Infantile Digital Fibromas

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A series of 15 digital lesions in 8 pediatric patients is presented. All infantile digital fibromas appear early in life at the distal or proximal interphalangeal joint level, characteristically involve the ulnar three digits, may be multicentric, and undergo rapid growth within a short time. Histology reveals deep dermal origin and intracytoplasmic inclusion bodies not observed in any other fibrous proliferative disorders. A viral etiology has never been proved. The lesions may recur despite wide excision followed by resurfacing with a flap or graft. This distinctive lesion has no adult counterpart and should be considered in the differential diagnosis of hard, immobile distal digital masses in children. (J Hand Surg 1995;20A:1014-1020.)

Infantile digital fibroma (IDF) is an unusual fibrous proliferative disease of the upper extremity occurring in childhood. Although there are many similarities to other adult and pediatric fibrous lesions, this tumor characteristically demonstrates a predictable growth pattern and a distinctive histology. Its etiology is not known. Familiarity with the typical features of this tumor should permit its ready discrimination from other fibrous proliferative lesions, including malignancies with a potential for metastasis, and should direct appropriate therapy.

Case reports of tumors that probably represented IDF appeared as early as 1924. A series of seven patients whose presentation was consistent with this diagnosis was cited in 1957 from the Children's Hospital, Boston, and the term neurofibrosarcoma was used. Infantile digital fibromatosis was recognized as a distinct entity and definitively described in 1965 by Reye. Since then more than 100 cases of IDF have appeared under various appellations, but citations within the hand literature have been scarce.

This series of multiple lesions observed in eight pediatric patients accentuates the specific characteristics of this particular lesion in contrast to fibrosarcomas occurring early and later in life.

Materials and Methods

This retrospective review includes eight patients treated between 1977 and 1991 (Table 1). Each child presented between 5 months and 6 years of age. All were managed with initial observation followed by surgical excision. Two patients were treated at other hospitals and referred for follow-up examination; one of these required re-excision and grafting. Patients with similar lesions involving toes were excluded from this study. Those with a follow-up period of less than 3 years were also excluded.

Results

Eight pediatric patients were treated for lesions involving 10 separate digits, 4 of which contained 2 separate masses. The average age of presentation was 18 months, with a range of 5 to 68 months. No lesions were noted at or shortly after birth, the earliest lesion being recognized at age 2 months. The follow-up period ranged from 3 to 14 years (Table 1).

The growth in all cases was described as slow but progressive. Characteristically, small rapidly growing lesions first appeared dorsal or lateral to the distal interphalangeal joint, and in four digits a second, distinctly separate lesion appeared over the proximal

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Table 1. Infantile Digital Fibromas

<table>
<thead>
<tr>
<th>Patient No.</th>
<th>Sex</th>
<th>Age First Examined</th>
<th>Digit</th>
<th>First Recognized</th>
<th>Treatment</th>
<th>Recurrence</th>
<th>Follow-up Period (y)</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>F</td>
<td>5 months</td>
<td>R fifth</td>
<td>2 months</td>
<td>XF flap, FTSG</td>
<td>no</td>
<td>4</td>
</tr>
<tr>
<td>2</td>
<td>M</td>
<td>28 months</td>
<td>R long</td>
<td>18 months</td>
<td>FTSG</td>
<td>no</td>
<td>6</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>R ring, DIP</td>
<td>30 months</td>
<td>FTSG</td>
<td>no</td>
<td>5</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>R ring, PIP</td>
<td>31 months</td>
<td>Flap</td>
<td>no</td>
<td>5</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>R fifth, DIP</td>
<td>38 months</td>
<td>FTSG</td>
<td>no</td>
<td>3</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>R fifth, PIP</td>
<td>38 months</td>
<td>Flap</td>
<td>no</td>
<td>3</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>R fifth, metacarpal</td>
<td>56 months</td>
<td>Closure</td>
<td>no</td>
<td>4</td>
</tr>
<tr>
<td>3</td>
<td>F</td>
<td>8 months</td>
<td>L ring</td>
<td>3 months</td>
<td>FTSG</td>
<td>no</td>
<td>7</td>
</tr>
<tr>
<td>4</td>
<td>F</td>
<td>30 months</td>
<td>L index</td>
<td>23 months</td>
<td>FTSG</td>
<td>yes</td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>L index</td>
<td></td>
<td>FTSG*</td>
<td>no</td>
<td>8</td>
</tr>
<tr>
<td>5</td>
<td>M</td>
<td>22 months</td>
<td>R long, DIP</td>
<td>16 months</td>
<td>FTSG</td>
<td>no</td>
<td>11</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>R long, PIP</td>
<td>20 months</td>
<td>Flap</td>
<td>no</td>
<td>11</td>
</tr>
<tr>
<td>6</td>
<td>F</td>
<td>18 months</td>
<td>R ring</td>
<td>22 months</td>
<td>XF flap, FTSG</td>
<td>no</td>
<td>14</td>
</tr>
<tr>
<td>7</td>
<td>M</td>
<td>6 years</td>
<td>L long</td>
<td>5 years</td>
<td>Local flap</td>
<td>no</td>
<td>3</td>
</tr>
<tr>
<td>8</td>
<td>F</td>
<td>11 months</td>
<td>R ring</td>
<td>9 months</td>
<td>FTSG</td>
<td>no</td>
<td>4</td>
</tr>
</tbody>
</table>

FTSG, full-thickness skin graft; PIP, proximal interphalangeal joint level; DIP, distal interphalangeal joint level; flap, local dorsal rotation flap; XF flap, cross-finger flap.

