

CASE REPORT

A Case of Giant Folliculosebaceous Cystic Hamartoma

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Clinically, folliculosebaceous cystic hamartoma (FSCH) lacks distinct features, but it has been reported as an asymptomatic, 1- to 3-cm, dome-shaped nodule on the face. Histopathologically, FSCH is characterized by adnexal and folliculosebaceous cystic proliferation with various mesenchymal changes. This case presented an unusually large, small-fist-sized mass in the right mandibular area that was accompanied by intermittent itching. Histopathologic findings demonstrated appropriate features of FSCH. We present an interesting case of giant FSCH in a 48-year-old female. (*Ann Dermatol (Seoul)* 21(1) 63~65, 2009)

-Keywords-

Folliculosebaceous cystic hamartoma, Giant

INTRODUCTION

Kimura et al¹ originally reported eruptive folliculosebaceous cystic hamartoma (FSCH) in 1991. Clinically, FSCH is usually a solitary, 0.5- to 1.5-cm, flesh-colored, sessile or pedunculated papule on the head, especially on the central part of the face and nose².

Because few cases of giant FSCH have been reported, there is no distinct definition of "giant". According to previous reports³⁻⁶, these lesions range from 5 cm to 23 cm in diameter. Along with its larger size, and unlike ordinary FSCH, giant FSCH occurs on areas other than the face.

CASE REPORT

A 48-year-old female presented with a pruritic skin-colored soft tumor on her right lower cheek that had appeared as

small papules 20 years earlier and had been growing gradually. The skin lesion was a skin-colored, cerebriform mass, sized 10×8×5 cm, on the right mandibular area (Fig. 1A). Its smooth surface contained scattered, dilated hair follicle-like pores with a waxy discharge (Fig. 1B).

Histological examination showed numerous sebaceous lobules radiating from cystic follicular structures with mesenchymal changes. Numerous mature sebaceous lobules surrounded the cystic structures. The cystic structures were lined with stratified squamous epithelium and showed well-developed, dilated infundibular portions of hair follicles with infundibular keratinization. There were some increases of muscle components near the sebaceous lobules (Fig. 2A), but no secondary hair follicles. The folliculo-cystic structures were wrapped in densely laminated collagen bundles with various vascular proliferation (Fig. 2B). In immunohistochemical staining, muscle components were immunoreactive to desmin and smooth muscle actin (Fig. 3A). Increased vascular structures showed reactivity to CD31 (Fig. 3B), but tumor stromal cells were negative for S-100 protein and neurofilament. The size of the lesion decreased slightly after treatment with isotretinoin (30 mg/d) for 3 months, but returned to its original size after cessation of drug. We recommended that the patient undergo excision and skin grafting but she did not want further treatment.

DISCUSSION

FSCH is a distinct cutaneous hamartoma formed from epithelial and stromal elements. Among the cases of FSCH in the literature¹⁻²⁰, only a few cases of giant FSCH have been reported (Table 1)³⁻⁶. The clinical features of our case were similar to those of the reported giant FSCH cases, except for cerebriform surface and chin involvement. FSCH is slightly more common in females. The sites of predilection are the nose, cheeks, forehead, and scalp. In most cases, the lesion does not exceed 3 cm in diameter⁷. The four cases in the literature described as "giant" ranged

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Fig. 1. (A) The skin lesion is a skin-colored, cerebriform mass, 10.0×8.0×5.0 cm, in the right mandibular area. (B) The smooth surface has scattered, dilated hair follicle-like pores with a waxy discharge.

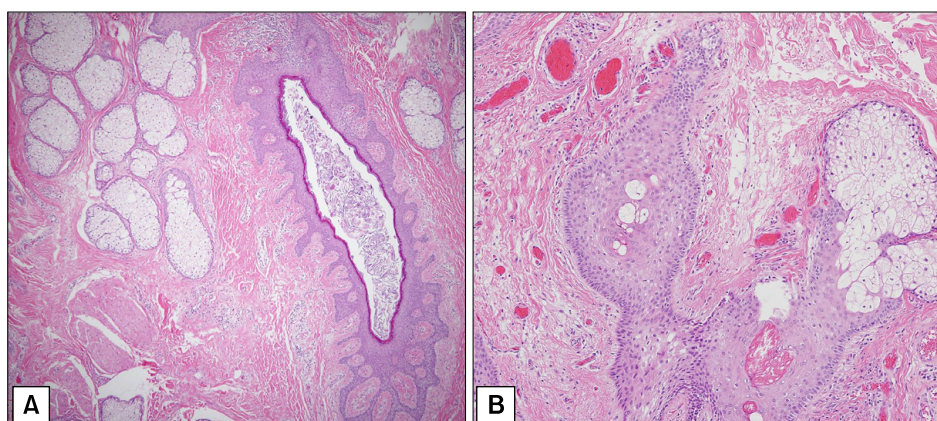


Fig. 2. (A) Numerous mature sebaceous lobules surround the cystic structures. There is some increase of muscle components on the left lower quadrant (H&E, ×40). (B) Increased numbers of small venules are shown around the fibroepithelial units (H&E, ×100).

