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Atypical Presentation of Isolated Peripheral T-cell Lymphoma in the Hand: Case Report

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Abstract

Peripheral T-cell lymphoma is a rare malignancy that characteristically demonstrates generalized lymphadenopathy and extranodal involvement. We describe an atypical case of peripheral T-cell lymphoma manifesting as isolated hand swelling without evidence of nodal disease or systemic symptoms. This report emphasizes the highly variable clinical presentation of T-cell lymphomas and the importance of expeditious incisional biopsy in the setting of an undiagnosed hand mass.

Keywords

hand tumor; peripheral T-cell lymphoma; not otherwise specified

Introduction

Neoplasms arising from mature (post-thymic) T-cells are collectively known as peripheral T-cell lymphomas. This rare heterogeneous entity consists of a variety of specific lymphoid disorders that are classified by the World Health Organization using morphologic, immunophenotypic, genetic, and clinical features [1]. Peripheral T-cell lymphoma frequently presents with generalized lymphadenopathy and constitutional symptoms as well as extranodal sites of involvement that include bone marrow, spleen, and liver [2]. Cutaneous manifestation occurs in approximately 10% of patients [3], but T-cell lymphoma presenting in the hand is exceedingly unusual with only a few reports available in the literature [4-8].

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Case Report

A 78-year-old right-handed man presented with progressive and painless left hand enlargement over the course of 1 year. He denied any history of trauma or systemic symptoms such as fevers, chills, or weight loss. His only medical problems were osteoarthritis and hypothyroidism. The patient had no personal or family history of autoimmune disease. Examination revealed diffuse swelling primarily affecting the volar aspect of the hand and also the proximal portions of the fingers and distal forearm (Fig. 1). No sensory deficits were noted in the hand. The skin was edematous, and several small blisters were present dorsally. There was no tenderness or discrete mass noted on palpation. Joints were supple, and active range of motion was limited only by swelling. The patient had no discernible lymphadenopathy on physical examination. Despite his hand deformity, the patient remained an avid piano player and was especially interested in preserving hand function.

The patient had been evaluated by multiple physicians prior our hand surgery consultation. Routine laboratory studies did not indicate chronic infection and did not suggest an autoimmune etiology. Radiographs of the left hand showed no osteomyelitis or other bony abnormality. Magnetic resonance imaging (MRI) with gadolinium contrast revealed a T2 hyperintense infiltrating mass diffusely involving the soft tissues in the left palm extending from the distal forearm to the proximal phalanges. There was no definite invasion of the bone, but the flexor tendons and intrinsic muscles were clearly surrounded by the mass. Based on these findings, the differential diagnosis included rheumatoid arthritis, atypical infection, fibromatosis, infiltrative tumor, and vascular malformation.

We performed an incisional diagnostic biopsy through an extended carpal tunnel incision and did not attempt to debulk the tumor. We encountered extensive inflammatory tissue resembling synovitis (Fig. 2). The infiltrative tumor demonstrated no respect for natural barriers. The transverse carpal ligament was attenuated and the median nerve, its recurrent thenar branches, and the superficial palmar arch were markedly displaced from their expected positions. These structures were identified and separated from the tumor. The flexor digitorum superficialis and profundus tendons were intact, although these too were surrounded by tumor. A full thickness sample of skin and fat and several specimens of the lesion were taken for pathologic examination and cultures. Frozen section evaluation of the tumor favored lymphoma. The incision was closed and a soft dressing was applied. When the patient was seen 2 weeks later, the skin was healing without complications.

Pathologic examination of the specimens revealed dense inflammatory infiltrate consisting predominantly of lymphocytes permeating through skin, muscle, adipose tissue, and synovium. Immunohistochemical stains identified the lymphocytes as T-cells expressing CD4, CD3, and CD5, with loss of CD7 (Fig. 3). The final diagnosis was peripheral T-cell lymphoma, not otherwise specified. All intra-operative cultures were negative for bacterial or fungal growth.

