

# Dupuytren in a Child: Rare Presentation of a Rare Clinical Entity

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Dupuytren disease in children younger than 10 years is rare and only 8 histologically proven cases have been reported. A histologically proven Dupuytren disease in a 10-year-old with an uncommon clinical presentation as a nodule on the radial side of the middle phalanx of the little finger is documented. Dupuytren's disease should be in the differential diagnosis in cases of nodules and contractures in the palm and fingers of children. (*J Hand Surg Am.* 2016; ■(■): ■–■. Copyright © 2016 by the American Society for Surgery of the Hand. All rights reserved.)

**Key words** Dupuytren disease, child, finger contracture, nodule.



**D**UPUYTREN DISEASE IN CHILDREN younger than 10 years is rare with only 8 histologically proven cases previously reported.<sup>1–8</sup> All the cases presented with either a nodule in the palm or a contracture in 1 of the fingers. We report a histologically proven case of Dupuytren disease in a 10-year-old child with an uncommon presentation as a nodule in the ring and little fingers.

## CASE REPORT

A 10-year old boy presented to our institution with a nodule on the radial side of the distal interphalangeal joint of the little finger that was associated with a contracture of the joint (Fig. 1). The family reported that the finger developed the contracture gradually over a period of about 4 months. The finger could not be

passively extended to a straight position. There was no family history of Dupuytren disease, diabetes, antiepileptic drugs, Scandinavian or Northern European ancestry, Ledderhose or Peyronie disease, nor alcohol consumption. A magnetic resonance imaging study was obtained and reported as a suspected fibrous histiocytoma. During surgery, the lesion was excised to the underlying bone because it was adjacent to it and this allowed the finger to be fully extended. A skin graft measuring 1 cm × 2 cm was placed on the periosteum to achieve wound closure. The histopathological examination of the lesion showed uniform bland spindle cells with elongated normochromatic nuclei without atypia and infrequent mitoses (Figs. 2, 3). These cells were arranged in fascicles in collagenous stroma. Perinuclear cytoplasmic inclusions were not observed.

Immunohistochemically, the cells were positive for smooth muscle actin and muscle-specific actin and showed focally weak cytoplasmic positivity for anaplastic lymphoma kinase (ALK). Nuclear positivity for ALK was not observed. The histology report indicated Dupuytren disease. Over a 4-month period following the surgery, the skin graft contracted and the finger developed a radial drift at the level of the distal interphalangeal joint. During this time a new mass appeared on the ulnar side of the middle phalanx of the ring finger (Fig. 4A). Magnetic resonance imaging again suggested that the lesion was a fibrous histiocytoma. This new lesion was excised

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Received for publication April 5, 2016; accepted in revised form August 15, 2016.

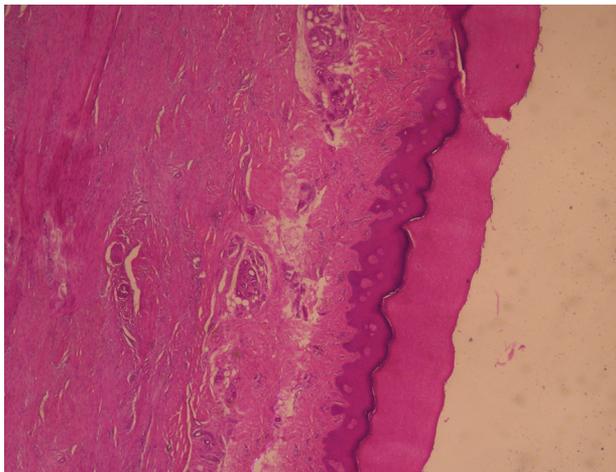
No benefits in any form have been received or will be received related directly or indirectly to the subject of this article.

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<http://dx.doi.org/10.1016/j.jhsa.2016.08.011>

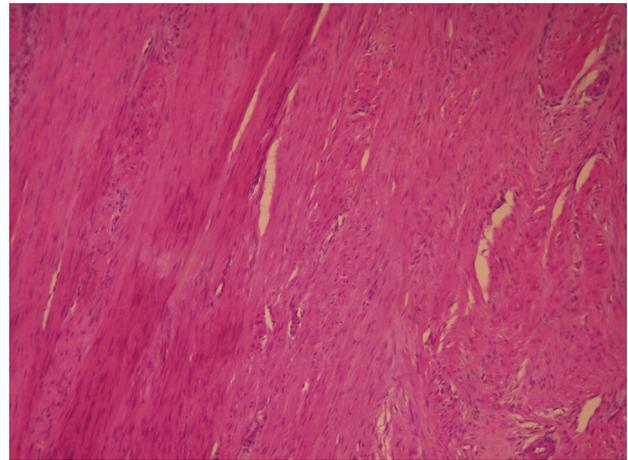


**FIGURE 1:** Presence of a nodule in the radial side of the middle phalanx of the little finger in a 10-year-old child. Consequent contracture occurred gradually over a period of 4 months.



