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A case of folliculosebaceous cystic hamartoma: a rare and clinically indistinct lesion

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Abstract

We report a case of a slowly growing papule on the nasal bridge of an elderly woman. Histopathological findings revealed a very unusual type of folliculosebaceous cystic hamartoma with a retiform and primitive epithelial proliferation associated with the stromal component. Folliculosebaceous cystic hamartoma is an uncommon lesion that involves a cystically dilated follicle embedded in a stroma of distinct mesenchymal tissue. Because folliculosebaceous cystic hamartoma is a rare and clinically indistinct lesion, clinical photographs of the lesion are not widely available. A recent search on PubMed revealed less than 50 published articles on folliculosebaceous cystic hamartoma; among these manuscripts there was a scarcity of clinical images of the lesion. This disparity is to be expected as the lesion typically has a very indistinct presentation. We thus describe an inconspicuously appearing case of folliculosebaceous cystic hamartoma to bring attention to the indistinct clinical morphology of the lesion and to report a histopathologically uncommon variant.

Keywords: folliculosebaceous cystic hamartoma, papulonodular cysts

Introduction

First reported by Kimura et al. in 1991, folliculosebaceous cystic hamartoma (FSCH) is a rare, benign, slow growing, cutaneous hamartoma composed of follicular and sebaceous units [1]. FSCH is likely underdiagnosed owing to the indistinct clinical presentation [2]. It typically presents on the

face as a flesh colored papule or nodule that can be dome shaped, sessile, or pedunculated [3]. It is most commonly found on the face in the middle aged or elderly yet multiple reports of FSCH throughout the body have also been described [4]. Typically the size of the lesion is less than 2cm [4]. Clinical findings alone rarely lead to the diagnosis of FSCH [5]. Indeed the vague presentation of the lesion leads to a broad differential diagnosis often including melanocytic nevi, epidermal cysts, basal cell carcinoma, soft fibromas, and sebaceous hyperplasia [2].

Case Synopsis

A 75-year-old woman presented with a two-year history of a dome shaped 0.6×0.6cm exophytic flesh-colored papule on the left lateral side of the nasal bridge (Figure 1). The lesion had slowly grown over the past two years and had recently been irritated by **the patient's glasses. The papule was not tender to palpation.** The patient had no other cutaneous abnormalities and had no family history of similar lesions.



Figure 1. A 0.6×0.6 cm exophytic papule on the left lateral side of the nasal bridge.

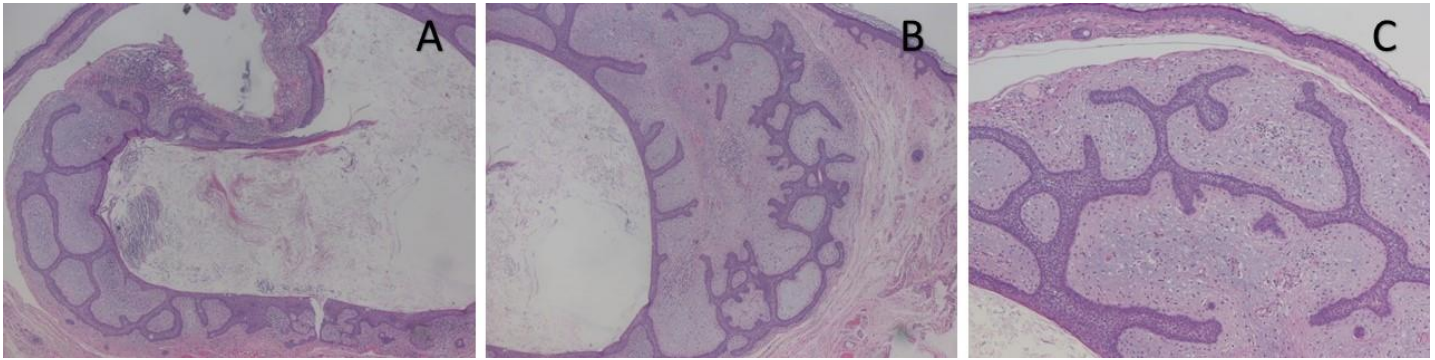


Figure 2. A) Low power (100 \times) view of H&E stained cystic structure with basaloid proliferations. Dense stroma clefting from the surrounding dermis is also appreciated. B) Lower power (100 \times) of another view of the cystic tumor revealing fibroepithelioma components including mucin infiltration. C) High power (200 \times) view showing fibroepithelioma features with abundant mucin in background stroma.

Histology revealed an exophytic mass comprised of a central dilated infundibular cyst connected to sebaceous lobules (Figure 2). The lesion was well-circumscribed and did not adhere to the surrounding tissue. The tumor showed features of a fibroepithelioma with a central cyst. The epidermis did not show any abnormalities. Basaloid nests with focal sebaceous differentiation were found in the dermis within the wall of the cyst (Figure 3). A distinct cleft was apparent between the fibroepithelioma components and the adjacent stroma. The lesion was removed by simple excision and the patient has had no complications or recurrence.

