A Case of Solitary Fibrofolliculoma

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Fibrofolliculoma is a benign follicular neoplasm which usually occurs in multiple and rarely solitarily. We have found only seven cases of previous reports of solitary fibrofolliculomas worldwide and only two in Korean literature.

Herein we report on a 40-year-old female patient with a solitary flesh-colored bean sized mass on the scalp which histopathologically proved to be a fibrofolliculoma.

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Key Words : Fibrofolliculoma, Solitary

Fibrofolliculoma is a recently described perifollicular connective tissue tumor which usually occurs in multiple with trichodiscomas and acrochordons, constituting the Birt-Hogg-Dubé syndrome. The solitary form is much rarer, nonhereditary, and unassociated with other abnormalities. Both have identical characteristic histopathological features showing infundibular epithelial proliferation and perifollicular fibrous proliferation[1-3].

We herein report a rare case of solitary fibrofolliculoma, exhibiting typical clinical and histological features in an otherwise healthy 40-year-old woman

CASE REPORT

A 40-year-old Korean woman noticed a bean-sized mass on her left scalp for about a year prior to attending our clinic. On examination of the skin, there was a 0.7× 0.6× 0.5cm sized skin-to-pink colored protruding mass slightly umbilicated in the center which was situated on the left parietal scalp area (Fig. 1). It had a smooth surface with a rubbery consistency. She had no other significant skin lesions and was in good general health without any previous medical problems. There was no family history of similar lesions.

An excision biopsy was performed and histological examination revealed a well defined tumor mass involving a group of adjacent pilosebaceous follicles (Fig. 2). Each individual lesion showed proliferative epithelial cords and spurs in the center, and a fibromucinous mesenchymal component surrounding it (Fig. 3). It also showed characteristic appearances of proliferating infundibular epithelial strands, 2-4 cells thick, anastomosing to form an epithelial network (Fig. 4). No signs of recurrence or newly appearing lesions have been found to date.

DISCUSSION

As each of the mesodermal components of the skin can be the source of neoplasms, the connective tissue component of the pilar apparatus gives rise to three different forms of tumors which include fibrofolliculoma, perifollicular fibroma, and trichodiscoma[4]. They are rare and generally occur in multiple in pure forms or in mixed combinations of these three tumors and share a common clinical presentation[3].

Among them, fibrofolliculoma was first described in 1977 by Birt, Hogg and Dubé[1]. They reported 15 members of a family with numerous small papular skin lesions which proved to be pilar hamartomas involving both epithelial and fibrous tissues associated with trichodiscomas and acrochordons. The name 'fibrofolliculoma' was introduced for

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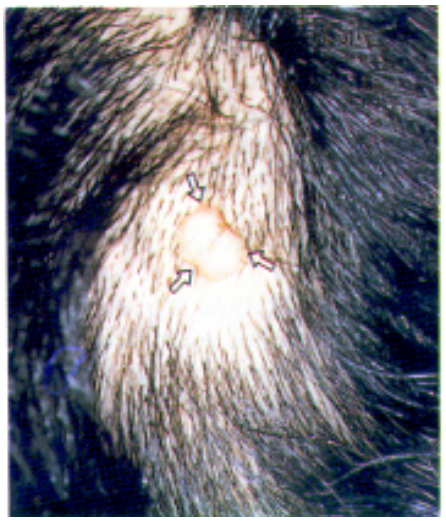

Fig. 1. Skin-colored protruding mass with a shallow central dell on the left parietal scalp area (open-spaced arrows).

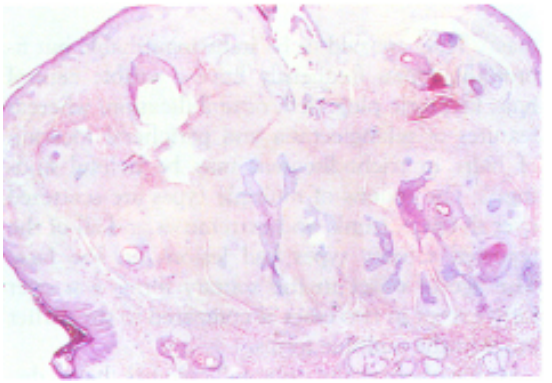

Fig. 2. Low power view showing circumscribed masses of involved adjacent pilosebaceous follicles (H&E, × 20).

