

# Dupuytren's Disease in Children—Differential Diagnosis

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**Background/Purpose:** Dupuytren's disease in children is uncommon; only a few histologically confirmed diagnoses are found in literature. In its early, proliferative phase with many fibroblasts, it can resemble the early stadium of an epitheloid sarcoma, which also is a rare tumor. The purpose of this report is to show pediatric surgeons and pathologists the importance of differential diagnosis of nodules and fibrotic bands in children's hands, especially to exclude a malignancy.

**Methods:** Between 1998 and 2000, 3 children at ages of 2½ years, 9 years, and 10 years, respectively, presented with the clinical signs of Dupuytren's disease of the hand. Each of them had fibrous bands with a flexion contracture of the fingers. All of them underwent fasciectomy.

**Results:** In patient 1, a 10° extension lag of the index finger resulted after 3 operations. Histology findings showed the typical features of Dupuytren's disease. In patient 2, histology findings showed a recurring digital fibroma of child-

hood. After consultation with the pathologist and reevaluation of the slides, the histologic diagnosis was corrected to Dupuytren's disease. Full extension and thumb abduction has been achieved. In patient 3, histology of the first and second operation was misdiagnosed as Dupuytren's disease. In the third operation 2½ years later, the histologic diagnosis of epitheloid sarcoma was made. Amputation of the forearm was necessary.

**Conclusions:** Exact anamnesis, location of the lesion, and suspicious diagnosis must be mentioned to the pathologist in the case of biopsy or excision of Dupuytrenlike lesions in children's hands. Awareness of the possibility of the epitheloid sarcoma may help prevent misdiagnosis.

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**INDEX WORDS:** Dupuytren's disease in children, epitheloid sarcoma.

**D**UPUYTREN'S DISEASE in children is uncommon. There only are a few reports of histologically confirmed Dupuytren's contractures in children under 10 years of age.<sup>1-6</sup> Differential diagnosis is important to exclude other causes of flexion contractures in hands with enlarging nodules and fibrotic bands, especially malignancies. We report on 3 children under 10 years of age with the clinical diagnosis of Dupuytren's disease. One of them had an epitheloid sarcoma of the hand and, therefore, needed amputation of the forearm.

## MATERIALS AND METHODS

Between 1998 and 2000, 3 children (ages 2½ years, 9 years, and 10 years, respectively) presented with the clinical signs of Dupuytren's disease of the hand. Each of them had fibrous bands with a flexion contracture of the fingers. In 1 case, age of onset was birth. There was no history of trauma. All of them underwent fasciectomy.

### Patient 1

A 2½-year-old girl presented with a 6-month history of progressive flexion contracture of the left index finger without preceding trauma. Clinical examination showed a fibrous band typical for Dupuytren's disease, extending from the metacarpophalangeal (MP) joint to the distal phalanx, resulting in a 50° extension lag of the proximal interphalangeal (PIP) joint (Fig 1A). X-ray was unsuspecting. Excision of the fibrotic band was performed.

### Patient 2

A 10-year-old boy presented with an adduction contracture of both thumbs, which had been present since birth. At the age of 5, an operation was performed on the left thumb by an orthopedic surgeon

elsewhere for trigger finger. Clinical examination showed a fibrous band on the palmar-radial aspect of both thumbs, each extending from the thenar to the interphalangeal (IP) joint. The left dominant hand had the following contracture: the MP joint was S 0-40-60, the IP joint S 0-0-80, radial abduction was 35°, and palmar abduction 45° (Fig 2A). A fasciectomy was performed on the left thumb (Fig 2B), and the skin was closed with Z-plasties.

