J Korean Surg Soc 2011;81:61-65
DOI: 10.4174/jkss.2011.81.1.61

JKSS
Journal of the Korean Surgical Society

pISSN 2233-7903 · eISSN 2093-0488

ORIGINAL ARTICLE

Fibrous hamartoma of infancy: an experience of a single institute

Guhyun Kang, Yeon-Lim Suh, Joungho Han, Gee-Young Kwon, Suk-Koo Lee[1], Jeong Meen Seo[1]

Departments of Pathology and [1]Surgery, Samsung Medical Center, Sungkyunkwan University School of Medicine, Seoul, Korea

Purpose: Fibrous hamartoma (FH) of infancy is a distinctive fibrous growth that most frequently occurs at birth and during the postnatal period. It is important for clinicians and pathologists to recognize this entity to avoid an aggressive approach. **Methods:** We herein describe the clinicopathologic features of 9 FHs diagnosed at a single institution between 1997 and 2010. **Results:** There were 7 boys and 2 girls, and the mean age of presentation was 14.7 months. The common locations were the lower back and gluteal region (n = 3) and scrotum (n = 2). They were solitary lesions, and measured 1.0 to 7.0 cm in maximum diameter (mean, 4.9 cm). The excised masses tended to be poorly circumscribed, and consisted of an intimate mixture of firm, gray-white tissue with fat. Histologically, these lesions were composed of 3 components forming a vague, irregular, organoid pattern: well-defined intersecting trabeculae of fibrocollagenous tissue; loosely textured areas of small, rounded, primitive mesenchymal cells; and mature fat. Over a median follow-up of 72 months, no patient showed recurrence. **Conclusion:** FH should be distinguished from other forms of fibromatosis and malignant tumors because it is benign and usually cured by local excision.

Key Words: Hamartoma, Infant, Soft tissue neoplasms, Differential diagnosis, Fibromatosis

INTRODUCTION

Subcutaneous soft tissue masses in infants and children encompass a diverse clinical and pathologic spectrum of entities including hamartomatous or choristomatous lesion, malformation, benign, intermediate, and malignant neoplasms. The majority are benign, but the rarity and diversity of infantile soft tissue tumors present a diagnostic and therapeutic challenge. The most common category is vascular, followed by fibroblastic-myofibroblastic, fibrohistiocytic, neurogenic and adipose [1,2]. Fibrous hamartoma (FH) of infancy is an uncommon fibroproliferative lesion that develops during the first 2 years of life, and up to 25% are discovered at birth [3,4]. It presents as a solitary mass-like lesion in the subcutaneous layer with rapid growth, and is occasionally attached to the underlying fascia [4,5]. The clinical and radiologic features of this lesion are nonspecific, and can mimic those of malignant soft tissue tumors [4,6,7]. Diagnosis is made by histologic examination to identify the characteristic triphasic pattern consisting of fibrocollagenous tissue, primitive mesenchymal cells and mature fat [3]. FH is a benign le-

Received January 31, 2011, Accepted March 23, 2011

Correspondence to: Joungho Han
Department of Pathology, Samsung Medical Center, Sungkyunkwan University School of Medicine, 50 Irwon-dong, Gangnam-gu, Seoul 135-710, Korea
Tel: +82-2-3410-2800, Fax: +82-2-3410-0025, E-mail: hanjho@skku.edu

sion that is cured by local excision [4,5]. In these points of view, it is important to recognize characteristics of this entity for proper diagnosis and clinical management. For this purpose, we present the clinicopathologic features and treatment results of 9 patients with FH diagnosed at a single institution.

METHODS

The cases in this study were 9 patients with FH, who had undergone local excision at our institution between 1997 and 2010. As this lesion had been classified under a variety of terms, it is likely that the patients presented here are a part of such cases in the pathology files of our department. The clinicopathologic information was obtained by medical record review. To protect patients' confidentiality, the database was password protected. After the data were exported to a spreadsheet, all specific identifiers (e.g., name, medical record number) were removed. The hematoxylin and eosin-stained slides were available in all cases, and the histologic features, including tumor circumscription, mitotic activity, celluarity, cytologicatypia and presence of necrosis or calcification, were assessed by 2 pathologists (J. Han and G. Kang). Complementary immunohistochemistry was not performed.

