A Case of Fibrous Hamartoma of Infancy

Seung-Hyun Lee, M.D., Haeng-Seok Kim, M.D., Tae-Jin Yoon, M.D.,
Tae-Heung Kim, M.D., Kye-Yong Song, M.D.*

Department of Dermatology, College of Medicine, Gyeongsang National University, Chinju, Korea
Department of Pathology, College of Medicine, Chung-Ang University, Seoul, Korea*

Fibrous hamartoma of infancy (FHI) is an uncommon, benign, solitary intradermal or subcutaneous tumor. It occurs typically in the axillary or shoulder region. The histopathologic examination of affected lesion shows the characteristic elements: dense fibrous tissue, adipose tissue, and primitive mesenchymal tissues. A 15 month-old girl had the multiple, asymptomatic, discrete, and skin-colored tumors that scattered on the scalp. These were present at birth. We report a rare case of FHI occurred on the scalp with multiple and congenital characteristics. (Ann Dermatol 11(4) 283~285, 1999).

Key Words : Congenital, Fibrous hamartoma of infancy, Multiple, Scalp

Fibrous hamartoma of infancy (FHI) is a histologically distinctive fibrous proliferative disorder, which occurs solitarily on the axilla, upper arm and shoulder of an infant in the first two years of life.

The tumor is located in the subcutaneous tissue or reticular dermis. There is usually poor demarcation from the adjacent soft tissues[1,2]. The lesion is composed of three characteristic histopathologic components: 1) dense fibrocollagenous areas, often appearing as bands or septae coursing in different directions; 2) mature adipose tissue; and 3) foci of collagen-poor, moderately or densely cellular myxoid mesenchymal tissue[1].

The tumor is usually benign, and local excision is generally curative although occasionally the lesion recurs locally.

We report herein a rare case of FHI. Unlike typical form, it had multiple tumors and occurred congenitally on the scalp. The tumor was excised partially and has been followed with close observation.

Received January 15, 1999.
Accepted for publication July 27, 1999.
Reprint request to : Tae-Jin Yoon*, M.D., Assistant Professor. Department of Dermatology, Gyeongsang National University Hospital, 90 Chilam-Dong, Chinju, Gyeongnam, 660-702, Korea
Tel : 82-591-750-8187, FAX : 82-591-762-8179
E-mail: s_pibu@gshp.gsnu.ac.kr

CASE REPORT

A healthy 15 month-old girl with normal full-term delivery visited our department with her left temporal lesion. The clinical feature of skin lesion was the seven or eight, 1.5 to 3 cm sized, hairless, skin-colored, flat-topped, non-tender and firm tumors on the left temporal scalp (Fig. 1). These were noted at birth by her mother. The tumors were asymptomatic and increased slightly in size by growing up. Physical examination was otherwise normal. The past and family history were non-contributory.

Laboratory studies including complete blood cell count, blood chemistry study, urine analysis, chest X-ray and EKG were within normal limits or negative.

The specimen taken from the tumor was stained with hematoxylin and eosin, alcian blue of pH 2.5, and toluidine blue. There were non-specific findings in the epidermis. The tumors were located in the reticular dermis and subcutaneous tissue and were not encapsulated. These were composed of rather cell-poor fibrous trabeculae, whorls of immature appearing spindle cells in a mucoid matrix and mature adipose tissue (Fig. 2). And the fibrous trabeculae were composed of well-oriented, spindle-

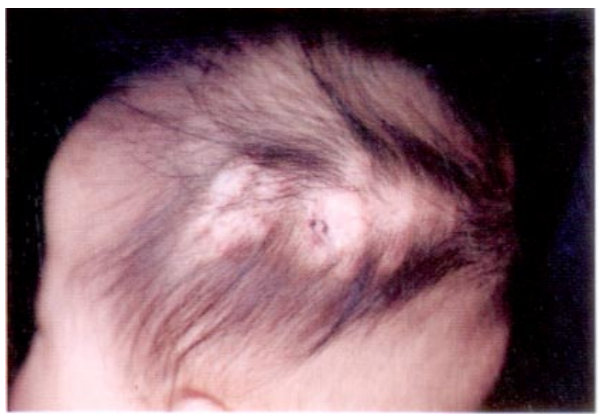

Fig. 1. The lesions on the scalp showing the erythematous to skin-colored, flat-topped and round-shaped nodules or tumors.

