Congenital Fibrous Papule of the Face

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A 2-month-old Korean boy presented with a solitary papule on the cheek which was noted at birth. Histopathologic findings were consistent with angiofibroma of fibrous papule of the face (FPF). FPF is known to affect adults, and congenital occurrence has not been reported to the best of our knowledge.

We report a case of congenital FPF which showed a facial papule clinically and an angiofibroma histologically. (Ann Dermatol 8:(4)257~259, 1996).

Key Words : Angiofibroma, Congenital fibrous papule of the face

Fibrous papule of the face (FPF) is an uncommon condition, which appears as a small facial papule and shows histologic features of an angiofibroma. It is usually single and occurs chiefly on the nose of adults[1].

The angiofibroma associated with tuberous sclerosis develops in childhood and is characterized by multiple lesions in a symmetric centrofacial distribution[2].

This case is a congenital FPF which was a solitary facial papule since birth.

There is no previous report of congenital FPF on review of the literature.

REPORT OF A CASE

A 2-month-old boy had a solitary papule on the right cheek. The dome-shaped, 3 mm sized, reddish papule was noted at birth (Fig. 1). His growth and development was in normal range and there was no contributable family history.

Shave biopsy was performed. The epidermis showed focal hyperplasia and slightly increased numbers of melanocytes in the basal layer. Many activated fibroblasts such as plump, stellate, kite, and spindle cells were found in the dermis. Capillary proliferation and dilatation, fibrosis, focal adnexal and perivascular lymphocytic infiltration were also observed (Fig. 2). Some mast cells were seen around the dermal vessels in toluidine blue stained section.

We made the diagnosis of congenital FPF with all these clinical and histopathologic findings. There were no signs of mental retardation, seizure, and developmental abnormalities during the 6 month follow-up period.

DISCUSSION

Fibrous papule of the face refers to a small facial papule with a distinctive fibrovascular component on histological examination[1]. This lesion was first described as "perifollicular fibroma" by Zackheim in 1960[3], and was termed "fibrous papule of the nose" by Graham et al. in 1965[4]. The designation of FPF first named by Meigel and Ackerman in 1979 is now widely accepted and frequently used[1].

The histogenesis of FPF has been controversial. Graham et al. proposed its origin from a cellular nevus[4], and Saylan et al. also regarded it as a variant of nevus cell nevi on the ground of the binucleated kite-shaped cells as seen in Spitz nevus, and increased number of melanocytes in the basal layer[5].

Received November 13, 1995.
Accepted for publication July 4, 1996.
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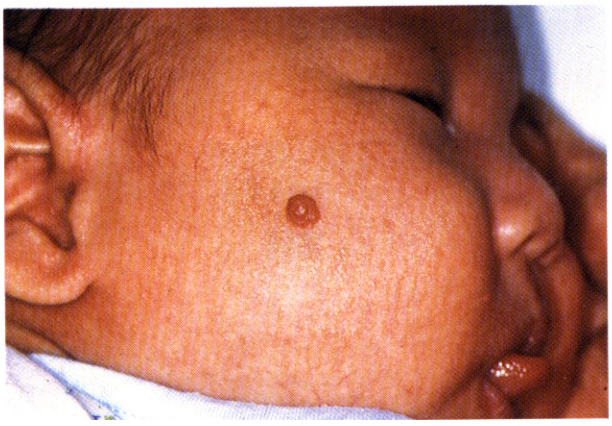

Fig. 1. A dome-shaped, reddish papule on the right cheek.

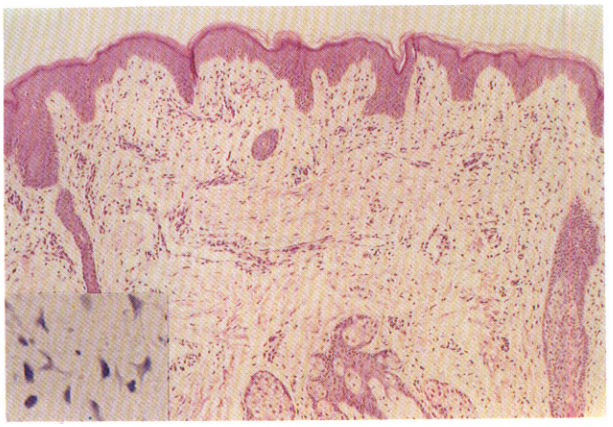

Fig. 2. Skin biopsy shows small increased numbers of melanocytes in the basal layer of the epidermis, and many activated fibroblasts and capillary proliferation and dilatation in the dermis. (Hematoxylin & eosin; magnification 100×.) Inset: High power view of the activated fibroblasts in the dermis. (Hematoxylin & eosin; magnification 400×).

