Folliculosebaceous cystic hamartoma: an unusual histopathological entity: a case report

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Abstract

The skin covers our body and exhibits a complex structure that is adapted to the different body areas. The scalp skin is distinctive, and the histopathological alterations in its diseases also present distinctive features. We present the clinical case of a 28-year-old female patient, with no significant medical history. She reported a nodular tumor on the left parietal region of the scalp, which had been present for 10 years and showed slow growth. The patient had no major discomfort other than an occasional discharge of scant whitish material upon applying pressure. Intralesional corticosteroids were administered but had no effect. Over the following year, the discomfort increased, leading to the decision to remove it. The anatomopathological study found dilated follicular hairy structures in the dermis filled with keratin and debris, surrounded by multiple sebaceous glands that flow there, all surrounded by a characteristic mesenchymal cells population, diagnosing a folliculosebaceous cystic hamartoma.

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Introduction

Folliculosebaceous cystic hamartoma (FSCH) is an unusual, benign, and slow growing cutaneous hamartoma composed of follicular and sebaceous units. The first description of this distinct entity was made by Kimura et al. in a series of five cases.[1] FSCH, trichofolliculoma (TF), trichodiscoma, fibrofolliculoma, and pilomatricoma are considered hamartomas of pilosebaceous origin.[2] FSCH can be found in all age groups; however, most cases occur in adults, with females being more affected than males.[3] The age of patients ranges from 4 to 84 years.[4]

The lesions typically present as single nodules or papules, with a predilection for the central area of the face and scalp.[5] They have a skin colored appearance and can be sessile or pedunculated.[6] Reports of FSCH in other body areas exist.[7] Except for rare giant cases of FSCH, most lesions rarely exceed 2 centimeters in size.[8]

The main histopathological criteria include the presence of an epithelial component in the form of a cystic infundibular structure, with radially connected lobules of sebaceous glands, and a mesenchymal component represented by collagenous stroma, adipocytes, and small blood vessels.[9] It usually involves the entire dermis and presents a cleft separating the lesion from the unaffected dermis.[10]

FSCH is often underdiagnosed due to its nonspecific clinical presentation,[11] and clinical information alone rarely leads to a definitive diagnosis.[10] A histopathological study is required for definitive confirmation.[12]

Case Report

A 28-year-old female patient with no significant medical history presented with a solitary nodular tumor on the left parietal

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region of the scalp. The tumor had been present for ten years, with chronic and slow growing evolution. Initially, the patient reported no associated symptoms other than the lesion itself, and an intralesional corticosteroid was administered without apparent improvement. Subsequently, the lesion increased in size, and upon acupressure, a small amount of whitish material would be released approximately every three months, without concomitant pain. Over the following year, the patient experienced increased discomfort in the area, mild pain upon pressure, and an increase in size, leading her seeking medical consultation with a decision to excise the lesion. Physical examination showed an oval shaped, skin colored, well defined, firm lesion with regular borders, with no other particularities. Macroscopic studies described a diamond shaped piece of scalp skin measuring 12x8x7 millimeters, with a brownish color and an elastic consistency. There was a nodular lesion on its deep side, measuring 7x6x4 millimeters, with well defined borders, an oval shape, a whitish color, and an elastic consistency (Figure 1 A). Upon sectioning, the cut surface appeared heterogeneous with a white yellowish color (Figure 1 B), and the entire specimen was processed. Microscopic observation identified a lesion composed of a disordered mixture of histological elements typical of the scalp in the deep dermis. These included dilated infundibular portions of hair follicles containing keratin and cellular debris but lacking hair. Multiple sebaceous glands converged radially into these dilated spaces, draining into them through their ducts (Figure 1 C). There were various inflammatory cells, forming accumulations that tended to surround the dilated hair follicles (Figure 1 D). These cells were immersed in a diverse population of mesenchymal lineage products, including fibroblasts, collagen with dense areas, scarce adipocytes, neuro-like areas, and blood vessels (Figure 1 E). None of the described cells showed alterations suggestive of dysplasia or neoplasia, displaying typical characteristics. Some tissue clefts clearly separated the lesion from the normal dermis (Figure 1 F). Multiple histological sections did not reveal any connection or opening of the cysts to the surface of the scalp. The combination of clinical, macroscopic, and microscopic criteria led to the diagnosis of FSCH, a rather uncommon entity. Follow-up and monitoring for up to 9 months after surgery did not demonstrate recurrence or complications.

Discussion

The skin is an extensive and complex tissue that changes and adapts to the different areas of the body it covers. There is a notable histological difference between the skin that covers the