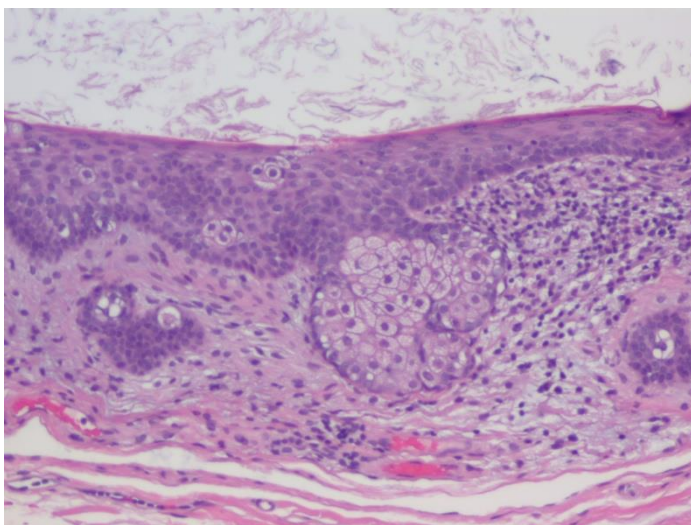


Figure 3. High power (400 \times) view revealing focal sebaceous differentiation with sebaceous islands within the wall of the cyst. Clefting is apparent between the tumor stroma and normal dermis.

Case Discussion

FSCH refers to a follicular proliferation in which sebaceous components are found within a cystic infundibular structure [5]. This follicular proliferation is itself embedded in a stroma of distinct mesenchymal tissues including adipocytes, blood vessels, collagen, and mucin as was prominent in our case [5]. FSCH completely involves the dermis with a **distinct cleft separating the lesion's stroma from the unaffected dermis** [5]. Histopathological exam of our **patient's lesion demonstrated** the cystic and follicular architecture associated with FSCH, yet also displayed a retiform and slightly primitive epithelial proliferation associated with the stromal component. In our knowledge, the case represents a rare variant of FSCH, as a retiform stromal epithelial proliferation has not been regularly described in this entity. Although not fitting all the features of most previously described FSCH, the lesion fits within the spectrum of this hamartoma better than any other tumor given the follicular and sebaceous features with cystic changes.

Histologically FSCH shares similarities with nevus lipomatosus superficialis, fibrofolliculoma, trichofolliculoma, and sebaceous trichofolliculoma [3]. Examination of fibrofolliculoma often reveals keratinous components in a cystically dilated folliculosebaceous unit. However, in fibrofolliculoma there is a paucity of mucin-filled-stroma and mature sebocytes that are classically found in FSCH [2, 6]. In contrast to FSCH, nevus lipomatosus superficialis

does not have epithelial components and instead consists completely of adipose cells [7]. In trichofolliculoma the infundibular cyst wall is connected to multiple vellus hair follicles, whereas in sebaceous trichofolliculoma the cystic wall is linked to mature sebaceous lobules [2, 4, 5]. Unlike both trichofolliculoma and sebaceous trichofolliculoma, FSCH is marked by a distinct mesenchymal stroma that is delineated by a cleft from the surrounding fibroepithelial components of the dermis [2, 4].

Immunohistochemical analysis regarding the histogenesis of FSCH remains unclear. Misago et al. suggested that nestin positive multipotent stem cells give rise to the mesenchymal changes, particularly the presence of adipocytes, found in FSCH [8]. In certain FSCH lesions, they found increased nestin expression in multiplying spindle cells and in sebaceous ducts [8]. Immature adipocytes, identified by positive staining with S100, were also found surrounding the sebaceous ducts, thus implying lipogenesis [2, 8]. In contrast to the view that FSCH uniquely arises from nestin positive stem cells, Suarez-Peñaranda et al. used antibodies

targeting CD10, CD34, p63, and Factor XIIIa, to highlight the shared immunohistochemical profile of the epithelial tissue in FSCH lesions with that of normal sebaceous structures [9].

We report this case not only because of the rarity of FSCH but also owing to the paucity of clinical photographs of the lesion. We encourage an open-minded approach when managing ordinary appearing papulonodular cysts. Clinicians should be aware that dermatoscopy can be helpful, as dermatoscopy of FSCH reveals orange-yellow globules and a whitish-yellow network [10]. Treatment centers on simple excision as there have been no reports of recurrence.

Conclusion

In summary, we describe this case to highlight the often-vague presentation of FSCH and to report a rare variant with retiform stromal epithelial proliferation. Clinicians should consider FSCH as part of their differential diagnosis when managing papulocystic lesions on the face.

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