RESULTS

The clinical features and treatment results of the 9 patients were summarized in Table 1. There were 7 boys and 2 girls, and the mean age of presentation was 14.7 months (median, 5 months). Family history did not reveal any pertinent findings. The lesion presented as a rapidly growing mass in two patients (case no. 1 and 5), and appeared as a subcutaneous swelling with ill-defined mass in one patient (case no. 6). They were all solitary, usually freely movable in the subcutaneous layer. The lesions were most commonly located in the lower back and gluteal region (n = 3), followed by the scrotum (n = 2), posterior neck, upper arm, axilla and groin. Ultrasonography showed a subcutaneous mass with ill-defined margin and heterogeneous echogenicity in 3 patients, which was suggestive of hemangioma, lymphangioma or fibrolipoma. Magnetic resonance imaging examination of case no. 5 revealed a heterogeneous low-signal mass arising from the spermatic cord on T2-weighted image.

There being no tendency to regress, all patients underwent complete excision. Fig. 1 shows the macroscopic and microscopic appearance of the FHs in our series. The le-

Table 1. Clinical features of 9 patients with fibrous hamartoma of infancy

Case No.	Gender	Age of presentation	Site of involvement	Clinical presentation	Ultrasonographic findings	Size (cm)	Follow-up period (mo)
1	Male	4 mo	Upper arm	Enlarging movable, firm mass	-	4.5	128
2	Female	1 mo	Upper buttock	Palpable soft mass	Isoechoic mass in the subcutaneous fat layer	7.0	72
3	Male	9 mo	Posterior neck	Movable, firm mass	Heterogenous echogenic mass	3.6	22
4	Female	10 mo	Axilla	Movable, firm mass	Heterogenous echoic mass with fuzzy margin	4.0	22
5	Male	8 yr	Scrotum	Rapidly growing firm mass	-	4.0	20
6	Male	5 mo	Lower back	Subcutaneous swelling	-	7.0	117
7	Male	At birth	Lower back	Movable, firm mass	Heterogenous high-echogenic mass with internal vascularity	2.3	4
8	Male	6 mo	Scrotum	Movable, soft mass	-	2.0	154
9	Male	At birth	Groin	Fixed, firm mass	-	1.0	141