*Recurrent mass hard to distinguish from hypertrophic scar.

Discussion

Infantile digital fibroma has a number of features in common with other fibromatous conditions, that distinguish them from nonproliferative lesions, such as scar on one end of a wide spectrum and malignant neoplasms on the other.3,4 The defining characteristics of fibromatoses, introduced by Stout in 1954 and refined by Becker and Chait in 1979, include (1) proliferation of well-differentiated fibroblasts; (2) an infiltrative pattern of growth; (3) the presence of a variable (but usually abundant) amount of collagen between proliferating cells; (4) a lack of cytologic features of malignancy and scanty or absent mitotic activity; and (5) aggressive clinical behavior characterized by local recurrences but without the capacity to produce distant metastases.

Infantile digital fibroma is one of several fibromatous disorders appearing in childhood (Table 2). In its characteristic form, it is not known to occur in adulthood.3,4,9-15 The incidence of IDF is highest during the first 3 years of life, with as many as one third of patients presenting soon after birth. The few cases that have been reported in older patients were atypical in history and in anatomic location.4,5,7

The characteristic lesion of IDF is a firm, broad-based and nontender nodule, which appears innocuously on the side of a digit. Its color may be pale red or may be identical to that of the surrounding skin. Nodules may occur alone or may be multicentric. Lesions are usually confined to the digits—fingers, toes, or both. One of our patients developed lesions over the dorsal metacarpal portion of the hand. The lesions have a predilection for the distal portion of the digit and for opposing surfaces of adjacent digits and most often appear on the sides or dorsum. In the present study, lesions at the distal interphalangeal joint level were located on the side of the digit; those at the proximal interphalangeal level were dorsally situated (Figs. 1, 3). The nodules of the IDF present preferentially on the ulnar three digits and have not been reported to occur on the thumb or great toe.
Figure 1. Infantile digital fibromas typically appear along the midlateral borders of the digit at the distal interphalangeal joint or within the dorsolateral skin at the proximal interphalangeal joint. On presentation to the hand surgeon, lesions may characteristically have grown rapidly to this size. Many are misdiagnosed as fibrosarcomas.

Figure 2. Options for surgical reconstruction after en bloc removal include local flap advancement at the proximal interphalangeal joint (top, center), hypothenar full-thickness skin grafts, and cross-finger flaps. Matching the glabrous and non-hairbearing portions of the graft to the digit will yield improved conformity to the graft (left). Cross-finger flaps from the side of adjacent digits are used only for joint exposure.

The usual course of IDF is one of gradual progression. Lesions may enlarge grossly and have been reported to produce deformity of interphalangeal joints. No joint distortion was noted in the present series. Following excision, the rate of recurrence at the same site or elsewhere in the digit has been as high as 60%. Spontaneous regression has also been reported; the true incidence of regression is not known, since most investigators have chosen surgical intervention over observation.

The morphology of IDF lesions is similar to that of other fibromatoses: a firm, poorly circumscribed mass with a white cut surface. The proliferative process appears to be limited to the dermis, with extension up to the epidermis and deep structures and compression of the same. Epidermal changes are minimal and generally consist of hyperkeratosis and acanthosis.

By light microscopy, the lesions of IDF display a relatively uniform proliferation of fibroblasts in a
Table 2. Fibrous Proliferations of Infancy and Childhood

