Recurrent infantile digital fibromatosis

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We present a case of an 8-year-old-boy with recurrent infantile digital fibromatosis (IDF) who presented with new fibrotic lesions. IDF is a benign fibrous growth of childhood. Typically affecting the fingers and toes of children, the condition is nonmalignant and has a high recurrent potential following surgical excision. Although ~200 cases of IDF have been described to date, a generalized consensus does not seem to exist on an approach to treatment. The current treatment modalities for IDF are reviewed. ***Ann Pediatr Surg*** **10:27–29 © 2014 Annals of Pediatric Surgery.**

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Introduction

Infantile digital fibromatosis (IDF), also known as inclusion body fibromatosis, infantile digital myofibroblastoma, or Reye tumor, is a benign fibrous growth of childhood belonging to the family of fibromatoses. This was first described as a distinct entity by Reye in 1965 [1]. Typically affecting the fingers and toes of children, the condition is nonmalignant, although there is a very high rate of recurrence following surgical excision. Despite the fact that ~200 cases of IDF have been described to date [2], a generalized consensus does not seem to exist on an approach to treatment.

We present a case of an 8-year-old-boy with recurrent IDF who presented with new fibrotic lesions in the fingers and palmar region. The current treatment modalities for IDF are also reviewed.

Case report

An 8-year-old boy with recurrent right hand fibromatosis and clinodactyly presented with a new palmar lesion proximal to his small finger metacarpophalangeal joint with worsening of flexion contracture. His first presentation to our institute was at the age of 10 months, when he was referred for enlarging nodules on the dorsal aspect of his small finger's proximal interphalangeal joint. Subsequent to this presentation, he had undergone three separate fasciectomy procedures for his recurrent digital fibromatosis in his long, ring, and small fingers. In addition, he had recurrent painful lesions on the dorsum of his proximal interphalangeal joints, which were excised, and skin grafted.

The histopathological assessments indicated fibromatosis with an intermediate filament on electronic microscopy, most consistent with recurrent IDF.

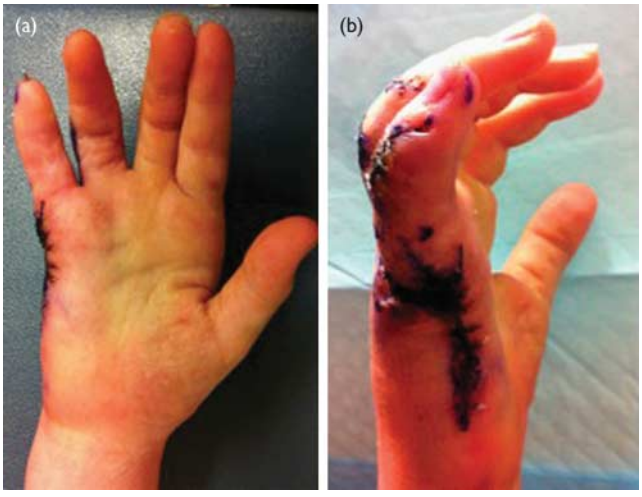
At the current presentation, he had a new 2×2 cm lesion at the hypothenar region with worsening of his flexion contracture. Fasciectomy with lesion excision and Z-plasty for lengthening was performed (Fig. 1).

Interestingly, despite compatible results in previous histopathological assessments with IDF, the palmar lesion did not indicate the presence of any intracytoplasmic inclusions, and yet, still compatible with IDF and a differential diagnosis, includes a more diffuse fibromatosis (Fig. 2).

Discussion

Fibromatoses, which represent ~9% of all soft tissue tumors in children, are a broad family of tumors ranging from completely benign to malignant. IDF represents about 2% of all of these fibromatoses, and is characterized by both its location and histological appearance [3]. The disease may present as a single or as multiple firm, pink, dome-shaped lesions that almost exclusively affect the lateral side of the distal phalanx extensor surface of the fingers and toes with sparing of the thumb and great toe. These nodules can be up to 2 cm in size and tend to

Fig. 1



Palmar (a) and ulnar (b) surface of the affected hand at 4 weeks postoperatively.

