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Fibrous Hamartoma of Infancy Manifested as Multiple Nodules

— A Case Report —

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Fibrous hamartoma of infancy is an uncommon benign fibrous proliferation, usually presenting as a solitary nodule.

A rare example of multiple fibrous hamartoma of infancy is described. Two masses which developed in the upper arm of a 10-month-old boy were successfully excised, and one month later three small nodules 2 cm below the previous operation field appeared.

It will be further elucidated whether multiple forms and early recurrence are related or not as cases accumulate.

Key Words: *Fibrous hamartoma of infancy, Multiplicity, Recurrence*

INTRODUCTION

Fibrous hamartoma of infancy is a relatively rare subcutaneous tumor showing unique clinical and pathological features. It was first described by Reye in 1956 (Reye, 1956) and later defined as an entity by Enzinger in 1965 (Enzinger, 1965). Enzinger (Enzinger, 1965) reviewed 30 cases and termed the lesion as "fibrous hamartoma of infancy", hereby, clearly characterizing the unique clinical and pathological features of this lesion.

As a rule, the tumor develops as a rapidly-growing single subcutaneous tumor in children below the age of two. Although poor circumscription of the tumor makes local recurrence occaisional, it always pursues a completely benign course. Virtually all cases are solitary, and multiple forms are exceedingly rare (Enzinger, 1965); after more than 20 years, 120 cases have been collected including only one case of multiple form (Enzinger, 1988). A single report was found in Korean literature (Suh et al., 1985), where three cases of fibrous hamartoma of infancy were described. All represented as a single mass.

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The case presented here is a multiple form of fibrous hamartoma of infancy in a 10-month-old boy, which recurred one month later after the first local excision. The recurrence might be the result of multiple development de novo rather than incomplete excision.

CASE REPORT

Clinical Findings

A normally developed, healthy, 10-month-old male infant was admitted because of two firm, non-tender and non-movable subcutaneous nodules located in the middle of the left upper arm. The nodules had grown larger over the previous three months, and one of them was firmly attached to the overlying skin. A complete physical examination revealed no palpable nodules in other sites. Symptoms or signs related to these masses were absent. The nodules were completely excised, and there as no remaining mass at the time of operation. The larger one, 4 x 2cm, was in the ventro-lateral aspect and the smaller one, 2×1cm, was in the dorso-medial aspect. Two masses were 3cm apart and separated by normal subcutaneous tissue.

One month after the initial excision, the patient was brought back to the hospital for a newly growing mass, 2×2cm, in the medial aspect of the left upper arm 2cm below the previous operation field (Fig. 1). This recur-

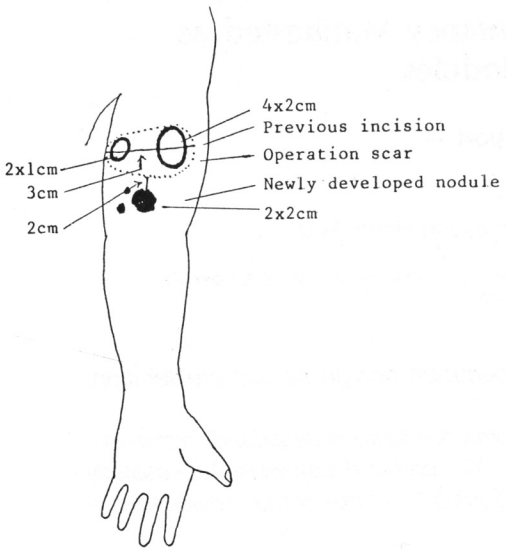

Fig. 1. Two discrete nodules were found in the upper arm initially. One month later, multiple nodules developed 2cm below the operation scar.

rent mass had grown very rapidly over one month period and was freely movable. At surgery, one thumb tip sized and two pea-sized nodules were found in the subcutaneous tissue. There was no recurrent mass at the previous excisional site. Wide excision was carried out.

The child has been well up until today, at the age of 20 months, without recurrence.