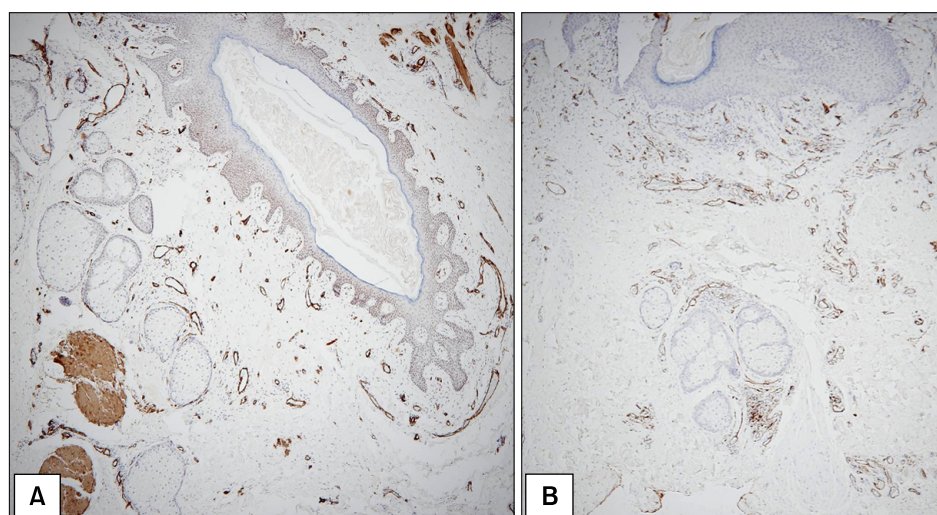


Fig. 3. (A) A slight increase of smooth muscles is shown around fibroepithelial units (SMA, ×40). (B) Increased numbers of small venules are shown around fibroepithelial units. Some of these vessels are congested with red blood cells (CD31, ×40).

from 5 cm to 23 cm in diameter. The lesions in giant FSCH cases occurred in various sites, such as genital areas, upper extremities, and upper back.

FSCH has distinct, unique, classical histopathologic features. Kimura et al¹ established the histological criteria for diagnosis of FSCH as: 1. an infundibular cystic struc-

Table 1. The clinical features of the reported cases of giant folliculosebaceous cystic hamartoma

Author	Sex/Age	Site	Size (cm)	Shape
Sturtz et al ³	F/32	Left upper arm	16.7×8.6×1.2	Multinodular, plaque-like
Yamamoto et al ⁴	F/24	Upper back	7.0×4.5×2.5	Polypoid, exophytic
Hamada et al ⁵	F/40	Genital area	5.0×3.0	Aggregated nodules
Brücher et al ⁶	M/74	Genital area	23.0×16.0	Coalesced nodules
This case	F/48	Right mandible	10.0×8.0×5.0	Polypoid, cerebriform

ture attached to sebaceous lobules via sebaceous ducts; 2. laminated fibroplasias around the entire fibroepithelial units; 3. mesenchymal changes around fibroepithelial units; 4. clefts between fibroepithelial units and surrounding altered stroma; 5. confinement of the process primarily to the dermis. Several variations in FSCH reflect the predominance of either epithelial or non-epithelial components. Fogt and Tahan⁸ reported large expanses of tufted aggregates, separated or enveloped within the epithelial units. Donati and Balus⁹ presented a case with neural proliferation, while Aloï et al¹⁰ described copious mucin deposits in a fibrillary stroma. Our case involved several mesenchymal changes, such as laminated collagen, vascular proliferation, and slightly increased muscle components. Because of its peculiar histopathologic features, diagnosis of FSCH is not difficult. But, in some cases, FSCH must be differentiated from sebaceous trichofolliculoma. Sebaceous trichofolliculoma is associated with pilosebaceous hyperplasia and overgrowth⁵. It presents as a central cystic cavity, usually containing a hair shaft, with sebaceous lobules and secondary hair follicles. However, sebaceous trichofolliculoma has minimal surrounding stromal proliferation. In FSCH, there are no secondary hair follicles and hair shaft in the cystic cavity. Mesenchymal change in FSCH is another point of differential diagnosis. Giant FSCH is less common than ordinary FSCH and this particular case occurred on the face unlike other giant FSCH cases. We need to gather more cases to determine how FSCH and giant FSCH differ from each other in clinical features and prognosis.

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