A Case of Folliculosebaceous Cystic Hamartoma

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Folliculosebaceous cystic hamartoma (FSCH) is a recently-recognized cutaneous hamartoma composed of follicular, sebaceous and mesenchymal elements. We describe an unusual case of FSCH in a 61-year-old male, who had a relatively large, 3 × 2.5 cm sized, smooth subcutaneous nodule on the occipital area of the scalp, an uncommon location for FSCH.
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Key Words: Folliculosebaceous cystic hamartoma, Subcutaneous, Occiput

INTRODUCTION

FSCH is a rare, distinct, cutaneous hamartoma of follicular, sebaceous and mesenchymal components[1,2]. It usually presents as a small, exophytic papule or nodule on the central part of the face. Histopathologically, it is characterized by infundibulo-sebaceous cystic proliferation with specific mesenchymal changes including packed fibrillary bundles of collagen, cleft formation between fibroepithelial units and surrounding stroma, sparsely distributed adipocytes, and increased numbers of small venules.

Since FSCH was first described in 1991, there have been approximately 30 reported cases including 2 cases of a giant variant in the English literature and 6 cases of FSCH in the Korean literature (Table 1)[1-12].

We describe a clinically interesting case of FSCH in a 61-year-old male, whose lesion was relatively large, reaching to 3 cm in diameter, and developed on the occiput.

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CASE REPORT

A 61-year-old man presented with a 2-year history of a large, subcutaneous nodule on the scalp. Physical examination revealed an asymptomatic, 3 × 2.5 cm sized, firm, subcutaneous nodule on the occipital area of the scalp (Fig. 1). Under the clinical impression of a "pilar cyst", "epidermal cyst" or "lipoma", the lesion was removed surgically. On histopathologic examination, there was found to be a cystically-dilated follicular infundibulum with numerous sebaceous lobules around it (Fig. 2). Mature sebaceous lobules were attached, through sebaceous ducts, to the dilated infundibulum. The stroma was composed of compactly-laminated

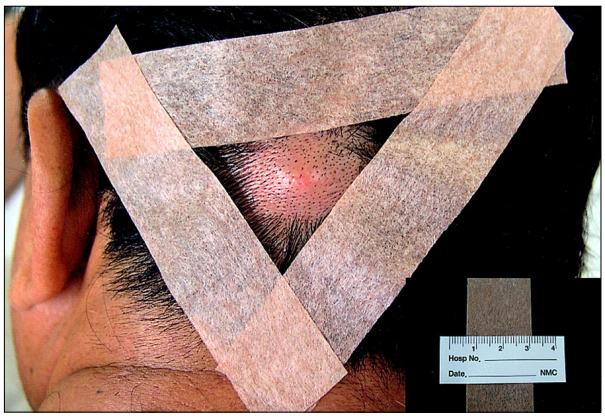

Fig. 1. A smooth, 3 × 2.5 cm sized, subcutaneous nodule on the scalp.

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Table 1. Summary of FSCH in the Literature

Cases	Age/sex	Location	Size	Duration
Kimura et al.[1]				
Case 1	42/F	Scalp	1.0 × 1.0 cm	3 years
Case 2	50/M	Nose	1.3 × 1.3 cm	6 years
Case 3	58/M	Forehead	0.6 × 0.6 cm	6 years
Case 4	48/F	Cheek	1.0 × 1.0 cm	15 years
Case 5	85/F	Nose	0.9 × 0.8 cm	Unclear
Donati et al.[3]	32/F	Ear	1.0 cm in diameter	3 years
Yamamoto et al.[4]	24/F	Back	7.0 × 4.5 cm	24 years
Aloi et al.[5]	8/M	Auditory canal	1.0 × 0.7 cm	Since birth
Bolognia et al.[6]	34/F	Labia major	4 cm	-
Sturtz et al.[7]	32/F	Arm	15 cm at greatest dimension	Several years
Lee et al.[8]				
Case 1	23/M	Scalp	Pea-sized	1 year
Case 2	46/M	Face	Pea-sized	Several years
Ahn et al.[9]	34/F	Face	0.5 × 0.6 cm	2 years
Choe et al.[10]	39/F	Back	0.8 cm	8 years
Kwon et al.[11]	37/M	Face	0.5 × 0.5 cm	1 year
Jang et al.[12]	36/F	Scalp	-	3 months
Our case	61/M	Scalp	3 × 2.5 cm	2 years