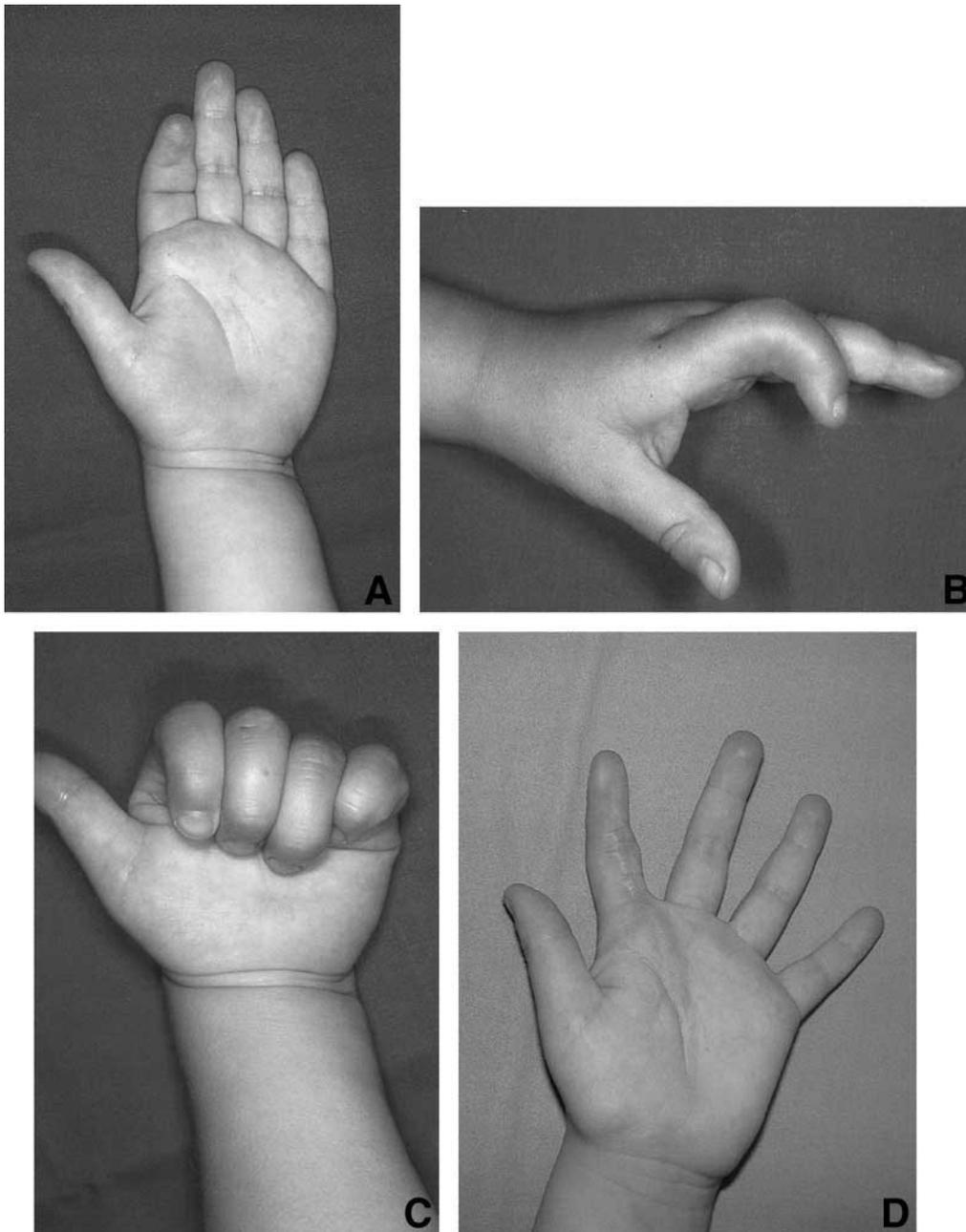
### Patient 3

A 9-year-old boy with a 1-year history of inability to fully extend his right fourth and fifth finger was operated on for "Dupuytren's disease" elsewhere; a partial fasciectomy and an arthrolysis was performed. The wound healing was bad with ulceration of the skin. An extension lag of 10° remained in the MP joints. Histology findings confirmed the clinical diagnosis of Dupuytren's disease. In spite of physical therapy, the flexion contracture progressed to 50°. Finger abduction was not possible and sensibility was normal. Electroneurography of the ulnar nerve was unsuspecting. Magnetic resonance imaging (MRI) showed postinflammatory granulation tissue of the interosseous and lumbrical muscles and discreet erosive cortical lesions of the metacarpals. Operative exploratory 10 months after the first operation showed massive

