

# Inflammatory myofibroblastic tumor of the hand: A case report

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**Abstract.** The present study reports the case of a 58-year-old male patient who repeatedly presented with hand ulcers that were diagnosed as cellulitis. Upon histological analysis, however, an inflammatory myofibroblastic tumor (IMT) was diagnosed. IMTs rarely occur in the hands. The involved tissue was removed with the tumor, with the exception of the tendons and cutaneous nerve. The involved tendon sheath and epineurium were carefully resected. After 2 years of follow-up, the patient showed no signs of tumor recurrence and the hand function was good. IMTs in the hands are relatively rare, and are often easily misdiagnosed as infection. An early and correct diagnosis is the key to successful treatment. A biopsy is necessary following debridement of the infected lesion, particularly for recurrent infected lesions. The surgical approach should be conservative, in order to maintain maximum hand function.

## Introduction

Over the last two decades, inflammatory myofibroblastic tumors (IMTs) have emerged from within the broad category of inflammatory pseudotumors, with distinctive clinical, pathological and molecular features (1). The etiology and pathogenesis of IMT remains unclear, however, infection, surgery, autoimmunity and chromosomal variation have all been hypothesized to contribute to IMT development (2). IMT has been hypothesized to represent an inflammatory reaction to viral infection, including human herpes virus 8 and Epstein-Barr Virus, however, it has also been considered as a type of autoimmune disease (3). Recently, IgG4-related disease has been associated with the pathogenesis of IMT (4);

however, the exact mechanism remains unclear (5,6). At the molecular level, positive immunohistochemical staining of ALK is observed in ~40-100% of IMT cases, depending on the anatomical sites at which they arise (2,7). Clinically, the majority of IMTs are benign, but they require adequate surgical treatment, as they have a tendency for local recurrence (8). IMTs exhibit a predilection for children and adolescents are considered to metastasize in ≤5% of cases (2). The most common anatomical locations are the abdominopelvic region, the lungs and the retroperitoneum (9), however, it rarely occurs in the limbs (10). Previously described cases of IMT have been described in the somatic soft tissue and bone (10). We present a case of an IMT in the hand of an adult. Written informed consent was obtained from the patient.

## Case report

In November 2009, a 58-year-old male presented to the Department of Hand Surgery at The First Affiliated Hospital of Zhejiang University (Hangzhou, China) with right dorsal hand ulcers that had been recurring for more than one year. The patient had previously undergone debridement and drainage without biopsy three times. Local clinics had diagnosed the condition as cellulitis (infection of the tissues below the skin), and found that the ulcer in the dorsal hand was refractory, with a trend for spreading. A physical examination showed a red and swollen hand and fingers, several ulcerous wounds with previous incision scars and bone exposure in the proximal phalanx of the index finger. The flexion and extension of the metacarpophalangeal joint was limited. X-ray showed no destruction of the bones and joints. Metal foreign matter was also found in the hand, which was caused by an injury 10 years ago (Fig. 1). Thus, the magnetic resonance imaging (MRI) scan was canceled. The laboratory findings were normal, except for normocytic, normochromic anemia. A fungal culture of the wound revealed the presence of *Candida albicans*.

Following one week of antifungal treatment (nystatin; 500,000 units every 6 h; YunPeng Inc., Linfen, China), debridement was performed under general anesthesia. A tumor with granulation of the vascularization in the subcutaneous tissue, which bled easily, was found. The dorsal extensor tendons, dorsal vein and cutaneous nerve were involved. The diffuse tumor grew within the superficial layer of the dorsal interosseous. No tumor capsule was noted. The involved tissue was removed with the tumor, except for the tendons and cutaneous

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Figure 1. X-ray of the right hand showing metal foreign matter in the second head of the metacarpal bone and a shadow of soft-tissue swelling.



Figure 2. Image of the tumor consisting of granulation with vascularization. The diffusely-growing tumor occurred under the skin without clear boundaries and easily hemorrhaged.