Pathologic Findings

The specimen at the first operation consisted of two lumps of firm fibroadipose tissue which measured 4.5×2.5×2cm and 5×3×2cm, respectively. They were not encapsulated. The cut surfaces were glistening gray-white with irregular islands of yellow fat and focal myxoid areas.

The specimen from the second operation consisted of several pieces of fibroadipose tissue, ranging from 1.2cm to 3.5cm. The gross appearance was identical to that of the previous operation.

Histologic examination: Multiple nodules were strikingly similar in histologic findings. Three distinct types of tissue, that is, fibrous trabeculae, myxovascular area and adipose tissue, were intermixed, forming a vague organoid pattern (Fig. 2). The fibrous trabeculae, which was composed of bundles or fascicles of fibrocytes and considerable amounts of mature collagen, comprised the major component of this tumor. The second, myxovascular tissue consisted of bipolar or stellate spindle cells in myxoid and richly vascular stroma. Lymphocytic infiltration was present in these myxoid areas. Alcian blue stain revealed that the myxoid area contained acid-mucopolysaccharide. There were also islands of mature adipose tissue closely intermixed with the above two components (Fig. 3). There was no capsulation and the tumor showed infiltrative growth to the surrounding normal subcutaneous tissue. No cellular anaplasia or mitotic figures were encountered (Fig. 4).

Minute nodules from the second operation also disclosed same histologic features. Deposition of hyalinized material in the myxovascular area was seen more frequently than in the previous operation.

DISCUSSION

Fibrous hamartoma of infancy 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. The principal location is the axilla, followed by the upper arm, thigh, inguinal region, shoulder, back, and forearm (Enzinger, 1988). Until now, single cases have been reported emphasizing unusual locations (Dehner, 1987; MItchell et al., 1982; Robbins et al., 1970; Wexler et al., 1979). A case report showing development in an internal organ was also reported (Baarsma, 1979). It is three times more common in boys than in girls.

Virtually all cases are solitary. Multiple forms are exceedingly rare. The present case is a rare example of multiple fibrous hamartoma of infancy in that two nodules developed synchronously at the same site, and three other nodules occurred a short time after the first excision. Although the latter nodules developed in the same upper arm after local excision, the site was 2 cm distant from the first operation site. There is no difference clinically or histologically from single to multiple forms. Further elucidation of multiple forms may be possible as cases accumulate. There is no evidence of familial incidence (Enzinger, 1988). Although congenital forms are not uncommon, no associated maliformations or other neoplasms have been found and the rarity of multiple forms differentiate this tumor from other congenital fibrous proliferations of infancy.

The tumors are usually poorly circumscribed and blend with the surrounding subcutaneous tissue (Enzinger, 1965). When the tumor is incompletely excised due to poor demarcation, recurrence may follow. However, its clinical course is completely benign. It is important to recognize this tumor properly and not overtreat it (Lee et al., 1988).