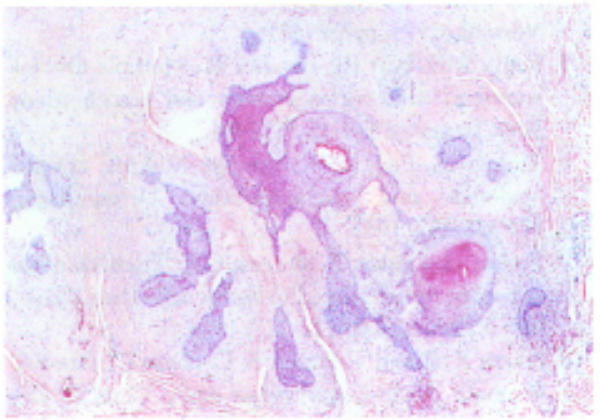

Fig. 3. Central dilated follicle with proliferative epithelial cords and spurs extending out into the surrounding fibromucinous mantle (H&E, ×40).

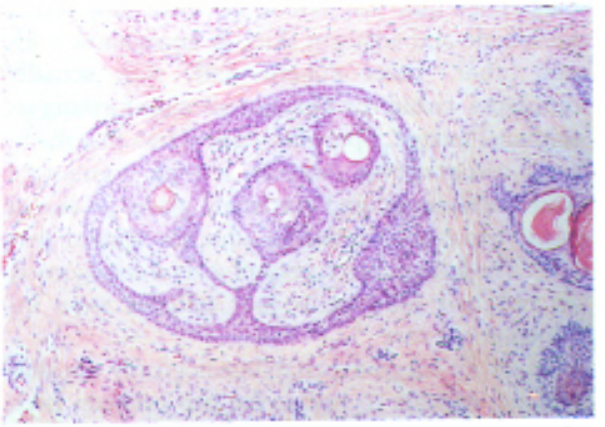

Fig. 4. Typical appearances of proliferating stroma and anastomosing epithelial strands (H&E, ×100).

the previously unrecognized tumor referred to in this report. Thereafter this syndrome has been known as the so-called Birt-Hogg-Dubé syndrome.

The multiple type usually appears after the age of twenty-five and shows autosomal dominant inheritance but sporadic cases have also been reported[5,6]. Most reports of multiple fibrofolliculomas have been associated in various combinations with trichodiscomas, acrochordons[1,2,5,6], perifollicular fibromas[7], and connective tissue nevus[2] and only two cases have been presented in the pure form unassociated with other significant cutaneous findings[3,8].

On the other hand, the solitary type of fibrofolliculoma is more uncommon and to our knowledge, only seven cases have been published previously worldwide and only two in Korean literature[3,9-12]. It has been shown to have the same clinical and histological features as in the multiple type and to be relatively trivial nonhereditary lesions[3].

Clinically fibrofolliculomas are asymptomatic 2-4mm yellowish white to skin-colored, smooth, dome-shaped lesions that look quite similar to trichodiscomas, perifollicular fibromas, or angiofibro-

mas[13]. They are said to be indistinguishable but fibrofolliculomas frequently have a visible central hair, keratotic plug, dell, or umbilication, whereas features of telangiectasia and peripheral location of hairs in trichodiscomas may be helpful findings[14]. The lesions of multiple types are scattered over the face, trunk, and extremities and all of the reported solitary types had lesions on the face. The age of onset in the solitary forms was over forty except in two cases as compared to the earlier onset in the multiple forms.

Histopathologically, fibrofolliculomas have distinctive and characteristic features with minor variations. The center of the lesion shows a hair follicle that is sometimes dilated and contains keratin material with a moderately well-circumscribed thick mantle of fibrous tissue surrounding it. The infundibular follicular epithelium extends out into this fibrous mantle forming epithelial strands, cords, or spurs. They may just extend outward, may rejoin the follicle and adjacent skin, or anastomose with each other. Three dimensionally they actually form septa within the fibromatous mass forming an 'epithelial sponge'. Special staining shows that the fibrous tissue stroma contains a high content of mucin substances, and extremely sparse or absent elastic tissue in contrast to adjacent normal dermis[1,2]. The lesions may consist of several contiguously affected hair follicles as four follicles were involved in our case[1,5,6].

Birt et al[1] suggested that histologically fibrofolliculoma appears to bridge the gap between perifollicular fibroma and trichofolliculoma based on the findings that fibrofolliculoma has epithelial proliferation with a perifollicular fibrous reaction whereas perifollicular fibroma has an essentially normal epithelial component and that fibrofolliculoma has a simpler and less prominent epithelial proliferation than that found in trichofolliculoma and there is also no formation of hair bulbs. Weintraub and Pinkus[2] suggested that the proliferation of both components in fibrofolliculoma is a manifestation of the close interaction between ectoderm and mesoderm in the follicle.

The present case showed typical histological features of fibrofolliculoma and occured solitarily on the scalp without any family history. Both previously reported cases of fibrofolliculoma in Korea were also the solitary type[11,12]. The fact that multiple types have only been reported in caucasians with light skin may be attributed to racial genetic differences, though the lack of medical attention of both patient and doctor should also be considered.

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