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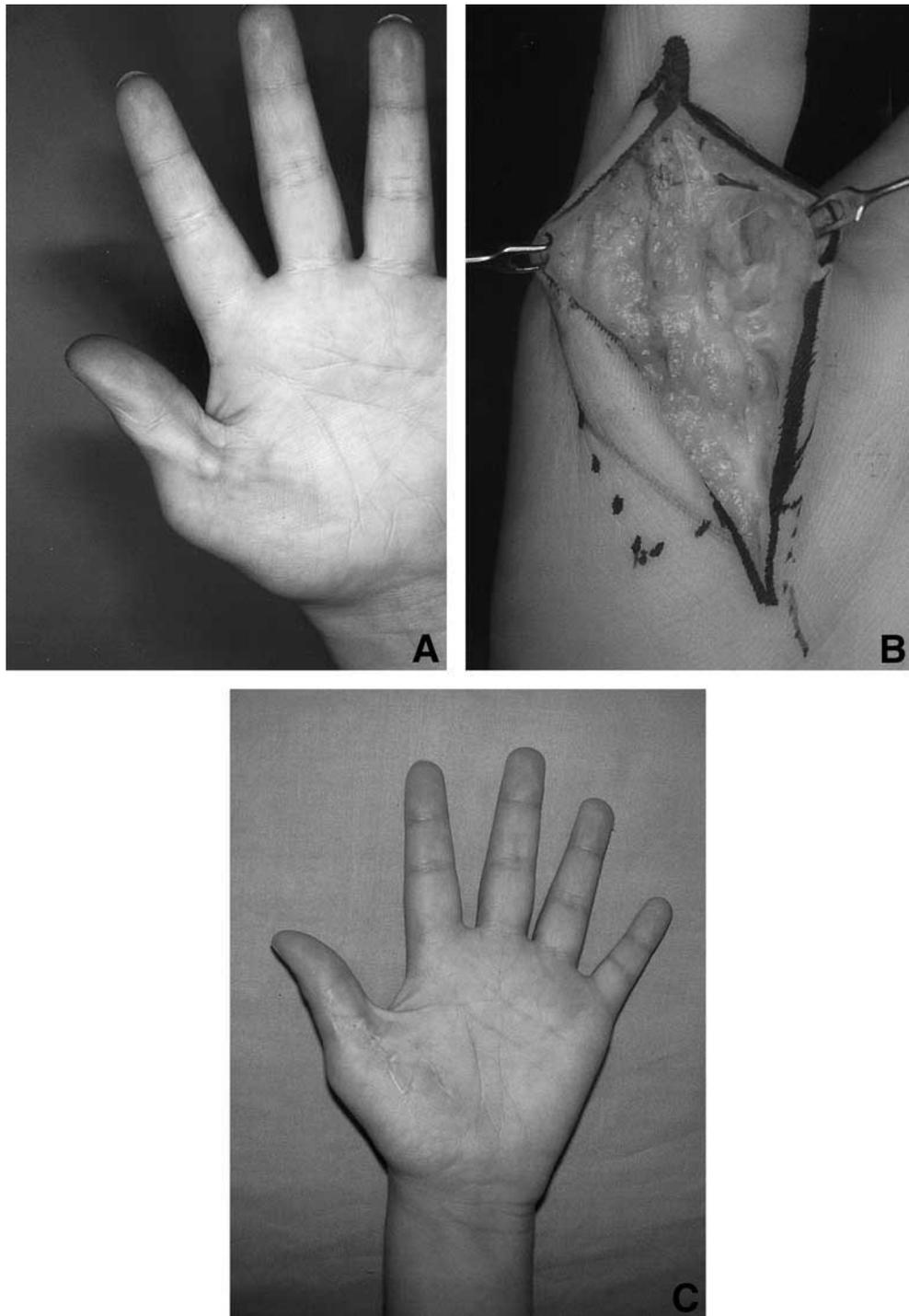
**Fig 1.** Patient 1, a 2½-year-old girl. (A) Fifty-degree extension lag of the left index finger caused by Dupuytren's contracture. (B) Bad result, 4 months after excision of the fibrotic band. Postoperative physical therapy was insufficient because of child's poor cooperation. (C) Possibility of nearly complete flexion of the index finger 4 weeks after the third operation. (D) Ten-degree extension lag of the left index finger after 3 years.

scar tissue around the fourth and fifth metacarpal bones. There was no fifth lumbrical muscle discoverable. A skin defect of the palm resulted, which had to be covered with an abdominal pedicled flap. Histology of the fibrous tissue showed again the features of Dupuytren's disease. Two weeks later, the flap's pedicle was cut (Fig 3A and B).