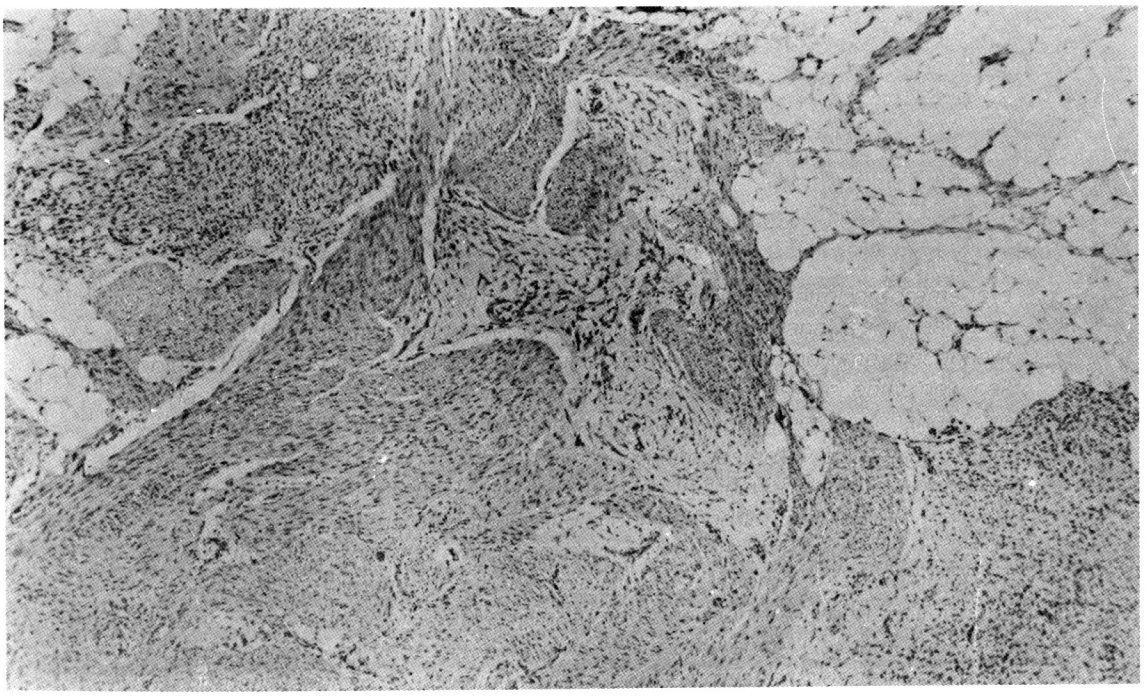

Fig. 2. Close intermingling of distinct three patterns in one field. Fibrous trabeculae is the major component of this tumor (H & E, ×40).

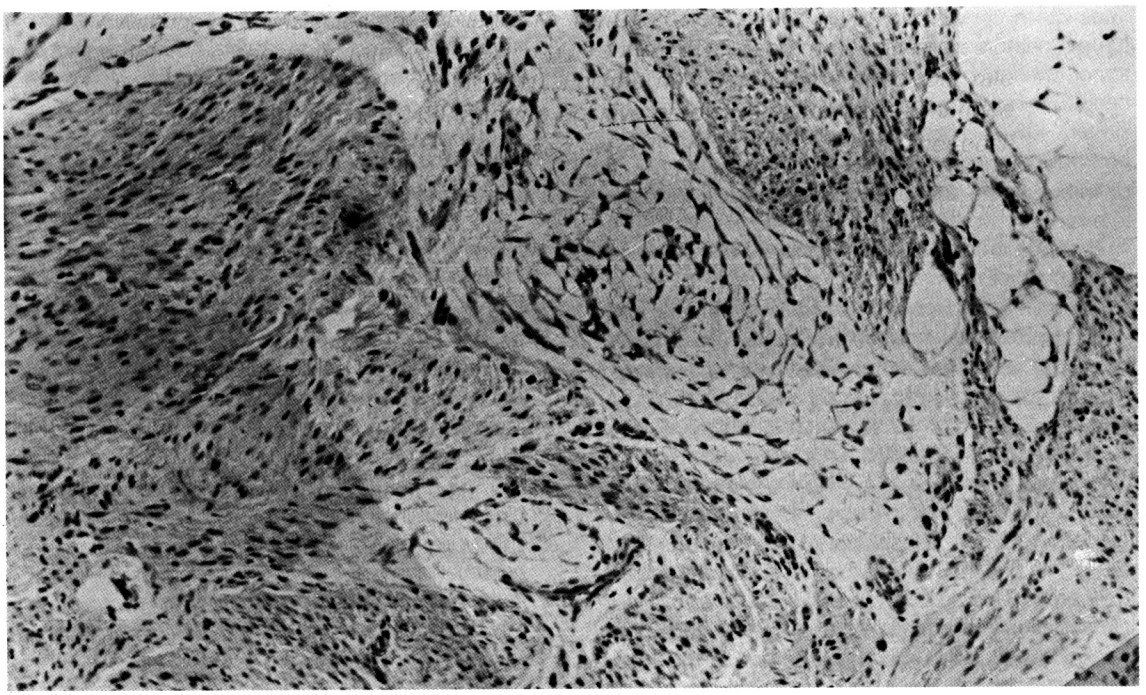

Fig. 3. Spindle or stellate shaped mesenchymal cells and blood vessels in myxoid area, admixed with mature fibrous tissue and fat (H & E, ×100).