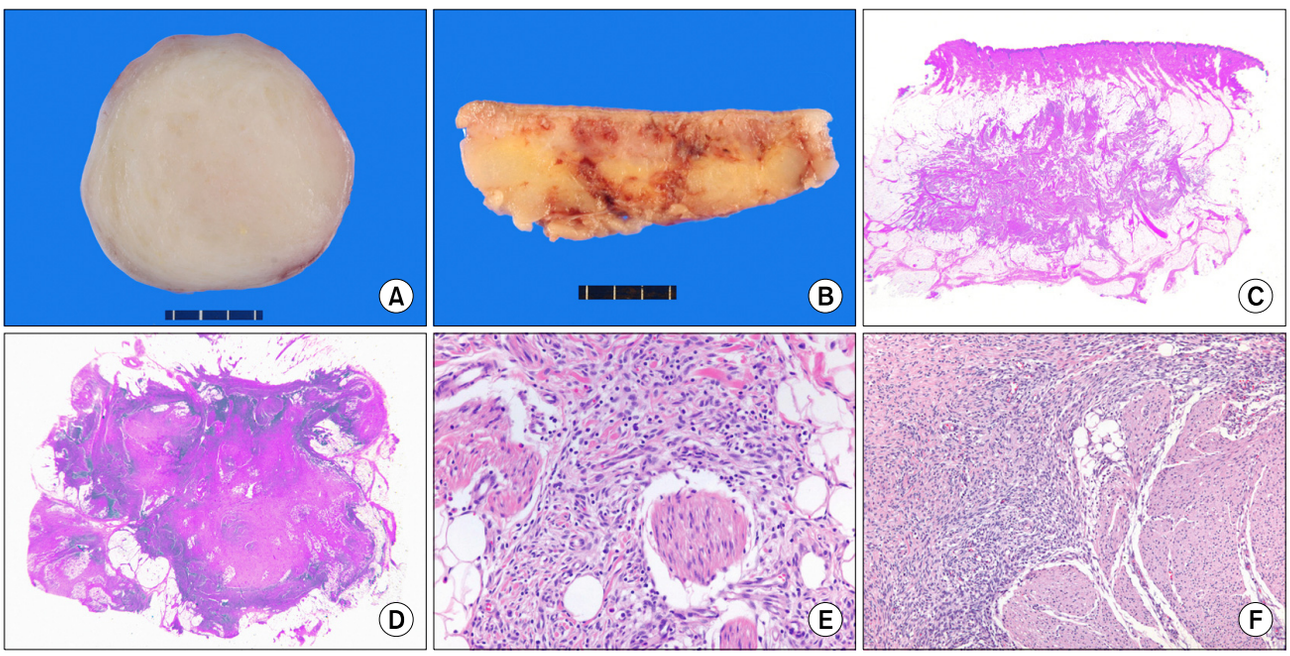

Fig. 1. (A) The mass is relatively well-defined, and exhibits rubbery, gray-white fibrous tissue (case no. 5). (B) The lesion is poorly circumscribed, and has a soft, gray-white surface with interposing yellowish fat (case no. 6). (C, D) At scanning power (H&E), there are poorly circumscribed fibrotic (or sclerotic) lesions in the dermis (case no. 3 and 4, respectively). (E, F) The lesions are characterized by an organoid mixture of 3 distinct components: loosely textured area of small, round or spindle, primitive mesenchymal cells, fibrocollagenous tissue and mature fat at ×100 and ×40 magnification (H&E), respectively (case no. 3).

sions were poorly defined from the surrounding tissue except for the cases in the scrotum and groin, and the cut surfaces were rubbery or soft and gray to white with interspersed yellowish fat. The amount of the fatty component varied among areas, and it occupied a large proportion in some cases, thereby resembling a fibrolipoma. The involved area measured 1.0 to 7.0 cm in maximum diameter (mean, 4.9 cm; median, 4.0 cm). All cases demonstrated essentially similar histologic features, and were composed of 3 different tissues forming an organoid structure. The well-defined trabeculae of the dense fibrocollagenous tissue consisted of fibroblastic spindle cells with bland, straight or wavy nuclei. The loosely textured islands were disposed among the fibrous trabeculae, which comprised immature-appearing, small, round or stellate, primitive mesenchymal cells with scanty cytoplasm. The mature adipose tissue was interspersed between the two components. The relative proportion of these 3 components varied considerably from area to area, and fat was recognized only at the periphery in some cases. Mitotic figures were absent in the fibroblastic and primitive cell areas, and nuclear atypia or hyperchromasia was not observed in any of the sections. Surgical radicality was achieved in all patients, but microscopic residuals were found in 3 cases. All remained well without local recurrence at 4 to 154 months (median, 72 months) including the patients with residual disease.

DISCUSSION

To our knowledge, this is the first comprehensive study in Korea describing the clinicopathologic features and follow-up results of patients diagnosed with FH at a single institution over 13 years. This lesion was first reported by Reye [8] in 1956 as subdermal fibromatous tumor of infancy, and the current term was later coined by Enzinger [3] in 1965. FHs are histologically characterized by an organoid mixture of 3 components: well-defined intersecting trabeculae of fibrocollagenous tissue; loosely textured areas of immature-appearing, small, rounded, primitive mesenchymal cells; and mature fat. In some cases, a pro-

nounced sclerosing process replaces the majority of the lesion [3].

FHs can be found in a variety of anatomical locations, although the principal sites are the axilla, shoulder, upper arm, back and groin. Cases involving the scalp, scrotum, perianal area and lower extremities have been also described [3,9-13]. The most common site in this study was the lower back and gluteal region. Like other fibrous tumors in children, this lesion is more common in boys, but there is no evidence of familial tendency [3,9]. FH has been reported in an infant and a 4-year-old boy with desmoids-type fibromatosis and tuberous sclerosis (TS), respectively [5,14]. The genetic tendency for TS to give rise to hamartomas might explain the coexistence with FH, but more detailed studies are needed to prove a genetic link between FH and TS [14]. A reciprocal translocation [t(2;3)(q31;q21)] was present in a previous report, and it is worthy of further study to determine whether chromosomal aberrations are related to its pathogenesis [15]. The lesions were solitary in our series, but several cases of multiple nodules in the same patient have been described [3,16,17]. Most FHs were less than 5 cm in diameter as described in the literature, and can reach up to larger than 10 cm [7,18]. They are usually firm, and may be affixed to the underlying tissue, thus causing the concern for malignancy [7].