However, Meigel and Ackerman in 1979 introduced an alternate histogenesis that the plump, stellate, and multinucleated cells in the dermis were fibroblasts[1]. This opinion was supported by the findings of many studies: the lack of S-100 protein as an immunohistochemical marker of neuroepithelial elements[6], no detectable melanosomes, premelanosomes, and desmosomes by electron microscope[7,8], and the intense immunostain reaction against the factor XIIIa antibody, staining in reactive and neoplastic fibroblastic and fibrohistiocytic lesions[9].

FPF is an angiofibroma histologically, which shows a localized area of fibrosis and vascular proliferation in the upper portion of the dermis, and scattered large triangular and stellate cells[1,10]. This angiofibromatous lesion of FPF is very similar to those that show in acral fibrokeratomas, pearly penile papules and oral fibromas. These diseases differ from FPF by the absence of perifollicular fibrosis because they contain no hair follicles, and the developing site[1]. The histologic appearance of adenoma sebaceum in tuberous sclerosis is similar to that of FPF, and may be differentiated only by clinical data[11].

FPF clinically has no characteristic findings, it should be differentiated from appendage, fibrous, or vascular tumors. In this case, we first thought FPF, trichofolliculoma or sebaceous hyperplasia. FPF can be differentiated from trichofolliculoma by the absence of a central core and a wool-like tuft resembling immature hair[12], and from sebaceous hyperplasia by the lesion number, size, and clinical course.

Our patient had a congenital facial papule, which was confirmed as an angiofibroma on histologic examination. To our knowledge, there is no reported case of congenital FPF.

REFERENCES

1. Meigel WN, Ackerman AB : Fibrous papule of the face. Am J Dermatopathol 1: 329-340, 1979.
2. Fitzpatrick TB, Szabo G, Hori Y, Simone AA, Reed WB, Greenberg MH : White leaf-shaped macules. Arch Dermatol 98: 1-6, 1968.
3. Zackheim HS, Pinkus H : Perifollicular fibromas. Arch Dermatol 91: 913-917, 1960.
4. Graham JH, Sanders JB, Johnson WC, Elson BH : Fibrous papule of the nose. J Invest Dermatol 45: 194-203, 1965.
5. Saylan T, Marks R, Wilson JE : Fibrous papule of the nose. Br J Dermatol 85: 111-118, 1971.
6. Spiegel J, Nadji M, Penneys NS : Fibrous papule: An immunohistochemical study with an antibody to S-100 protein. J Am Acad Dermatol 9: 360-362, 1963.
7. Santa Cruz DJ, Prioleau PG : Fibrous papule of the face: An electron microscopic study of the two cases. Am J Dermatopathol 1: 349-352, 1979.

8. Ragaz A, Berezowsky V : Fibrous papule of the face: A study of five cases by electron microscopy. Am J Dermatopathol 1: 353-355, 1979.
9. Albert JN, Neal SP, Helene BB : Fibrous papule: A tumor of fibrohistiocytic cells that contain factor XIIIa. J Am Acad Dermatol 19: 1102-1106, 1988.
10. McGibbon DH, Jones EW : Fibrous papule of the face (nose): Fibrosing nevocytic nevus. Am J Dermatopathol 1: 345-348, 1979.
11. Nickel WR, Reed WB : Tuberous sclerosis: Special reference to the microscopic alterations in the cutaneous hamartomas. Arch Dermatol 85: 209-226, 1962.
12. Pinkus H, Sutton RL Jr : Trichofolliculoma. Arch Dermatol 91: 46-49, 1965.