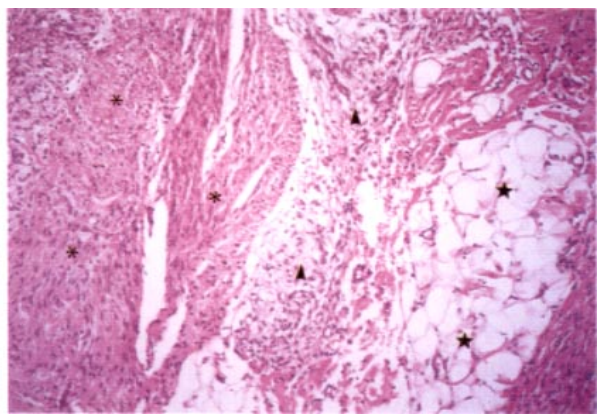

Fig. 2. Biopsy specimen taken from skin-colored tumor shows fibrous trabeculae(asterisk), myxoid mesenchymal tissue(arrowhead), and mature adipose tissue(star). (H&E, ×100).

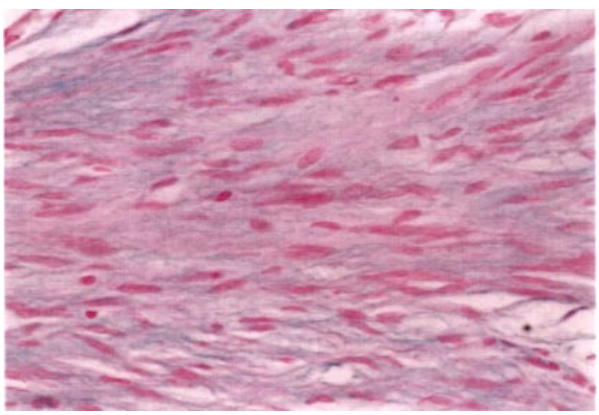

Fig. 3. alcian blue stained the cytoplasm of the spindle-shaped cells. It means that the stained materials are acid mucopolysaccharide. (alcian blue pH 2.5, ×400).

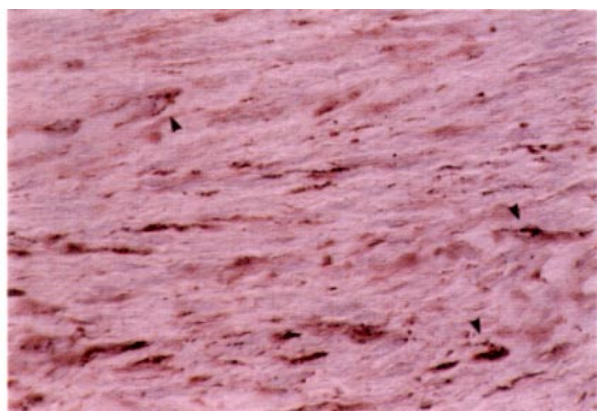

Fig. 4. The immunohistochemical staining with vimentin shows the positive reaction in the perinuclear cytoplasm(arrowhead). (Vimentin, ×400).

shaped cells with narrow or wavy nuclei arranged in loosely structured myxoid areas. Mature adipocytes were arranged in groups, and cellular areas consisting of immature oval or stellate mesenchymal cells were distributed within myxoid matrix (Fig. 2). The cytoplasms of the spindle-shaped cells were stained positively with alcian blue of pH 2.5 (Fig. 3) and toluidine blue (figure not shown).

The immunohistochemical staining with vimentin shows the positive reaction in the perinuclear cytoplasm of the spindle-shaped cells in the fibrous trabeculae and less weak reaction in the cellular myxoid areas (Fig. 4). The vimentin-positive cells are distributed diffusely on the dermal and subcutaneous area. But S-100 protein antibody, NSE, desmin and EMA staining were negative. Fibrous hamartoma of infancy was diagnosed by the clinical and characteristic histopathological findings.

Partial excision was carried out and the patient was followed up closely for 1 year. No new lesions occurred.

DISCUSSION

Fibrous hamartoma of infancy (FHI) usually develops during the first two years of life as a small rapidly-growing mass in the lower dermis or subcutis. In 15-20% of the cases, it presents at birth. It is usually solitary in nature, and only very rare cases of multiple lesions were reported[3]. The principal location is the axilla, followed by the upper arm, thigh, inguinal region, shoulder, back, and fore-

arm[4]. But it rarely occurred in the hand[5] and foot[6]. In our case, it had occured congenitally as multiple nodules or tumors on the scalp.