**FIGURE 2:** Fascicles of bland fibroblastic/myofibroblastic cells in collagenous stroma. (Hematoxylin-eosin stain, magnification,  $\times 25$ .)

under general anesthesia 5 months after the first operative procedure on the little finger. During surgery, the lesion seemed to originate from the collateral ligaments. During the same procedure, the radial drift of the little finger was corrected with z-plasties (Fig. 4B). The histology report for the new lesion indicated Dupuytren disease. There were no



**FIGURE 3:** The cells are uniform without nuclear atypia, mitoses, or perinuclear cytoplasmic inclusions. (Hematoxylin-eosin stain; magnification,  $\times 200$ .)

complications following the second procedure and no recurrence 12 months after surgery.

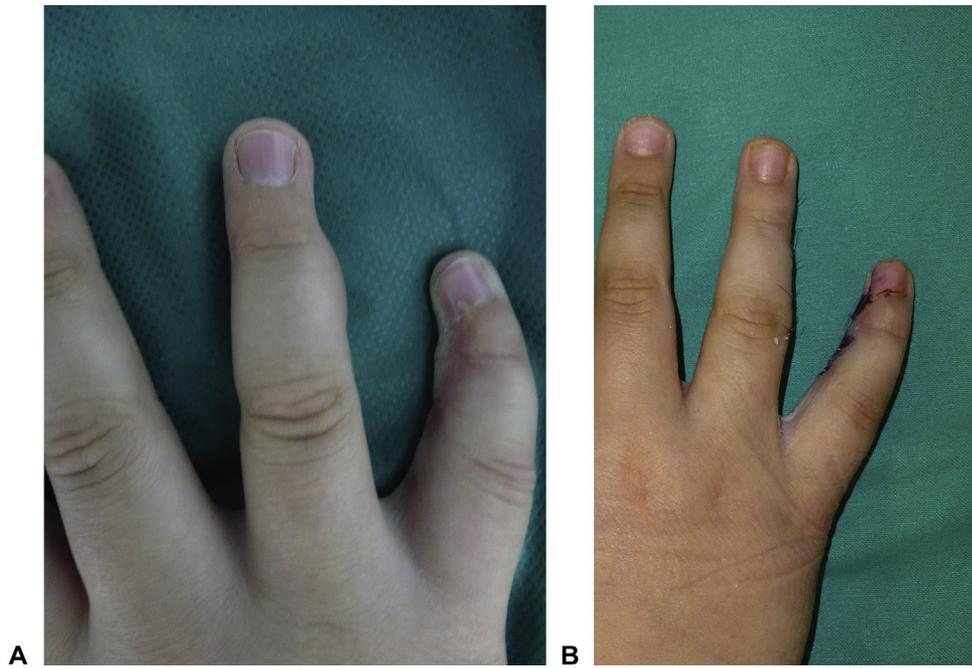
## DISCUSSION

Dupuytren disease in children is exceptionally rare. The most important clinical entity from which it must be distinguished is epithelioid sarcoma,<sup>4</sup> a tumor with epithelioid-like features that occurs mostly between the ages of 10 and 34 years. Ninety-seven percent of cases of epithelioid sarcoma are located in the extremities, mainly the hands and fingers. The tumor has a high recurrence rate (85%), and metastasis occurs in 30% of cases.

Apart from epithelioid sarcoma, the differential diagnosis for a firm mass in the hand includes extraabdominal fibromatosis, recurrent digital fibroma, juvenile aponeurotic fibroma, fibroma of the tendon sheath and giant cell tumor of tendon sheath.<sup>4</sup> Extraabdominal fibromatosis is a rapidly infiltrating nonmetastasizing tumor with a high recurrence rate. It originates from the aponeurosis, the fascia and connective tissue of the muscle. It affects hands in 1% of cases and children younger than 10 years in 10% of cases.<sup>4</sup> In our case, the absence of nuclear positivity for ALK excluded extraabdominal fibromatosis.

All the cases of Dupuytren contracture in children previously reported in the literature appeared as either a nodule in the palm<sup>5,7</sup> or a flexion contracture of a finger.<sup>1,4–6,8</sup> In this case, the presentation was atypical with a nodule appearing in a digit on the radial aspect of the middle phalanx that was associated with gradual contracture over a period of 4 months. Later, a second nodule appeared in the adjacent digit.

Dupuytren disease should be included in the differential diagnosis of a nodule in the palm or fingers



**FIGURE 4:** **A** Presentation of a second lesion at the ulnar side of the middle phalanx of the ring finger. The lesion corresponded to the level the tip of the crooked little finger rubbed against the adjacent finger. **B** Photograph 10 days after surgery for removal of the lump from the ring finger and correction of the radial drift of the little finger with z-plasties.

or contracture of the fingers of children. Imaging studies can be misleading as in this case in which magnetic resonance imaging suggested both lesions to be fibrous histiocytoma. A tissue diagnosis is mandatory because other serious clinical entities such as epithelioid sarcoma and extraabdominal fibromatosis can present with similar clinical features.

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