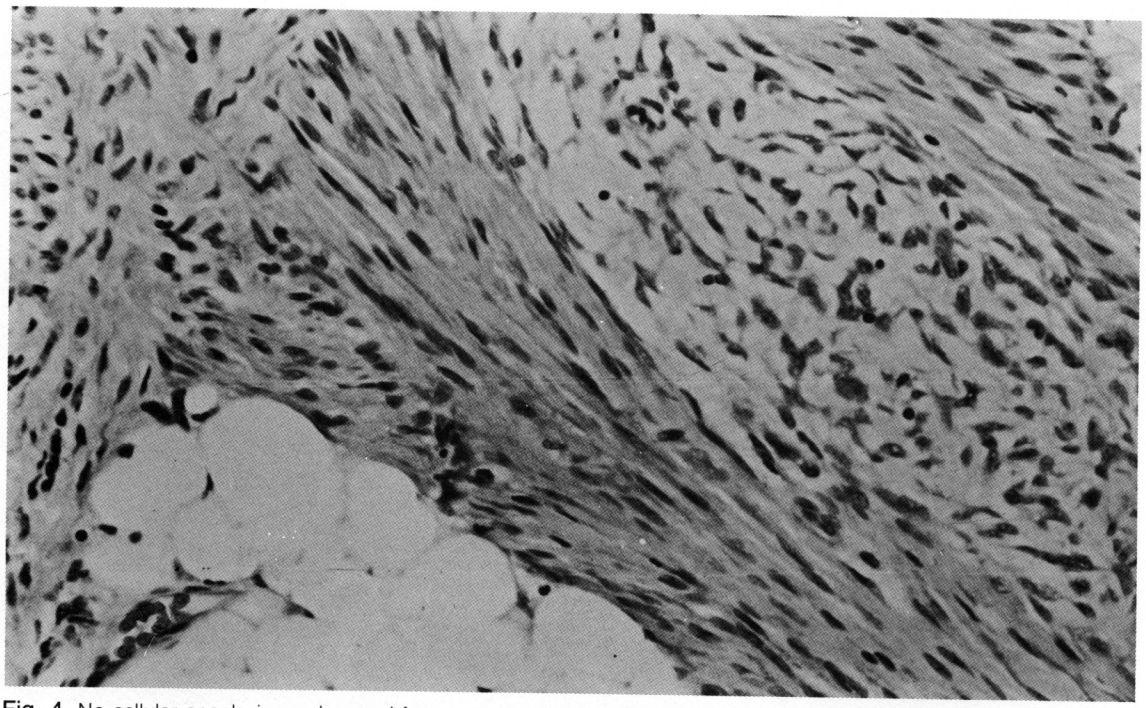

Fig. 4. No cellular anaplasia or abnormal forms are present in any three components of the tumor (H & E, ×200).

These tumors also have very distinct histologic features which clearly differentiate it from other fibrous tumors once a pathologist has experienced it. The tumor invariably shows three different tissue components forming a vague, irregular, organoid pattern (Enzinger, 1988). These elements occur in varying proportions. They include 1) intersecting trabeculae of fibrous tissue, 2) loosely textured areas of immature mesenchymal cells, and 3) mature adipose tissue. The immature appearance of primitive mesenchymal cells in myxoid areas mimicks some malignant lesions. However, there is no anaplasia, and mitotic figures are infrequent (Enzinger, 1965; Lee et al., 1988). Enzinger emphasized this remarkable uniformity of microscopic and clinical features. He suggested the term "fibrous hamartoma of infancy" due to its organoid microscopic structures and frequent occurrence at birth and in the immediate postnatal period (Enzinger, 1965), 1965). His assumption was later confirmed by ultrastructural study (Greco et al., 1984). The principal cells in the fibrous and myxovascular areas were spindle-shaped or stellate. The stellate cells showed immature mesenchymal cells, and the spindle-shaped cells showed fibroblasts. There were also transitional cells or preadipose fibroblasts, as well as mature fat cells in the fat, which were considered an early step in the maturation of adipose tissue. Myofibroblasts were also demonstrated (Benjamin et al., 1977; Greco et al., 1984). The blood vessels in the myxoid area were also prominent, showing features of growing capillaries. This ultrastructural study demonstrated the hamartomatous nature of the tumor.

