the anterior aspect of the chest. The lesions are flesh-colored to yellow, though they have a slate gray color in darker-skinned individuals. A rare facial variant has been reported in 2 patients of Asian descent.<sup>17</sup>

Eruptive vellus hair cysts are small cystic structures lined by a stratified squamous epithelium with a granular layer. The cyst cavity contains numerous small vellus hair shafts and laminated keratin. Eruptive vellus hair cysts originate from the infundibulum or less frequently the isthmus or infundibular-isthmic junction of the hair follicle.

**Characteristics of Trichofolliculomas**—Trichofolliculomas are solitary, 3- to 5-mm, flesh-colored papules that occur on the face. They are highly differentiated, benign, neoplastic proliferations of an actively trichogenic epithelium, with structural components reflecting all portions of the pilosebaceous unit. Trichofolliculomas consist of a central dilated primary follicle contiguous with the surface epidermis embedded in a fibrous stroma. Multiple small secondary follicles with varying degrees of follicular differentiation arise from the primary follicle.

**Co-occurrence of Lesions**—An association between SCM and EVHC has been recognized.<sup>5-10</sup> Steatocystoma multiplex and EVHC have similar clinical features but distinctive histologic features. They also have a similar age of onset, location/appearance of lesions, and mode of inheritance. Steatocystoma multiplex and EVHC can be distinguished by immunohistochemical techniques: SCM shows expression of keratin 10 and keratin 17, whereas EVHCs express only keratin 17.<sup>28</sup>

Steatocystoma multiplex and EVHC have only rarely been reported to occur together on the trunk. One case of SCM and EVHC occurring on the forehead has been described.<sup>3</sup> Other types of benign follicular neoplasms simultaneously developing in association with SCM or EVHC also are rare. Milia, SCM, and EVHC on the face and trunk have been reported in 1 family,<sup>4</sup> and facial steatocystoma associated with a pilar cyst and bilateral preauricular sinus was reported in 1 patient.<sup>19</sup> Although trichofolliculomas have not been reported to occur with SCM or EVHC, 2 related follicular neoplasms—trichoepitheliomas and trichoblastomas—have been

reported to occur in association with SCM on the face and chest and around the ears in 1 case.<sup>18</sup>

**Differential Diagnosis**—The clinical differential diagnosis includes multiple epidermoid cysts, dermoid cysts, Gardner syndrome, sebaceous adenomas, Muir-Torre syndrome, syringomas, milia, leiomyomas, lipomas, acneiform folliculitis, multiple familial and nonfamilial trichoepitheliomas, cylindromas, and angiofibromas.<sup>3,29</sup>

## Conclusion

Our patient represents a rare case of simultaneous occurrence of SCM, EVHC, and trichofolliculomas localized to the forehead. The patient had multiple neoplasms involving differentiation toward various regions of the pilosebaceous unit. This case gives further support to the hypothesis that these benign follicular neoplasms are closely related but are distinct conditions within the spectrum of the same disease process. They represent nevoid malformations of the pilosebaceous unit that can be sporadic or inherited in an autosomal-dominant pattern. Pure types of these lesions may represent one end of the spectrum, but in some patients, there are overlapping features or hybrids of each condition. Several biopsies from patients with multiple lesions should be performed to establish an accurate diagnosis.

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