### RESULTS

In patient 1, full extension of the index finger was achieved. Histology findings showed the typical features

of Dupuytren's disease. Physical therapy was difficult because of poor cooperation by the child; a flexion contracture of 80° developed in the PIP joint and of 10° in the distal interphalangeal (DIP) joint (Fig 1B). In a second operation, fibrous tissue was excised from the index finger and a Z-plasty was performed. Histology findings showed scar tissue. The postoperative result was bad with continuous flexion contracture. A third opera-

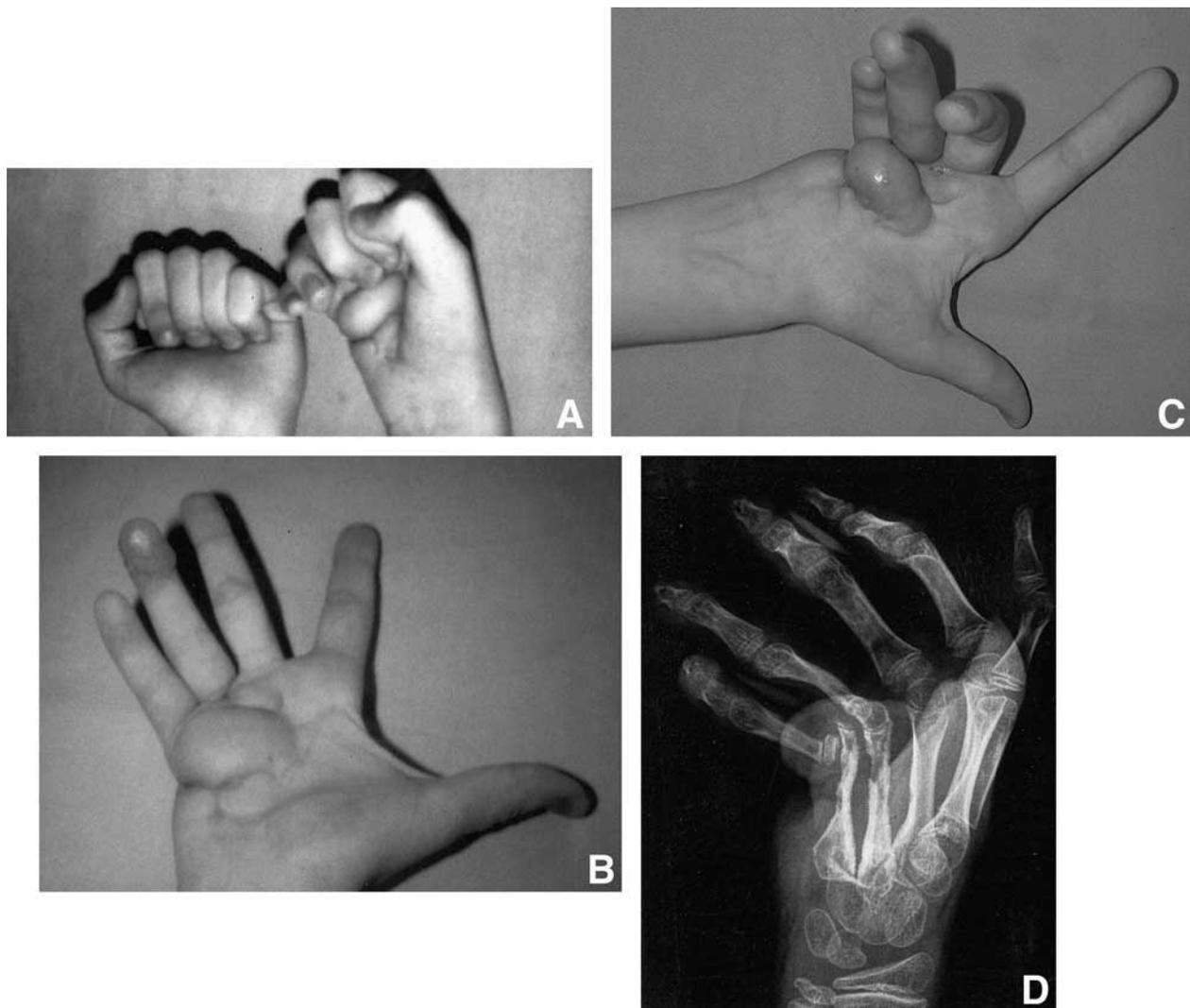


**Fig 2.** Patient 2, a 10-year-old boy. (A) Flexion contracture and adduction contracture of the left thumb. (B) Intraoperative view shows a fibrotic band. Histology first showed a recurrent digital fibroma of childhood, later it was corrected to Dupuytren's disease. (C) Full extension and abduction of the left thumb after 2 years.