The patient pursued oncology follow-up at another medical institution. Therefore, we ascertained the details of his postoperative course through phone interviews with the patient

and his oncologists. The patient underwent a positron emission tomography study that demonstrated isolated involvement of his right hand. A bone marrow biopsy was negative for malignancy. Work up did not reveal definitive evidence of nodal involvement. Given the patient's lack of systemic symptoms and advanced age, the tumor board recommended a course of radiation therapy to avoid the systemic side effects of chemotherapy. Shortly after completion of his radiotherapy, the patient noted a considerable reduction in the size of his hand and healing of the dorsal blisters. No immediate adverse effects from the radiation were reported. The patient stated that he was still able to play the piano.

Discussion

T-cell non-Hodgkin lymphomas comprise only 6% of all lymphoid malignancies. Mature T-cell neoplasms are known as peripheral T-cell lymphomas [9]. Within this category, there exists a subtype under the name peripheral T-cell lymphoma, not otherwise specified (or unspecified) that consists of T-cell lymphomas that cannot be classified by specific disease features. T-cell lymphoma, not otherwise specified, is usually seen in older patients presenting in advanced stage and exhibits a poor prognosis with an overall 5-year survival rate of 20-45% [3, 9, 10]. The typical clinical picture entails bulky lymphadenopathy, extranodal disease, systemic constitutional symptoms (fever, night sweats, and weight loss), and elevated lactate dehydrogenase levels [2]. From an immunophenotypic standpoint, most cases demonstrate a predominance of CD4 T-cells and downregulation of pan T-cell antigens with loss of CD7 being most common [1]. Given the relative rarity of peripheral T-cell lymphomas, there is a paucity of evidence-based recommendations that guide treatment and the optimal therapy remains controversial [2]. Among patients with cutaneous manifestations of peripheral T-cell lymphoma, those with multifocal skin lesions or concomitant disease at noncutaneous sites are generally treated with chemotherapy, whereas radiation therapy is considered first-line treatment for patients with localized disease [5, 10].

Extranodal T-cell lymphoma involving the hand is especially uncommon. A small number of case reports have been published and describe a wide array of clinical presentations (Table 1). These cases demonstrate solitary or multifocal cutaneous lesions relating to T-cell lymphoma ranging from ulcerating nodules to plaques extending over the fingers. Most of these cases exhibited associated clinical features such as lymphadenopathy, and therefore these patients were treated with chemotherapy [4, 6-8]. Sia et al [5] reported a 74-year-old man who sought evaluation for a rapidly growing (3 weeks) exophytic, 3 cm diameter, ulcerating mass on the dorsal-ulnar aspect of the right hand. The patient had a similar 4 cm lesion on the right heel. Both lesions were presumed to be squamous cell carcinoma, and thus wide local excision with 1 cm margins and immediate split-thickness skin grafting was performed. Once the diagnosis of anaplastic large cell lymphoma was established, definitive treatment via radiation therapy was pursued and resulted in remission. In contrast to these existing case reports, our patient presented with generalized hand and wrist swelling without other clinical findings. In addition, the infiltration of malignant T-cells was not limited to the skin, but instead was widely disseminated into the deeper soft tissue structures of the hand. Because our patient exhibited localized disease after extensive work up, radiation therapy was chosen.

This case report illustrates the importance of MRI and expeditious incisional biopsy in the setting of an undiagnosed hand tumor. In all cases listed in Table 1, an initial incorrect diagnosis was made based on patient history and clinical appearance. This led to a variety of misguided treatments, including administration of bexarotene [4], steroids [6], anti-mycobacterium antibiotics [7], and wide local excision [5]. Complications resulting from these attempted interventions were not provided in these case reports. Although our patient was not treated empirically for a presumed infectious or rheumatologic disorder, he underwent a battery of laboratory tests and visits with multiple physicians over the course of 1 year before an MRI was obtained. The effect on prognosis due to this delay in diagnosis cannot be determined. However, once the correct histopathologic diagnosis was attained, the patient was promptly evaluated and treated by oncology specialists.

