A Case of Folliculosebaceous Cystic Hamartoma on the Upper Back

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Folliculosebaceous cystic hamartoma(FCH) is a recently recognized adnexal tumor and is composed of follicular, sebaceous and mesenchymal elements such as small vessels, adipocytes and neural component in the fibrous stroma. It usually presents as an exophytic papule or nodule on the central part of the face. We describe a 39-year-old woman with FCH on the right upper back, which is an unusual site. A biopsy specimen demonstrated a follicular structure in infundibular nature, sebaceous lobules attached to it, and compactly laminated fibrous stroma with an increased number of vessels in it. Immunohistochemically, many of the vessels in the stroma showed positive reaction for CD34, and Factor Ⅷ.

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Key Words : Folliculosebaceous cystic hamartoma, Upper back

Folliculosebaceous cystic hamartoma(FCH) is a term first used by Kimura et al[1] 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[1].

Although about 20 cases have been reported, FCH is probably not as uncommon as its rarity in the literature would indicate. In Korea, 4 cases of FCH have been reported[2-4].

Histogenesis of FCH is still unclear. Some authors denied that FCH is a disease entity[5-6], whereas others regarded it as a distinct adnexal tumor despite the controversial nosology[1,7-11]. The controversy exists largely in the relationship between FCH and sebaceous trichofolliculoma. The most characteristic feature of FCH is the change of the mesenchymal stroma showing increased small vessels, adipocytes and neural component.

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CASE REPORT

A 39-year-old woman presented with a 8-year history of a papule on the right upper back. Physical examination revealed a 0.8 cm, skin colored, firm and sessile papule, which had neither pores nor hairs(Fig. 1). Under the clinical impression of an "epidermal cyst", the lesion was removed surgically. Histopathologic examination revealed that the epidermis showed no abnormal findings and was not connected to the dermal lesion(Fig. 2). In the dermis, there were keratin-filled, dilated infundibular structures and numerous sebaceous lobules attached through sebaceous ducts to them as well as an isolated lobule in the stroma(Fig. 3). The sebaceous lobules had normal features of sebocytes with clear cytoplasm and central nucleus. The stroma demonstrated compactly laminated fibrosis and diffuse proliferation of small vessels. However there were no increase of adipocytes and/or neural tissues. The tumor was separated by cleft from the surrounding, compressed normal tissue in the dermis(Fig. 3). Immunohistochemically, the vessels

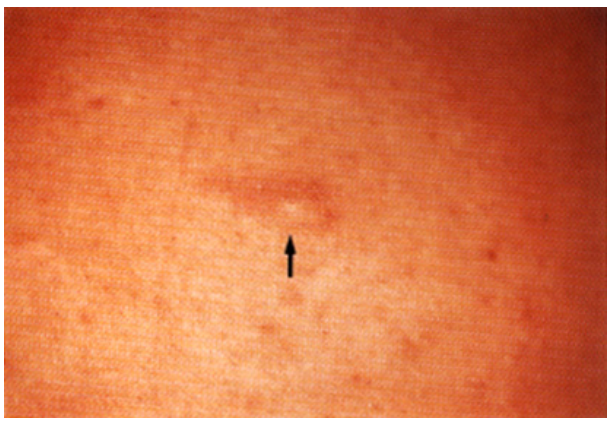


Fig. 1. A 0.8 cm, skin colored, sessile papule is seen on the right upper back(arrow).

in the stroma were positive for CD34(Fig. 4), and factor Ⅷ.

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

There was no evidence of recurrence in the following 14 months after removal.

DISCUSSION

Clinically, FCH is a solitary, smooth-surfaced, sessile or pedunculated papule or nodule on the face, scalp[7] and rarely on the back[8] like our case. The tumor size is usually less than 2 cm in diameter, but Yamamoto et al[8] reported a giant FCH reaching

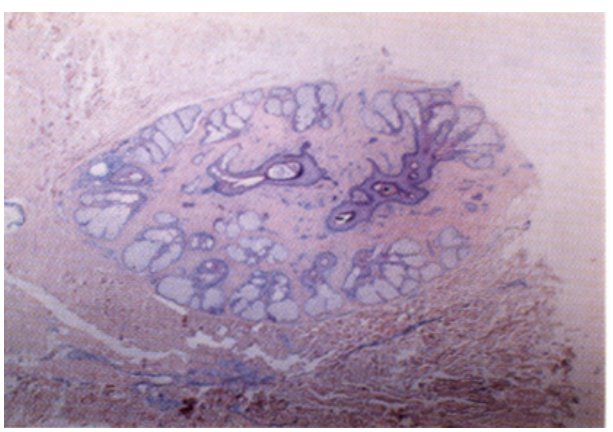


Fig. 2. This section is showing typical lesion, consisting of folliculosebaceous element and compactly laminated stroma separated by cleft from the surroundings(H&E, 10×).