tion was performed 5 weeks later with excision of fibrous tissue and arthrolysis of the PIP joint. Histology findings showed scar tissue again. Finally, by intensive physical therapy and by using a night splint, a good result was

achieved (Fig 1C). Three years after the first operation, an extension lag of only 10° resulted (Fig 1D).

In patient 2, histology findings showed a recurring digital fibroma of childhood. Because of the very rare



**Fig 3.** Patient 3, a 9-year-old boy. (A) Incomplete flexion of the fourth and fifth finger after defect coverage of the palm with a bulky abdominal flap. (B) Incomplete extension of the fourth and fifth finger after defect coverage of the palm. The lesion at the distal part of the fourth finger originates from a trauma and not from the original disease. (C) Sixteen months later; infiltration of the flap by a tumorlike lesion. Increasing flexion contracture of the third, fourth and fifth finger. (D) X-ray 16 months after defect coverage of the palm, shows osteolysis of the fourth and fifth metacarpal bones and partially of the ulnar aspect of the third metacarpal.

location of this disease in thumbs, the pathologist was asked to see the slides once again, after which the histologic diagnosis was corrected to Dupuytren's disease. The postoperative course was uneventful. The child achieved full range of motion of his left thumb. Two years later, there was no recurrence of the disease on the left thumb (Fig 2C) and no progression on the right thumb.

In patient 3, the fourth and fifth finger lost their motor and sensible function completely during the following 16 months after skin defect coverage of the palm (Fig 3C). X-ray after 16 months showed an osteolysis of the fourth and fifth metacarpal bones and partially of the ulnar aspect of the third metacarpal (Fig 3D). MRI scan

showed massive fibrous tissue around the fourth and fifth metacarpals and proximal phalanges. Because of complete loss of function, the fourth and fifth finger ray were resected with "scar tissue." Histology findings showed an epitheloid sarcoma. Computed tomography (CT) scan showed 3 suspicious lesions of the lung and enlarged lymph nodes of the right axilla. These lymph nodes were extirpated, and histology findings showed no malignancy. Finally, the child's parents were convinced of the necessity of amputation of the forearm, which was performed simultaneously with axillary lymph node dissection. After an interdisciplinary consultation with oncologists and parents, the decision was made not to apply chemotherapy unless enlargement of the metastases. Six

months later, the suspicious lesions of the lung had not enlarged. The boy was supplied with a myoelectric prosthesis.

### DISCUSSION

Dupuytren's disease is a slowly growing superficial fibromatosis. The etiology remains unknown. Usually it affects men between 40 and 60 years of age.<sup>7</sup> Histology is characterized by proliferation of fibroblasts in the early stadium and production of collagen fibers later in the course of disease.<sup>7</sup> In 1996, Urban et al<sup>1</sup> reviewed the literature on the occurrence of Dupuytren's disease in children younger than 13 years and stated that this rare diagnosis in children is only tenable where histologic confirmation is available. Large population studies describe a 1% incidence in children under 14 years of age,<sup>8</sup> whereas histologically confirmed diagnoses might have a much lower incidence. Fasciectomy is the choice of treatment in the stadium of flexion contracture (Figs 1A and 2A), whereas conservative treatment is reserved for the early stadium with nodules and fibrotic bands without contracture.<sup>9</sup> In patients 1 and 2, fasciectomy of the fibrotic bands led to good results (Figs 1D and 2C), whereas in patient 1, 3 operations were necessary. Finally, histology findings confirmed the clinical diagnosis in patient 1 and 2. In patient 2, history of adduction contracture of the thumb dated back to birth, which is the youngest histologically confirmed case found in literature.

