

## *A Case of Folliculosebaceous Cystic Hamartoma on the Right Nasal Ala*

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*Folliculosebaceous cystic hamartoma is a rare cutaneous hamartoma comprised of follicular, sebaceous, and mesenchymal elements. It usually presents as an exophytic papule or nodule on the central part of the face. The histological findings are intradermal cystic structures lined by infundibular epithelium, numerous sebaceous lobules radiating from cystic structures and a surrounding stroma composed of mesenchymal changes, including variable proportions of fibrous, adipose, vascular and neural tissues. We herein report a case of folliculosebaceous cystic hamartoma in a 73-year-old man who presented with a skin-colored, dome-shaped papule on the right nasal ala. (Ann Dermatol (Seoul) 19(4) 170~172, 2007)*

Key Words: Folliculosebaceous cystic hamartoma

### INTRODUCTION

Folliculosebaceous cystic hamartoma (FSCH) is a term first used by Kimura et al.<sup>1</sup> in 1991 to describe a cutaneous lesion of follicular, sebaceous and mesenchymal origin. It is usually found on the head, mainly around the nose, as an exophytic papule or nodule, occasionally with hairs through pores<sup>1</sup>.

The histogenesis of FSCH is still unclear. Some authors denied that FSCH is a disease entity<sup>2,3</sup>, whereas others regarded it as a distinct adnexal tumor despite the controversial nosology<sup>1,4-8</sup>. The controversy exists largely in the relationship between FSCH and sebaceous trichofolliculoma. The most characteristic feature of FSCH is the change in the mesenchymal stroma, which shows increased small vessels, adipocytes and a neural component.

### CASE REPORT

A 73-year-old man presented with a 40-year

history of a papule on the right nasal ala. Physical examination revealed a 1.0 cm, skin-colored, firm and sessile papule, which had neither pores nor hairs (Fig. 1). Under the clinical impression of a "soft fibroma", the lesion was excised. Histopathologic examination revealed that the epidermis showed no abnormal findings and was not connected to the dermal lesion. In the dermis, there were keratin-

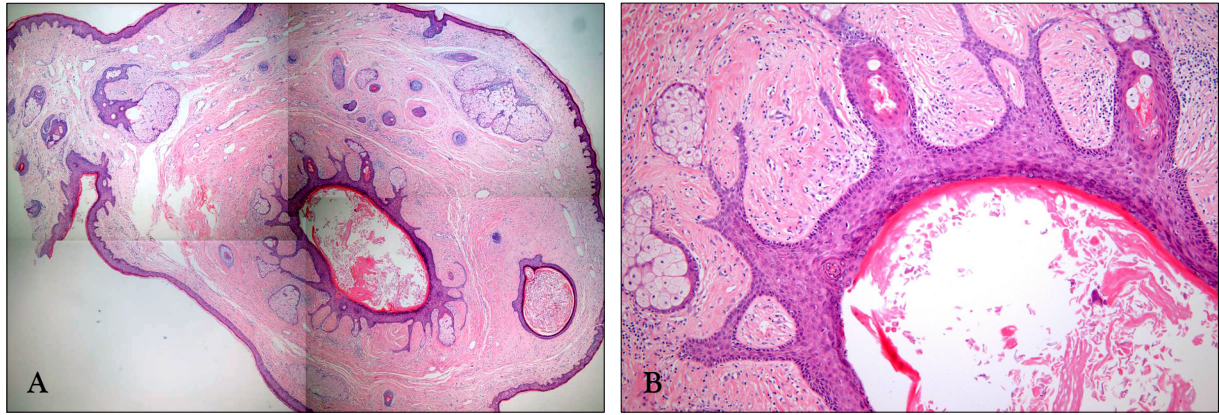


**Fig. 1.** A 1.0 cm, skin-colored, exophytic nodule is seen on the right ala nasi.

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**Fig. 2.** (A) This section is showing typical lesion, consisting of folliculosebaceous element and compactly laminated stroma separated by cleft from the surroundings (H&E stain,  $\times 10$ ). (B) Higher magnification of Fig. 2A shows numerous sebaceous lobules attached through sebaceous duct to the infundibular structure (H&E stain,  $\times 40$ ).

filled, dilated infundibular structures and numerous sebaceous lobules attached through sebaceous ducts to them as well as an isolated lobule in the stroma. The sebaceous lobules had normal features of sebocytes with clear cytoplasm and central nucleus. The stroma demonstrated a compactly laminated fibrosis and diffuse proliferation of small vessels. However, there was no increase in adipocytes and/or neural tissues. The tumor was separated by a cleft from the surrounding compressed normal tissue in the dermis (Fig. 2A, B).