The vast majority of hand tumors, benign or malignant, occur as solitary or multiple discrete entities and usually exhibits well-described clinical behavior. Although the differential diagnosis for our patient included atypical chronic infection and rheumatoid arthritis with severe pannus formation, there were no relevant symptoms that substantiated these conditions. Given an atypical presentation of painless generalized swelling of the hand and wrist along with non-diagnostic laboratory studies, radiographs and MRI are helpful to further investigate the characteristics of the lesion. Unless these studies provide convincing evidence of a benign process, we recommend proceeding directly to incisional biopsy. Intra-operative frozen sections may be helpful to guide subsequent immunohistochemical evaluation of specimens. Importantly, biopsy should be carried out prior to initiation of empiric therapy when the diagnosis is unclear to avoid the potential risks of unwarranted interventions and delay of definitive treatment.

Cutaneous manifestations of T-cell lymphoma can be highly variable and the diagnosis of these rare malignancies can be challenging. When patient history, physical examination, and laboratory studies exclude common causes of hand tumors, rarer diagnoses must be considered and appropriate imaging studies are indicated. Increased awareness of these established guidelines for evaluation of soft tissue masses of the hand among primary care physicians will expedite referral of patients with uncommon hand tumors. The hand surgeon can provide a diagnostic biopsy of the lesion and facilitate definitive management.

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Figure 1. Preoperative photographs. Note the ill-defined mass within the soft tissues of the left hand predominantly affecting the palm and wrist.

Figure 2. Intra-operative photograph. The tumor extended proximal to the attenuated transverse carpal ligament (*) and was intimately associated with the skin, subcutaneous tissues, intrinsic muscles, and tendons. The median nerve, recurrent branch, and superficial arch were completely surrounded by infiltrating tumor.

Figure 3.

Peripheral T-cell lymphoma, not otherwise specified. (A) Photomicrograph of biopsy specimen revealed infiltrating lymphocytes permeating between intrinsic muscle fibers (hematoxylin-eosin stain, 400×). (B) Immunohistochemical staining demonstrated CD4 positivity in the lymphocytes, indicating T-cell lineage (400×).

Table 1

Reported cases of cutaneous T-cell lymphoma in the hand.

Author (Date Published)	Age and Sex	Pathologic Diagnosis	Clinical Presentation	Initial Diagnosis and Management	Associated Clinical Features	Treatment and Outcome
Medvedofsky et al (2008)	51 M	Primary cutaneous T-cell lymphoma	Solitary ulcerating mass over left thenar eminence	Mycobacterium infection treated with empiric antibiotics	Nodular rash in axilla and groin, elevated lactate dehydrogenase level	Chemotherapy; partial remission
Fernandez-Torres et al (2009)	29 M	Extranodal NK/T-cell lymphoma, nasal type	Solitary ulcerating lesion on the right middle fingertip	Pyogenic granuloma	Axillary and inguinal lymphadenopathy, intranasal mass	Chemotherapy, no recurrence at 12 months
Park et al (2010)	52 M	Peripheral T-cell lymphoma, not otherwise specified	Multiple large nodular plaques over bilateral hands and fingers	Benign dermatologic condition unresponsive to steroids	Multiple enlarged iliac nodes, positive bone biopsy	Chemotherapy; partial remission
Wu et al (2011)	53 M	Acute T-cell leukemia/lymphoma with human T-cell lymphotropic virus type 1 infection	Multiple small nodules over bilateral upper and lower extremities	Mycosis fungoides unresponsive to treatment	Bilateral axillary lymphadenopathy	Chemotherapy; partial remission
Sia et al (2012)	74 M	Anaplastic large cell (T-cell) lymphoma	Ulcerating nodule on right hand and right heel	Squamous cell carcinoma treated with wide local excision	None	Radiation therapy to both lesions; no recurrence at 5 months
Kung et al (2014)	78 M	Peripheral T-cell lymphoma, not otherwise specified	Generalized swelling of left hand	Presumed infectious or rheumatologic etiology	None	Radiation therapy