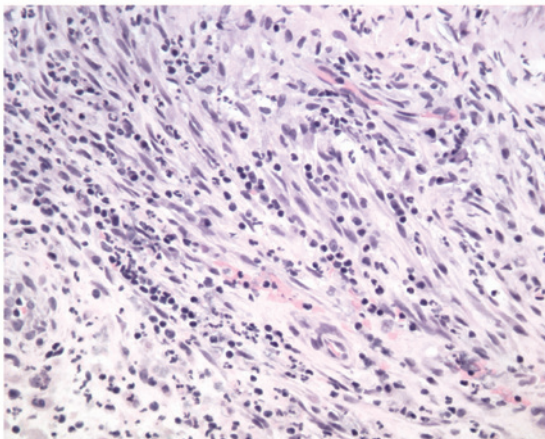


Figure 3. Spindle cells with evident cellularity and necrosis, accompanied by mononuclear cell infiltration consisting of dense lymphocytes and plasma cells. (hematoxylin and eosin stain, original magnification, x400).

nerve. The involved tendon sheath and epineurium were carefully resected (Fig. 2).

The intraoperative frozen section revealed that the tissue showed degeneration, necrosis and inflammatory granulation, with atypical hyperplasia. An extended resection of the

granulation tissue and inflammatory veins was performed. Relaxation incisions were designed in the dorsal hand to close the incisions. Histologically, the tumor was composed of spindle cells in a mesenchymal arrangement, with evident cellularity and necrosis, accompanied by mononuclear cell infiltration consisting of dense lymphocytes and plasma cells (Fig. 3). Immunohistochemical stains were positive for smooth muscle actin and desmin and negative for CD34, S100 and CD117. In addition, the spindle cells were not reactive with antibodies specific for cyclin D1, p53, B-cell lymphoma-2 or anaplastic lymphoma kinase. An IMT was consequently diagnosed.

Following one week of negative pressure draining, the relaxation incisions were closed. The patient underwent an uneventful post-operative course and was discharged. No signs of recurrence were detected on follow-up at 24 months.

## Discussion

IMTs are histologically characterized by dominant myofibroblast invasion and variable inflammatory infiltrates. The tumors behave as benign lesions, but occasionally can be locally aggressive. The literature to date shows that IMTs most typically occur in the lungs and retroperitoneum. Little focus has been placed on the localization of IMTs in the limbs and extremities (10). Currently, the etiology of IMT is unclear, however, an inflammatory origin has been suggested due to an association with minor trauma, surgery and/or infections (6,10). The causes of IMT in the present case arose from infection and debridement. Debridement without biopsy is the main cause of the misdiagnosis of IMTs. Using MRI, it is possible to evaluate the lesional extension of IMTs in the soft tissues, although this was not feasible in the present case due to the presence of metal foreign matter in the patient's hand, which was caused by an injury 10 years ago.

IMTs in the soft tissues have seldom been reported in the literature. The tumors usually displace and distort the soft tissues or bones (10). Meanwhile, IMTs in the hand have not yet been reported at all. In the present case, the tumor was found to be locally invasive. In the confined space of the dorsal hand, the dorsal extensor tendons and cutaneous nerve were involved. Thus, an enlarged resection would have destroyed the functional structure in the hand. The treatment of such IMTs can be challenging, as there is no established medical treatment protocol, and the proximity of the tumors to vital structures tumors can mean that they are not resectable. The decision to treat this lesion only by tumor resection with preservation of the extensor tendons, dorsal vein and cutaneous nerve turned out to be appropriate. A reconstructive approach after an enlarged resection would have been invasive with a less functional result, however, it may have reduced the risk of local recurrence, which is ~25% for IMTs (5).

IMTs in the hands are relatively rare and are often easily misdiagnosed as infection. An early and correct diagnosis is the key to successful treatment. A biopsy is necessary following debridement of infection lesions, particularly recurrent infected lesions. The surgical approach is should be conservative, in order to maintain maximum hand function.

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