When Reye first described this lesion, he suggested that progressive maturation to adult fibrous tissue might be a reparative process rather than a true neoplastic proliferation (Reye, 1956).

Since fibrous hamartoma of infancy 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. Benjamin et al. reported such a case, in which myofibroblasts were demonstrated by electron microscopy (Benjamin et al., 1977). They suggested that the contraction of myofibroblasts was responsible for tumor regression. In point of view of treatment, it may be that operative removal is not the best approach. A period of observation might be considered in the hope that the active phase will subside and the mass become reduced to an area of quiescent scar tissue (Reye, 1956).

Local recurrence of the tumor after incomplete excision has been reported, and it has been treated suc-

cessfully by wider local excision (Enzinger, 1965). The tumor has irregular boundaries, and outposts of the main growth are to be found at a distance as minute nodules embedded in the fat (Reye, 1956). For this reason, total extirpation cannot be achieved in the first operation. Lately-developed nodules in the present case may have originated from these satellite minute nodules however, the operative finding strongly suggests the possibility of truly multiple occurrence. The nodules developed in a very short time after the first operation. There is no clear description of whether there is any relation between multiplicity and recurrence. However, we think that metachronously developed nodules from multiple foci de novo may manifest as recurrent lesions clinically. When multiple nodules occur, close follow-up is recommended to find newly-developing nodules in the vicinity.

REFERENCES

Baarsma EA: *Juvenile fibrous hamartoma of the pharynx. J Laryngol Otol 93: 75-79, 1979.*

Benjamin SP, Mercer RD, Hawk WA: *Myofibroblastic contraction in spontaneous regression of multiple congenital mesenchymal hamartomas. Cancer 40: 2343-2352, 1977.*

Dehner LP: *Soft tissue, peritoneum, and retroperitoneum. In: Pediatric surgical pathology. 2nd ed. Baltimore; Williams & Wilkins, p887, 1987.*

Enzinger FM: *Fibrous hamartoma of infancy. Cancer 18:241-248, 1965.*

Enzinger FM, Weiss SW: *Fibrous proliferations of infancy and childhood. in Soft tissue tumors. 2nd ed. St. Louis, Missouri: The C.V. Mosby Company, pp164-168, 1988.*

Greco MA, Schinella RA, Vuletin JC: *Fibrous hamartoma of infancy. An ultrastructural study. Hum Pathol 15:717-723, 1984.*

Lee JT, Girvan DP, Armstrong RF: *Fibrous hamartoma of infancy. J Pediatric Surg 23:759-761, 1988.*

Mitchell ML, DiSant'Agnese PA, Gerber JE: *Fibrous hamartoma of infancy. Hum Pathol 13:586-588, 1982.*

Reye RDK: *A consideration of certain subdermal "fibromatous tumors" of infancy. J Pathol Bact 72:149-154, 1956.*

Robbins LB, Hoffman S, Kahn S: *Fibrous hamartoma of infancy. J Plastic Reconstr Surg 46:197-200, 1970.*

Suh EH, Cho KJ, Ahn KH, Chi JG: *Fibrous hamartoma of infancy: Three cases report. Kor J Pathol 19:202-295, 1985.*

Wexler JAH, Dana M, Silverman J: *A case of fibrous hamartoma of infancy. J Pediatric Surg 14:80-82, 1979.*