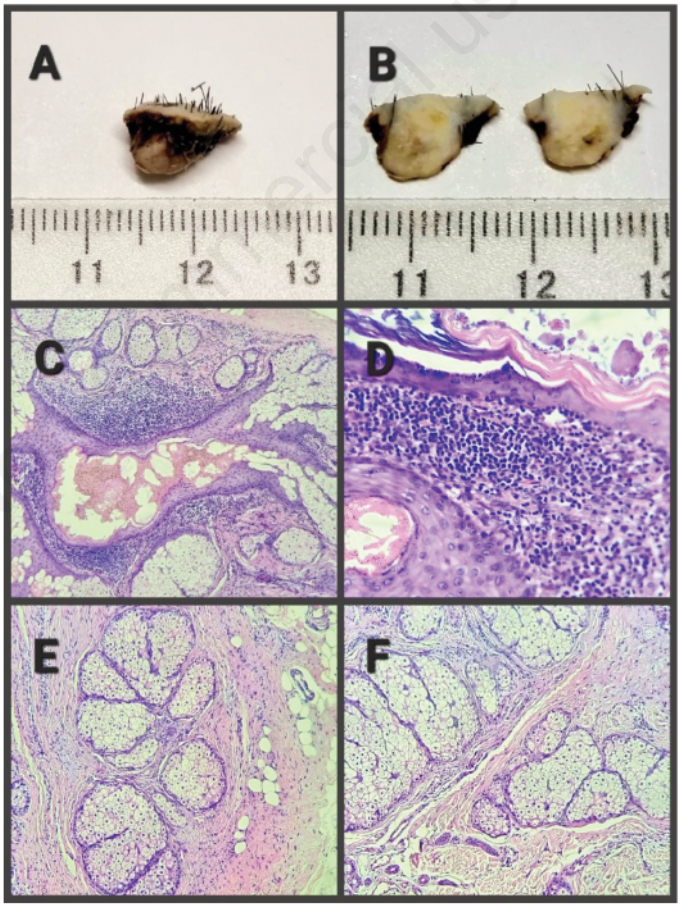

Figure 1. (**A**) Lateral view of the specimen, showing a welldefined oval shaped lesion in the deep dermis. (**B**) Heterogeneous cut surface with whitish yellowish areas. (**C**) Dilated infundibular portion of hair follicle containing keratin and debris, surrounded radially by sebaceous glands that drain into the dilation (40X). (**D**) Various inflammatory cells in relation to the dilated infundibular portion, with a noticeable superficial granular layer of the epithelium (100X). (**E**) Mesenchymal elements (fibroblasts, collagen, adipocytes) surrounding the epithelial component of the lesion (40X). (**F**) Central cleft separating the lesion (top left) from the normal dermis (bottom right) (40X).

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scalp, chest, genital area, palms, and soles, to name a few regions. Furthermore, the histological difference reflects the functional utility of the studied area.

Lesions that occupy space are often referred to as "tumors", a generic term that should not be mistaken as synonymous with neoplasia, which is characterized by uncontrolled cellular proliferation and can be either benign or malignant. With this previous consideration, a hamartoma is defined as the presence of disordered mature histological structures specific to the tissue in which it occurs and is not a neoplasm per se. It simply occupies space and tends to increase in size, which can be alarming for both the patient and the healthcare professional.

FSCH represents a disordered proliferation of structures found in the skin, composed of epithelial elements (follicular infundibulum, sebaceous glands) and mesenchymal elements (fibroblasts, collagen, adipocytes, blood vessels, inflammatory cells). It constitutes a slow growing lesion without other distinctive clinical features.

Precisely, the lack of distinctive clinical features and the unusual frequency of the condition allows for extensive and varied presumptive diagnoses. All of the above, combined with a large amount of cutaneous pathology, justifies the presentation of this case in order to guide a definitive diagnostic approach based on clinical pathologic knowledge.

The lesion exhibits very slow growth, mainly due to the accumulation of keratin and debris in the dilated infundibular portions, as well as the presence of multiple sebaceous glands, all within the proliferation of characteristic mesenchymal elements. Indeed, the presence of mesenchymal derived elements is an important differential criterion, as will be discussed below.

Ansai et al. reported the largest published series of FSCH cases, including 153 cases, of which 92 cases affected males and 61 cases affected females. The typical presentation was that of a papule or nodule, with normal skin color, measuring a few millimeters in diameter, predominantly located on the skin of the face, particularly the nose, in middle aged and elderly patients.[7]

Setting aside many cutaneous lesions with which it shares similarities, the main consideration in the differential diagnosis is sebaceous trichofolliculoma. This lesion presents as an oval shaped lesion with depression and frequent communication with the exterior, along with the presence of abnormal hair in the dilated infundibulum. Both criteria are absent in FSCH.

Furthermore, Schulz and Hastschuch proposed that FSCH represented a late stage of trichofolliculoma, but this hypothesis would not explain the congenital and neonatal cases of FSCH.[13]

In summary, sebaceous trichofolliculoma macroscopically exhibits an oval shape with flattening and superficial communication and, histologically, it shows rudimentary hairs in the infundibular cysts, with an absence of the mesenchymal derived elements described in FSCH.

Conclusions

The FSCH is a hamartomatous, non-neoplastic lesion that affects the dermis and represents a mixture of histological elements found in the skin, but in a disordered manner. It is characterized by the presence of epithelial tissues (dilated pilosebaceous infundibula, radiating sebaceous glands) and mesenchymal elements (fibroblasts, collagen, adipocytes, neuroid elements, blood vessels, and inflammatory cells). During its slow progression, these elements occupy space and cause an increase in the size of the lesion.

This lesion usually does not cause characteristic clinical symptoms, which is why it has received multiple presumptive diagnoses and treatments before excision. The definitive diagnosis of FSCH is provided by histopathological examination, based on the previously described microscopic criteria and a thorough differential diagnosis.

The rarity of the disease makes it poorly recognized and often confused with other skin conditions that occupy space. The purpose of this report is to gain a better understanding of this mysterious and unusual entity.

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