Extraabdominal fibromatosis<sup>10</sup> is—in contrast to Dupuytren's disease—a rapid growing and infiltrating, non-metastasizing tumor with a recurrence rate of 40%. It originates not only from the aponeurosis but also from the fascia and connective tissue of the muscle. Hands are affected in 1% and children under 10 years of age in 10% of these tumors. Etiology may be genetic, endocrine, or physical. Tumor size often is 5 to 10 cm. Histology describes spindle cells and abundant collagen fibers. Preoperative diagnosis is difficult as well as postoperative differentiation between scar tissue and a recurrent tumor. Therapy is by wide resection; amputation normally is reserved for repeated recurrence. Alternative therapy modalities—mostly used in combination with resection in recurrent tumors—are radiation, endocrine therapy, and chemotherapy.

Recurring digital fibroma of childhood<sup>11,12</sup> is a rare benign fibrous tumor that must be distinguished from Dupuytren's disease. It occurs on the dorsal and lateral aspects of the fingers and toes in children, sometimes multiple, but it usually does not affect the thumb and the great toe. The recurrence rate is high, and sometimes amputation is necessary. Histology findings show elongated spindle cells and abundant collagen fibers, and round eosinophilic inclusions are typical. In patient 2,

recurring digital fibroma was first histologically diagnosed. After giving more clinical information to the pathologist and reevaluating the slides, diagnosis was changed to Dupuytren's disease.

Epitheloid sarcoma was first described by Enzinger<sup>13</sup> in 1970, and then later by others.<sup>12,14-16</sup> Seventy percent of the tumors occur between the ages of 10 and 34 years. Ninety-seven percent are located in the extremities, 34% in fingers and hands. When located in the subcutis the lesion usually presents as a firm nodule that grows slowly and frequently becomes ulcerated 2 or 3 months later. Deep-seated lesions often manifest as a firm area of induration or as a multinodular, lumpy mass, which may resemble Dupuytren's disease. X-ray can show fine flecks of calcium in the tumor and cortical thickening or superficial erosion of the underlying bone, as was described in patient 3 (Fig 3D). Treatment is by excision, and in the case of repeated recurrence, by amputation. Surgical treatment can be accompanied by radiation or chemotherapy. Recurrence rate is 85% and metastases are found in 30%, mainly of the lungs, the pleurae, and the skin of the scalp.

Histology of epitheloid sarcoma shows 2 main cellular elements; spindle cells tend to be more conspicuous in earlier lesions, whereas the polygonal or epitheloid cells predominate in recurrent or metastatic tumors. Because of the slow growth and the harmless appearance during the initial period, the lesion may be mistaken for a benign process such as a fibromatosis, a fibroma, a chronic ulcer, or a granulomatous inflammation, which was described by MRI scan in patient 3. Dupuytren's disease in its proliferative phase with lots of fibroblasts can resemble the early stadium of an epitheloid sarcoma. Not being aware of the existence of the epitheloid sarcoma may contribute to misdiagnosis.<sup>14</sup> Reevaluation of the slides of the first and second histology in patient 3 by another pathologist 2 years later showed the features of an epitheloid sarcoma with nests of epitheloid cells. This proves the histologic misdiagnosis.

Further soft tissue tumors of the hand that have to be considered for differential diagnosis are the juvenile aponeurotic fibroma,<sup>17</sup> the fibroma of tendon sheath,<sup>18</sup> and the giant cell tumor of tendon sheath, now called *localized nodular tenosynovitis*.<sup>19</sup> They all are characteristic of a high recurrence rate because of infiltration of or attachment to the surrounding structures, but they do not metastasize. Histologically, the juvenile aponeurotic fibroma and the fibroma of tendon sheath show mainly fibroblasts and collagen fibers, whereas the giant cell tumor is characterized by giant cells, histiocytes, and collagenized stroma with hemosiderin pigment. Treatment is by excision, and cases of amputation of digits in recurring tumors are reported.

Because of the rare entity of Dupuytren's disease in

childhood, diagnosis of nodules and fibrotic bands in children's hands must be assigned with great caution. Exact anamnesis, location of the lesion, and suspicious diagnosis must be mentioned to the pathologist in the

case of biopsy or excision. Immunohistochemistry may be helpful in diagnosing an epitheloid sarcoma. Close-meshed check-ups are necessary to detect recurrence of disease in an early stadium.

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