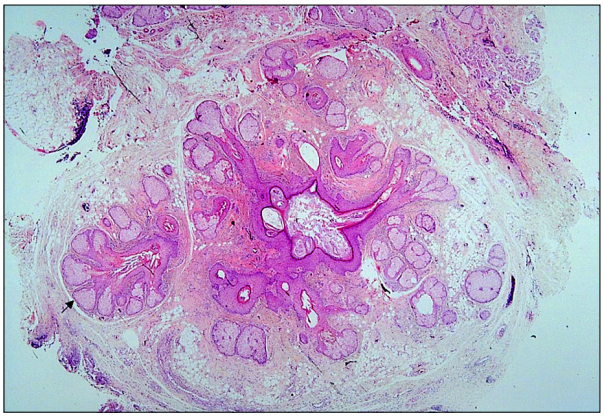

Fig. 2. Showing a cystically-dilated folliculosebaceous unit in the center (H&E, × 20). The tumor was separated by a cleft from the surrounding normal tissue (arrow).

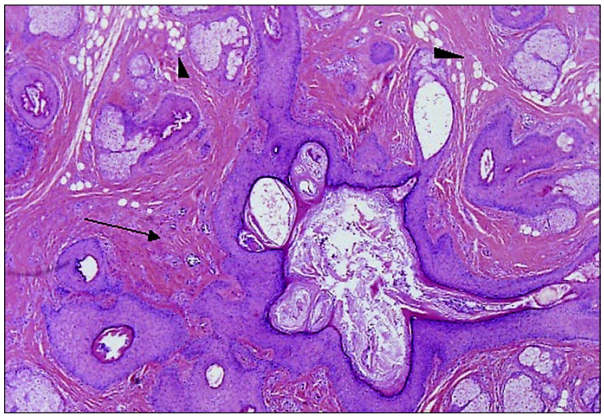

Fig. 3. Mesenchymal stromal components demonstrate dense fibrous tissue with increased blood vessels (arrow) and foci of adipocytes (arrow heads) (H&E, × 100).

fibrotic tissue, adipocytes, and small vessels (Fig. 3). The tumor was separated by a cleft from the surrounding, compressed normal tissues in the dermis.

On the basis of the above findings, a diagnosis of folliculosebaceous cystic hamartoma was made. There was no evidence of recurrence 5 months after removal of the tumor.

DISCUSSION

Clinically, FSCH is an asymptomatic, slow-growing, small papule or nodule usually occurring on the head or neck areas[1,2], but other uncommon locations such as the upper back, forearm, labia majora, and the scalp have been reported[3-12]. The tumor size is usually 0.5-1.5 cm in diameter, but 2 cases of a giant variant have been reported[4,7]. One was a 7.5 cm in diameter, exophytic, lobulated mass on the back and the other one was a 15 cm-sized multinodular plaque on the arm.

Originally, the term hamartoma meant a benign tumor-like nodule composed of an overgrowth of mature cells and tissues which normally occur in the affected area. It usually presents at the time of birth, and does not show active growth after birth. Hence, hamartoma used in FSCH is actually a misnomer. However, the term "hamartoma", Kimura[1] used in his original article, is intended to convey the fact that a jumble of tissue elements is normally present at the site of the lesion. Thus, FSCH is thought to be just merely one of the epidermal appendage tumors appearing in postnatal life and also having a potential for size enlargement. The tendency of tumor enlargement might be stronger in unusual areas such as the back, the labia, and the extremities and can result in morphologic changes including polypoid, plaque- like, or multinodular growth.

Although it was not a giant lesion and had none of the above morphologic changes, the tumor in our patient had continuously grown to 3 cm in diameter by the time of diagnosis. Because of its rarity, the importance of a large or giant variant is still unknown except for the cosmetic or therapeutic aspects.

Since FSCH carries no specific clinical characteristics, various tumors may be suspected prior to histopathologic examination. While intradermal nevus, neurofibroma, pilomatricoma and other adnexal tumors are clinical differential diagnoses for a typical FSCH lesion, pilar cyst, epidermal cyst or lipoma were suspected for our scalp lesion. Whatever the clinical impressions may be, the correct final diagnosis of FSCH can be made without difficulty from the well-known histopathologic features.

We report an unusual case of FSCH showing a relatively large, 3 × 2.5 cm sized, smooth, subcutaneous nodule on the occipital area of the scalp, an uncommon location for FSCH.

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