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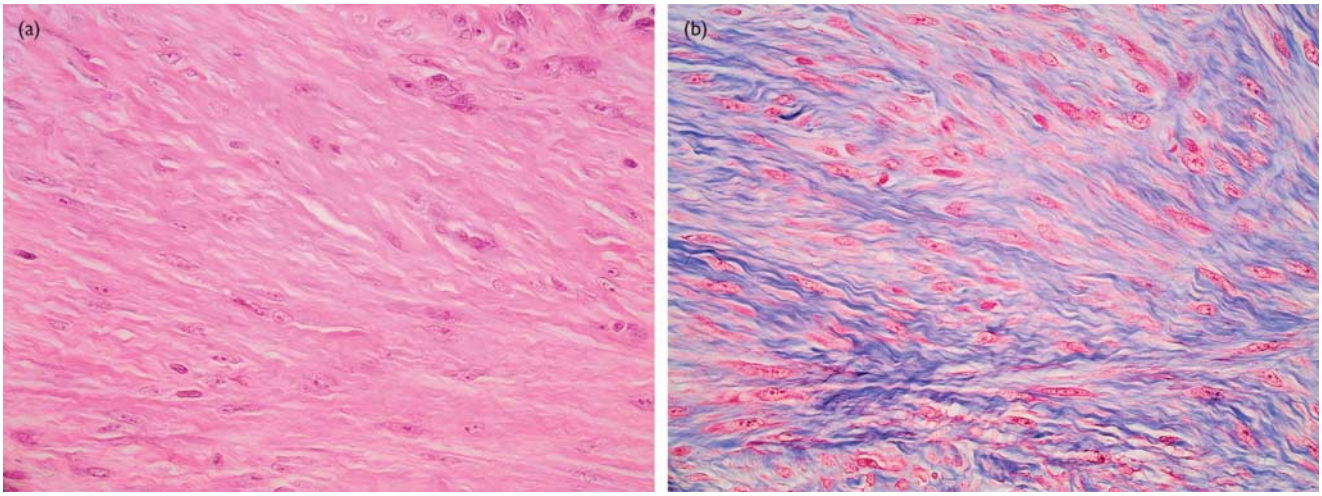
Fig. 2



Histopathological findings of most recent excised lesion indicating the absence of inclusion bodies (H&E and Masson's Trichrome stain, × 40 magnification).

be painless despite their ability to lead to joint mobility and functional deformity. Although very rare, nodules have also been noted to form in the breast, forearm, and even mouth. Most lesions appear during the first 3 years of life, and are present at birth in one third of the cases.

The etiology of IDF is largely unknown. Although a viral cause has been suggested by some authors [4], no virus has yet to be isolated. Other postulated causes include metabolic, leading to abnormal deposition of collagenous protein precursors [5], as well as congenital, although once again, no concrete evidence exists at this time [4].

Histologically, the presence of myofibroblasts with eosinophilic intracytoplasmic inclusion bodies on pathological examination is pathognomonic for IDF. There also appears to be an inverse relationship between the size and number of inclusion bodies and the degree of fibrosis, with older tumors having fewer, less dense inclusion bodies [6].

Some reports indicate that the tumor may undergo a decrease in the numbers of inclusion bodies and subsequent fibrosis with time. Thus, even as a form of fibromatosis, this entity may not have a consistently aggressive nature [6]. We believe that this may have been the case with our patient, where the most recent lesion lacked histological evidence of inclusion bodies despite having a clinical appearance that was very similar to the three previous histologically confirmed tumors 6, 7, and 8 years earlier.

The management of IDF remains a controversial issue. Although some authors recommend early surgical intervention, others advocate for a more conservation approach with hopes of spontaneous regression. When a surgical approach is adopted, wide excision and grafting remains the most commonly performed procedure [4].

Although early surgical intervention can be curative, recurrence is quite common. Baerg *et al.* [7] identified several factors that can be used to predict the rate of recurrence of fibromatoses. Patients presenting older than 5 years of age, extremity location of disease as well as incomplete surgical excision are all predisposing factors to postsurgical recurrence. Kanwar *et al.* [8] report that if the lesions are excised, usually between 2 weeks and 6 years later, there is a 75% chance of recurrence at the same site or on an adjacent finger or toe. Thus, they recommend that surgery should only be performed for correction of contractures, aggressive growth, or functional problems. Although conservative, nonsurgical management may result in regression, if this approach is chosen, it is imperative that the patient be monitored closely as joint deformity may occur.

Intralesional steroid for the management of IDF has been shown in a report by Holmes *et al.* [9] to be promising in select patients. However, further studies are required.

Other proposed treatment options include intralesional injections of fluorouracil, which, despite the pain of repeated injections, can yield good functional as well as cosmetic results [10]. Positive results have also been reported by some authors using Mohs micrographic resection [11]. Amputation is rarely required because of the benign nature of the disease and alternative treatment options.

Conclusion

In conclusion, IDF is a benign pediatric soft tissue fibromatosis with a high recurrence rate. Owing to the benign nature of the lesions and the high rate of postsurgical recurrence, we recommend a conservation watch-and-wait strategy for patients with histologically confirmed IDF nodules that do not cause joint deformity or functional impairment. One should not hesitate, however, to proceed with surgical resection of the tumor(s) if the patient develops deformity associated with the lesions. In our case, a surgical approach was chosen owing to the progressively worsening flexion contracture.

Acknowledgements

Conflicts of interest

There are no conflicts of interest.

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