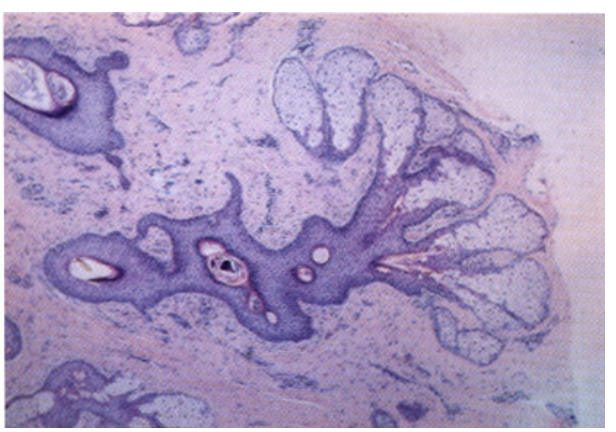
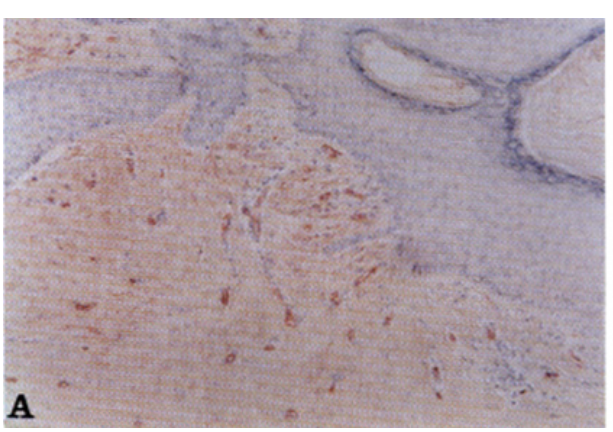


Fig. 3. Higher magnification of Fig. 2 shows numerous sebaceous lobules attached through sebaceous duct to the infundibular structure(H&E, 40×).



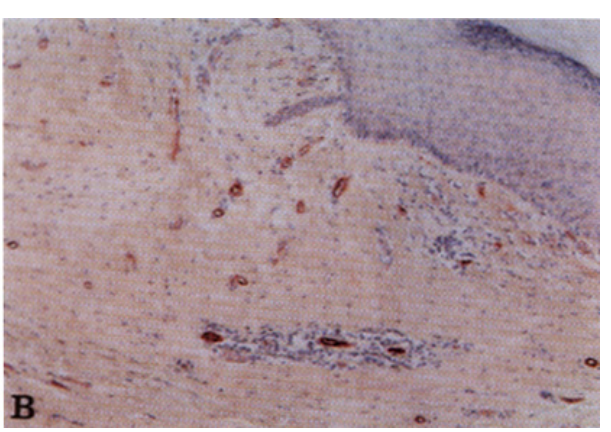


Fig. 4. Many small vessels are seen in the fibrous stroma(Immunohistochemical stain for CD34, 40×).

7.5 cm in diameter. FCH is typically asymptomatic and slow growing, and observed in adults older than 30 years of age[1-4]. Microscopically, the lesion shows a composite of follicular, sebaceous and mesenchymal elements. Kimura et al[1] proposed diagnostic histopathologic criteria of FCH as follows:

1. An infundibular cystic structures to which are attached sebaceous lobules via sebaceous ducts.
2. Compactly laminated fibroplasia around the entire epithelial component of fibroepithelial units.
3. Mesenchymal changes around fibroepithelial units that include fibrillary bundles of collagen, adipocytes, and an increased number of small venules.
4. Cleft between fibroepithelial units and surrounding altered stroma, and at the periphery between the altered stroma and adjacent compressed fibrous tissue.
5. Confinement of process primarily to the dermis in some instances, the subcutaneous fat may be involved.

In addition, several variants have been reported including FCH with perifollicular mucinosis[9], neural component[10] and apocrine gland[11].

Our case met the above criteria for FCH except adipocytes or neural tissues. But immunohistochemical staining using CD34 and Factor Ⅷ showed many small vessels in the stroma.

The histopathologic differential diagnoses include sebaceous hyperplasia, sebaceous trichofolliculoma, fibrofolliculoma, perifolliculoma and dermoid cyst[7]. Of these, sebaceous trichofolliculoma(ST) has the most similar features to FCH and there is a controversy about the relationship between these two conditions[5,6]. ST is a rare variant of tricho- folliculoma, usually occurring on the nose as numerous well-differentiated sebaceous lobules and ducts with occasional hair structures arranged around a central cystic "primary follicle"[12]. Schulz and Hartschuh[5] proposed that FCH 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 FCH. And they described that the various presentations of FCH are simply different stages of one and the same lesion. The other view to the relation of FCH and trichofolliculoma was proposed by Simon et al[6]. They proposed that FCH is but the sebaceous end of the tricho-sebo-folliculoma spectrum and that the name of FCH should be deleted and substituted by sebofolliculoma, a pole of the spectrum of tricho-sebo-folliculoma. On the other hand, Templeton[7] suggested FCH is a distinct hamartoma formed from epithelial and stromal element, and the latter is the most characteristic feature of FCH. ST has histopathologically a few similar findings to FCH 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, and, first of all, lacks mesenchymal proliferative stroma of FCH[12]. Clinically, whereas ST occurs around the nose and has central depression or pores, FCH 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 FCH belongs in a spectrum of folliculosebaceous hamartomas that vary based on the proportion of epithelial and mesenchymal components.

The clinical differential diagnoses include epidermal cyst, intradermal nevus, neurofibroma, pilomatricoma and other adnexal neoplasms.

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

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