On the basis of the histological findings, a diagnosis of folliculosebaceous cystic hamartoma was made.

There was no evidence of recurrence in 2 months after removal.

## DISCUSSION

Folliculosebaceous cystic hamartoma (FSCH) is a distinctive cutaneous hamartoma of follicular, sebaceous and mesenchymal components<sup>1,4</sup>. This hamartoma is characterized by infundibulosebaceous 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<sup>1,4</sup>. Since Kimura et al.<sup>1</sup> first described FSCH in 1991, over 30 cases have been reported in the literature, and about 8 cases have been reported in Korea<sup>13</sup>. The typical FSCH presents as a solitary, flesh-

colored, smooth-surfaced papule or nodule with a sessile or pedunculated shape<sup>1</sup>. This hamartoma usually occurs on the head, especially on the central part of the face and nose<sup>4</sup>. The size of the lesions in all reported cases on the head and neck did not exceed 25 mm in diameter. In three extremely large growths (giant variants), the lesions were located on the upper back, labia majora and upper arm<sup>5,9-10</sup>.

Physical examination shows FSCH lesions to be asymptomatic, usually rubbery to firm in consistency. They tend to grow slowly without any change in color or texture over time<sup>11</sup>. FSCH lacks distinctive clinical features and the initial diagnoses in all reported cases included disorders other than FSCH, such as intradermal nevus, sebaceous hyperplasia, basal cell carcinoma, lipoma and neurofibroma<sup>11</sup>.

Histopathological features of FSCH include (a) an infundibular cystic structure adjacent to the sebaceous glands, (b) compactly laminated fibroplasias around the entire epithelial component of fibroepithelial units, (c) mesenchymal changes around fibroepithelial units that include fibrillary bundles of collagen, adipocytes and an increased number of small venules, (d) clefts between the fibroepithelial units and the surrounding altered stroma and the adjacent normal skin structures and, (e) confinement of these processes primarily to the dermis<sup>1</sup>. Our case fulfilled the above criteria for FSCH except adipocytes or neural tissues.

The histopathologic differential diagnoses include sebaceous hyperplasia, sebaceous trichofolliculoma, fibrofolliculoma, perifolliculoma and dermoid cyst<sup>4</sup>.

Of these, sebaceous trichofolliculoma (ST) has the most similar features to FSCH and there is a controversy about the relationship between these two conditions<sup>2,3</sup>. ST is a rare variant of trichofolliculoma, usually occurring on the nose as numerous well-differentiated sebaceous lobules and ducts with occasional hair structures arranged around a central cystic "follicle"<sup>12</sup>. Schulz and Hartschuh<sup>2</sup> proposed that FSCH is the same disease as ST, which corresponds to a trichofolliculoma at its very late stage. They divided trichofolliculoma into 4 stages, that is, early, fully developed, late and very late stage, and explained that each stage reveals its characteristic findings through serial changes. Sebaceous differentiation was more pronounced than in fully developed trichofolliculoma, reaching from several foci of sebaceous cell nests to entire sebaceous lobules, linked to the dilated infundibular structure or located freely in the perifollicular sheath. Compared with fully developed trichofolliculoma, the stroma of the late stage of trichofolliculoma was also far more prominent, characterized by many dilated vessels, numerous fibrocytes, as well as fibrillary bundles of collagen and therefore resembled the stroma of FSCH. They described that the various presentations of FSCH are simply different stages of one and the same lesion. The other view to the relation of FSCH and trichofolliculoma was proposed by Simon *et al.*<sup>3</sup> They proposed that FSCH is but the sebaceous end of the tricho-sebo-folliculoma spectrum and that the name of FSCH should be deleted and substituted by sebofolliculoma, a pole of the spectrum of tricho-sebo-folliculoma. On the other hand, Templeton<sup>4</sup> suggested FSCH is a distinct hamartoma formed from epithelial and stromal elements, and the mesenchymal stroma is the most polymorphous component of FSCH. ST has histopathologically a few similar findings to FSCH such as dilated follicular structure with associated sebaceous elements and rudimentary follicles. However, there are far more different findings between them. For examples, ST locates more superficially in the dermis and has hair shafts within dilated follicular structures. It also lacks mesenchymal proliferative stroma of FSCH<sup>12</sup>. Clinically, whereas ST occurs around the nose and has a central depression or pores, FSCH occurs on any area in the face, on the scalp, and even on the back. Moreover, it does not always show pores or hairs, as in our case. Therefore, it has been suggested that

FSCH belongs in a spectrum of folliculosebaceous hamartomas that vary in aspects of the proportion of epithelial and mesenchymal components.

The treatment is a surgical excision. There has been no report of association with gastrointestinal malignancy or Muir-Torre syndrome.

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