Clinically, FHI should be differentiated from enlarged lymph node, juvenile fibromatosis, lipoma, sarcoma, dermatofibroma, neurofibroma, or hemangioma[3]. But, it can be clearly diagnosed by characteristic histopathological and immunohistochemical findings.

The microscopic observation showed three characteristic findings as 1) fibrous trabeculae of various thickness, which intersect fat lobules in the subcutis, 2) myxoid mesenchymal tissues, and 3) mature adipose tissues. The fibrous trabeculae were composed of spindle-shaped cells, fibroblasts, arranged in loosely structured myxoid areas and more densely collagenous foci. The stellate cells showed immature mesenchymal cells, and the spindle-shaped cells showed fibroblasts. There was no anaplasia, and mitotic figures are infrequent. By some literature, the fibrocollagenous regions of the tumor contained high amounts of myofibroblastic cells positively stained for desmin and actin. In our case, it was negative for desmin. Myofibroblasts are modified fibroblasts that are capable of assuming structural and functional properties of smooth muscle cells. These cells have been noted in many normal as well as pathological tissues[8]. But other literature showed different immunohistochemical results[9,10].

The precise histogenesis of FHI is unknown, although the morphology seems to fulfill the criteria for a hamartomatous tumor, which is defined as an excessive growth of tissue that is normal to the area in which it is found.

Since FHI is not seen after the age of three, one has to assume that its peak growth occurs in early childhood and thereafter becomes quiescent and regresses spontaneously[4]. But in fact, one study revealed that it did not show spontaneous regression[2]. The findings of this study showed that the tumor grew rapidly from birth up to the age of about 5 years; it then slowed down but growth did not stop, nor was there regression. There were no other changes from the microscopic pattern except for a better formed capsule. Malignant changes were not observed[2]. Although rapid infiltrative growth and local recurrence were observed, long-term clinical follow-up indicated an essentially benign clinical course[6]. Delayed surgery was not associated with increased risk of operative complications: in fact, it seemed to facilitate complete resection of the tumor[2]. The presence of superficial infiltration into underlying muscle, rapid recurrence, and multiple lesions did not necessarily imply a bad prognosis[3]. There was no clear description of whether there was any relation between multiplicity and recurrence. When multiple nodules occur, close follow-up is recommended to find newly-developing nodules in the vicinity[4]. In other literature, the tumor was said not to recur or metastasize[11]. It is important to recognize this tumor properly and differentiate from malignant tumors not to overtreat it.

REFERENCES

1. Paller AS, Gonzalez-Crussi F, Sherman JO : Fibrous hamartoma of infancy. Arch Dermatol 125:88-91, 1989.
2. Efem SE : Clinicopathological features of untreated fibrous hamartoma of infancy. J Clin Pathol 46:522-524, 1993.
3. Maung R, Lindsay R, Trevenen C et al : Fibrous hamartoma of infancy. Hum Pathol 18:652-653, 1987.
4. Jung PM, Hong EK : fibrous hamartoma of infancy manifested as multiple nodules. J Korean Med Sci 5:243-247, 1990.
5. Jebson PJ, Louis DS, Arbor A : Fibrous hamartoma of infancy in the hand: A case report. J Hand Surg 22A:740-742, 1997.
6. German DS, Paletta CE, Gabriel K : Fibrous hamartoma of infancy. Orthopedics 19:258-260, 1996.
7. Groisman G, Kerner H : A case of fibrous hamartoma of infancy in the scrotum including immunohistochemical findings. J Urol 144:340-341, 1990.
8. Michal M, Mukensnabl P, Chiumska A et al : Fibrous hamartoma of infancy A study of eight cases with immunohistochemical and electron microscopical findings. Path Res Pract 188:1049-1053, 1992.
9. Popek EJ, Montgomery EA, Fourcroy JL : Fibrous hamartoma of infancy in the genital region. J Urol 152:990-993, 1994.
10. Groisman G, Lichtig C : Fibrous hamartoma of infancy: An immunohistochemical and ultrastructural study. Hum Pathol 22:914-918, 1991.
11. Abara EO, Churchill BM, McClorie GA et al : Fibrous hamartoma of infancy. J Urol 140:1508-1509, 1988.