Despite its infiltrative growth and focal cellularity, the clinical course is benign [5,7,18]. The treatment of choice for FH is complete excision with an envelope of normal tissue [3,7]. Although as many as 16% locally recur within a few months after primary excision, recurrences are nondestructive and cured by re-excision [3-5]. It was reported that the tumor capsule was developed in protracted and untreated FH. It continued to grow without regression, but delayed excision was not associated with an increased risk of postoperative complications [18]. Clinical differential diagnosis includes a number of entities such as epidermal cyst, inclusion body fibromatosis, juvenile hyaline fibromatosis, palmoplantar fibromatosis, histiocytoma, dermatofibroma, leiomyosarcoma and fibrosarcoma [12]. When the myofibroblastic areas predominate, the lesion may be difficult to distinguish from lipofibromatosis, infantile myofibromatosis and calcifying aponeurotic fibroma. However, the typical triphasic feature is readily recognized in most cases with sufficient sampling of the lesion. The overlying skin was grossly unremarkable, and was not sampled for histologic evaluation in such cases. However, eccrine changes, increased terminal hair follicles and epidermal basaloid follicular hyperplasia were frequently seen in previous reports, and these can be useful clues to consider a diagnosis of FH when the biopsy is superficial [19,20].

In conclusion, both the clinician and pathologist should be aware of this entity in order to avoid misdiagnosis of other fibromatosis and malignant tumor and unnecessary aggressive treatment.

CONFLICTS OF INTEREST

No potential conflict of interest relevant to this article was reported.

REFERENCES

1. Coffin CM, Dehner LP. Soft tissue tumors in first year of life: a report of 190 cases. Pediatr Pathol 1990;10:509-26.
2. Gurney JG, Ross JA, Wall DA, Bleyer WA, Severson RK, Robison LL. Infant cancer in the U.S.: histology-specific incidence and trends, 1973 to 1992. J Pediatr Hematol Oncol 1997;19:428-32.
3. Enzinger FM. Fibrous hamartoma of infancy. Cancer 1965;18:241-8.
4. Dickey GE, Sotelo-Avila C. Fibrous hamartoma of infancy: current review. Pediatr Dev Pathol 1999;2:236-43.
5. Carretto E, Dall'Igna P, Alaggio R, Siracusa F, Granata C, Ferrari A, et al. Fibrous hamartoma of infancy: an Italian multi-institutional experience. J Am Acad Dermatol 2006;54:800-3.
6. Maung R, Lindsay R, Trevenen C, Hwang WS. Fibrous hamartoma of infancy. Hum Pathol 1987;18:652-3.
7. Lee JT, Girvan DP, Armstrong RF. Fibrous hamartoma of infancy. J Pediatr Surg 1988;23:759-61.
8. Reye RD. A consideration of certain subdermal fibromatous tumours of infancy. J Pathol Bacteriol 1956;72:149-54.
9. Sotelo-Avila C, Bale PM. Subdermal fibrous hamartoma of infancy: pathology of 40 cases and differential diagnosis. Pediatr Pathol 1994;14:39-52.
10. Mitchell ML, di Sant'Agnese PA, Gerber JE. Fibrous hamartoma of infancy. Hum Pathol 1982;13:586-8.
11. Harris CJ, Das S, Vogt PJ. Fibrous hamartoma of infancy in

the scrotum. J Urol 1982;127:781-2.
12. Albukerk J, Wexler H, Dana M, Silverman J. A case of fibrous hamartoma of infancy. J Pediatr Surg 1979;14:80-2.
13. Tay YK, Levy ML. Congenital plantar nodule in an infant. Pediatr Dermatol 2004;21:506-7.
14. Han HJ, Lim GY, You CY. A large infiltrating fibrous hamartoma of infancy in the abdominal wall with rare associated tuberous sclerosis. Pediatr Radiol 2009;39:743-6.
15. Lakshminarayanan R, Konia T, Welborn J. Fibrous hamartoma of infancy: a case report with associated cytogenetic findings. Arch Pathol Lab Med 2005;129:520-2.
16. Jung PM, Hong EK. Fibrous hamartoma of infancy manifested as multiple nodules: a case report. J Korean Med Sci 1990;5:243-7.
17. Yoon TY, Kim JW. Fibrous hamartoma of infancy manifesting as multiple nodules with hypertrichosis. J Dermatol 2006;33:427-9.
18. Efem SE, Ekpo MD. Clinicopathological features of untreated fibrous hamartoma of infancy. J Clin Pathol 1993; 46:522-4.
19. Grynspan D, Meir K, Senger C, Ball NJ. Cutaneous changes in fibrous hamartoma of infancy. J Cutan Pathol 2007; 34:39-43.
20. Fernandez M, Al-Hammadi A, Nahal A. Eccrine gland stroma changes in cutaneous hamartoma of infancy. J Cutan Pathol 2008;35:508-9.