A 45-year-old male presented with a single, asymptomatic swelling over the left cheek of 10-year duration. Cutaneous examination revealed solitary 7 mm × 7 mm skin colored to slightly hyperpigmented nontender nodule over the left malar area adjacent to the nose [Figure 1]. It was soft in consistency with limited mobility. The overlying skin was apparently normal with no punctum, surface telangiectasia, or hairs. Other mucocutaneous, general, and systemic examinations were within normal limits. The nodule was excised, and the histopathology revealed dilated cystic follicular infundibulum with multiple attached sebaceous lobules in the wall [Figure 2]. The surrounding stroma was fibroblastic that contained fibrillary bundles of collagen, increased number of dilated venules, clefts, and rudimentary hair follicles [Figure 3]. The dermis also showed numerous sebaceous lobules and adipose tissue.

**Question**

What is your diagnosis?
Folliculosebaceous cystic hamartoma (FSCH).

Discussion

FSCH is a rare nonneoplastic cutaneous hamartoma composed of follicular, sebaceous, and mesenchymal elements. It is also considered as a variant of trichofolliculoma by a few authors.[1] FSCH was initially reported in 1991.[1] It usually affects adults, but has also been reported in children as well as the elderly, including a few congenital cases.[2,3] Its occurrence is slightly more common in females compared to males[2] and has been observed to have Asian racial predilection.[4] It commonly affects the head and neck area, especially on or around the nose as in our case. Rarely, it may occur on genitals or breast.[4,5] FSCH usually presents clinically as an asymptomatic solitary papule, sessile or pedunculated nodule or polyp, rubbery to firm in consistency, with normal overlying skin with no surface opening or central umbilication. The size of the tumor is usually <3 cm, although giant variants have been reported.[2-3] Rare associations such as rosacea, nevus lipomatosus superficialis, and port-wine stain have been reported.[5]

The true origin of FSCH is controversial. The most accepted theory is that it is a distinct hamartomatous adnexal lesion, predominantly of the sebaceous glands. Another popular view is that FSCH represents the telogen stage of sebaceous trichofolliculoma (STF), a pilo adnexal tumor.[1] However, the occurrence of congenital FSCH refutes this school of thought.[1,3] In contrast to FSCH, STF is more common in children and presents clinically as a depressed lesion with an ostium containing protruding hairs. The other differential diagnoses include intradermal nevus, sebaceous cyst, neurofibroma, dermoid cyst of nose, and nodular basal cell carcinoma.[1]

The histological diagnostic criteria for FSCH[2] include (1) an infundibular cystic structure with several sebaceous lobules attached to it via sebaceous ducts, (2) compactly laminated fibroplasia around the epithelial units, (3) mesenchymal component around the epithelial units comprising fibrillary bundles of collagen, adipocytes, and many small venules, (4) clefts between the fibroepithelial units and surrounding altered stroma, and (5) dermal and subcutaneous location of the tumor. Rare pathological features of FSCH include the presence of neural and smooth muscle components, hair shaft fragments, and perifollicular mucinosis.[1,5] The closest differential diagnosis of FSCH is STF, which can be differentiated by the presence of central cystic cavity with connection to epidermis and contains hair shafts, sebaceous lobules, and secondary hair follicles. Stromal mesenchymal components and clefts are notably absent in STF histopathology.

Immunohistochemistry of FSCH shows p63 staining in all epithelial components, while few sebocytes show CD10 positivity. CD34- and Factor XIIIa-positive cells have been demonstrated in the stroma. Staining for androgen and alpha-estrogen receptors has also been reported.[5]

Excision is the primary modality of treatment with no reports of recurrence. In conclusion, this case highlights the importance of considering a differential diagnosis of FSCH while dealing with nodulocystic lesions situated in and around the nose.

Learning points

• FSCH is a cutaneous hamartoma composed of predominantly sebaceous glands
• It usually presents in adults in the head and neck area as solitary papule or nodule with normal overlying skin
• Histopathology of FSCH is characterized by an infundibular cystic structure with several sebaceous lobules attached to it and surrounding mesenchymal elements with clefts
• It should be distinguished from STF which usually presents in children as a depressed lesion with an ostium.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.
References

1. Hamada M, Kiryu H, Satoh E, Moroi Y, Urabe K, Furue M. A case of genital folliculosebaceous cystic hamartoma with an unique aggregated manifestation. J Dermatol 2006;33:191-5.

2. El-Darouty MA, Marzouk SA, Abdel-Halim MR, El-Komy MH, Mashaly HM. Folliculo-sebaceous cystic hamartoma. Int J Dermatol 2001;40:454-7.

3. Cole P, Kaufman Y, Dishop M, Hafez DA, Hollier L. Giant, congenital folliculosebaceous cystic hamartoma: A case against a pathogenetic relationship with trichofolliculoma. Am J Dermatopathol 2008;30:500-3.

4. Badr A, Lakshmiah GR. Folliculosebaceous cystic hamartoma of the nipple: A case report. J Cutan Pathol 2009;36:597-600.

5. Suarez-Peñaranda JM, Vieites B, Ramírez-Santos A, Fernández-Redondo V, Toribio J, Del Rio E, et al. Clinicopathological and immnuohistochemical findings in a series of folliculosebaceous cystic hamartoma. J Cutan Pathol 2009;36:251-6.