<table>
<thead>
<tr>
<th>Diagnosis</th>
<th>Reference</th>
<th>Location</th>
<th>Recurrence</th>
</tr>
</thead>
<tbody>
<tr>
<td>Infantile digital fibromatosis</td>
<td>Sakurane,¹ 1924, Reye,³ 1965</td>
<td>Digits</td>
<td>Common</td>
</tr>
<tr>
<td>Juvenile oponeurotic fibroma/calcifying fibroma</td>
<td>Keusbey,²⁶ 1953</td>
<td>Palmar and plantar surfaces</td>
<td>Common</td>
</tr>
<tr>
<td>Infantile myofibromatosis/congenital fibromatosis</td>
<td>Stout,⁷ 1954, Schnitka et al.,²⁷ 1958</td>
<td>Soft tissue, bone, and viscera</td>
<td>Rare</td>
</tr>
<tr>
<td>Infantile desmoid type fibromatosis/infantile and juvenile fibromatosis</td>
<td>Stout,⁷ 1954</td>
<td>Musculature</td>
<td>Common</td>
</tr>
<tr>
<td>Giant cell fibroblastoma</td>
<td>Shmookler and Enzinger,²⁶ 1982</td>
<td>Thigh, back, inguinal region</td>
<td>Common</td>
</tr>
<tr>
<td>Hyaline fibromatosis</td>
<td>Murray,²⁶ 1873</td>
<td>Dermis and subcutis</td>
<td>—</td>
</tr>
<tr>
<td>Fibrous hamartoma/subdermal fibromatous tumor of infancy</td>
<td>Reye,⁴ 1956</td>
<td>Axillary and inguinal regions</td>
<td>Rare</td>
</tr>
<tr>
<td>Fibromatosis colli</td>
<td>Taylor,⁴¹ 1875</td>
<td>Sternocleidomastoid muscle</td>
<td>Rare</td>
</tr>
</tbody>
</table>

Figure 3. (A) Appearance of original hand mold and (B) of the same hand 3 years following wide excision and grafting. Large lesions on the long (ulnar) and fifth (radial) have been grafted and one on the ring (ulnar) has been closed with a local flap. Residual soft tissue deformities are seen. (Figure continues.)
dense collagenous stroma (Fig. 4A). Mitoses are scarce but have been described. The presence of cytoplasmic inclusion bodies permits histologic distinction of IDF from other fibromatoses. These structures are small (3-15 μm in diameter), round or polygonal in shape, and usually located near the nucleus. Although variable in number, they are invariably present in each lesion (Fig. 4B).

The staining characteristics of these inclusion bodies are distinctive. In addition to being eosinophilic, they stain red with Masson trichrome, purple with phosphotungstic acid-hematoxylin, and black with iron hematoxylin. Before the advent of electron microscopy and advanced histochemical techniques, they were thought to contain either RNA, elastic fibers, or simply an “abnormal by-product of metabolically deranged fibroblasts.” More recently, Iwasaki and colleagues have shown conclusively that the inclusion bodies contain densely packed microfilaments; the significance of this finding is not understood.

The cellular component of IDF consists of fibroblasts and myofibroblasts. Contractile proteins have been identified free in the cytoplasm as well as within cytoplasmic inclusion bodies. The cells of IDF typically have abundant endoplasmic reticulum in the vicinity of the nucleus and a prominent Golgi zone.

The etiology of IDF is not known. Its proliferative histology and the tendency to recur locally and regionally are characteristic of a neoplastic process, but the lesion lacks cytologic features of malignancy and has not been associated with distant metastases. The occurrence of extradigital post-

Figure 3. (continued) (C, D) Postoperative appearance of a similar lesion in an 18-month-old child treated with a cross finger flap for coverage of an exposed distal interphalangeal joint.
traumatic lesions$^{16,31}$ that were histologically indistinguishable from digital fibroma has led to the suggestion that IDF is physiologically related to scar.

A viral origin has been proposed because of the anatomic distribution of synchronous and recurrent lesions on the same or adjacent digits.$^2$ The hallmark intracytoplasmic inclusions were previously thought to resemble viral particles,$^7,29$ but electron microscopy has not confirmed this observation.$^{24-26,31,32}$ Pohjanpelto et al.$^{24}$ isolated a filterable cell-transforming agent from nodules of IDF, a finding that has never been reproduced. In the present study no attempts were made at viral isolation.

The characteristic clinical presentation of a rapidly growing, hard nodule on the dorsal or lateral surface of a digit in a child less than 3 years of age is almost always diagnostic of IDF and easily distinguishes it from a wart (verruca vulgaris), scar tissue, a retained foreign body, or an inclusion cyst. Digital fibromas should not be confused with palmar and plantar fibromatoses, which also grow rapidly and frequently have a histologic appearance consistent with fibrosarcoma (Table 2). The dermal origin and spread of IDF tumors can be insidious, which relates to the high recurrence rate following inadequate excision. Early intralesional steroid injection did not affect growth of two tumors in one of our patients, although Tsuge$^{35}$ has reported complete resolution of one case with steroid tape.

Wide excision is presently the treatment of choice. Although proliferative cells have not been demonstrated to invade collateral ligaments, bone tendon, or retinacular sheaths, portions of periosteum and ligaments have been excised to ensure complete removal. Despite loupe magnification during dissection, it is difficult to discern a plane between these dense lesions and a normal ligament.

Although cross-finger flaps have been necessary for resurfacing exposed joint spaces, wound coverage with hypothenar skin grafts has been our preferred treatment. Matching the glabrous and nonglabrous portions of the hypothenar donor site to the defect on the digit provides excellent results. The ellipse of donor skin can be harvested in either a horizontal or vertical direction (Fig. 2).

References