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Case Report

Postsurgical Appearance of Chilblains Lupus Erythematosus

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Chilblain lupus erythematosus is a rare form of chronic cutaneous lupus erythematosus that presents as pruritic and painful cutaneous lesions. The lesions follow a relapsing and remitting pattern and are often located on the dorsal surfaces of the hands or feet. Its treatment is supportive in nature, and the lesions often recur. In this case report, we describe the course of a patient, with no history of any form of lupus, who developed chilblains lupus localized over the incision site after undergoing hand surgery. We discuss the differential and incorrect diagnoses made before determining the proper diagnosis of chilblains.

Chilblain lupus erythematosus is a rare form of chronic cutaneous lupus erythematosus (chilblains). Chilblain often presents with erythematous or purple, macular, or papular lesions on the dorsal surfaces of the hands or feet. Chilblains is more prevalent in colder and damper climates. The histopathology of chilblains is not highly specific, making their diagnosis difficult.[1] This case report details the unusual presentation of a 52-year-old woman in South Carolina with chilblains lupus diagnosed at the site of previous distal interphalangeal (DIP) joint arthrodesis. The intention of this report is to increase awareness among hand surgeons about a rare presentation of chilblains.

Case Report

A 52-year-old woman with a past medical history of myasthenia gravis, Raynaud disease, Hashimoto thyroiditis, and osteoarthritis presented with chronic, severe pain and a deformity in the DIP joint of the right middle finger; she had no history of trauma. The pain was worse with activity and when the joint was bumped. Nocturnal pain in the finger frequently woke her up from her sleep. Her maternal history was significant for osteoarthritis of the hands. Previous medical management included celecoxib and prednisone. After the medical management failed, DIP joint arthrodesis was indicated. Preoperative radiographs revealed DIP joint arthritis and ulnar deviation (Fig. 1). Distal interphalangeal fusion was performed under local anesthesia using a stainless steel, headless, 2-mm compression screw through a dorsal incision, without apparent complication. The incision healed without incident, and by 6 weeks after the surgery, the patient's preoperative pain and swelling resolved. Radiographic appearance at 6 weeks showed near-complete union (Fig. 2). Radiographic union was confirmed fluoroscopically 12 weeks after the surgery (Figs. 3, 4). Approximately 1 year after the surgery, the patient presented with a complaint of a progressively enlarging, firm, tender mass over the scar on the dorsum of the right middle finger. The mass was linear, in line with the previous scar, and raised by 3 mm. There was no palpable fluctuance or joint instability. Fluoroscopic evaluation in the office revealed solid DIP joint arthrodesis, with no signs of lucency or ectopic calcifications. The mass was exceedingly painful and ultimately biopsied in the office. At the time of the biopsy, the consistency of the mass was gritty, firm, and ill defined. It was incorporated into the dermis beneath the surgical scar. The biopsy included 4 mm of skin and subcutaneous tissue, obtained dorsal to the DIP arthrodesis on the right middle finger, and was sent contained in formalin to the pathology department.

The biopsy was read, showing the presence of chronic inflammation with associated foreign material and no evidence of malignancy. The pathology report stated that the foreign material was of uncertain origin and was likely present from the previous procedure or was a foreign body. Microscopic analysis showed the presence of dermal collagen, with mild fibrosis, and extensive small

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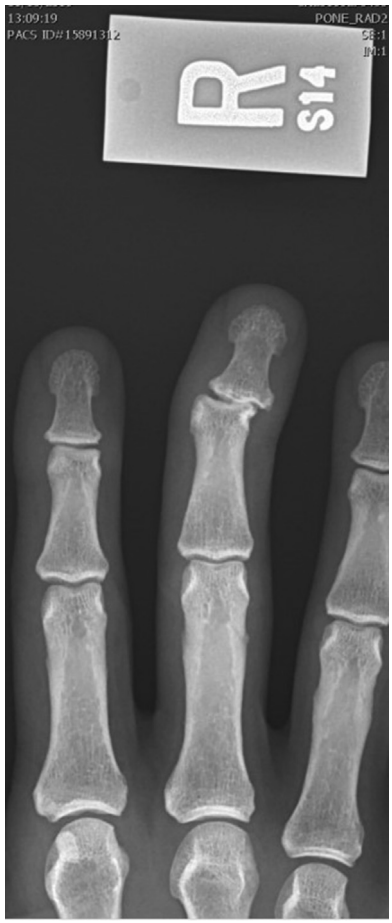

Figure 1. Preoperative radiograph of arthritic finger. Distal interphalangeal fusion was indicated and performed.

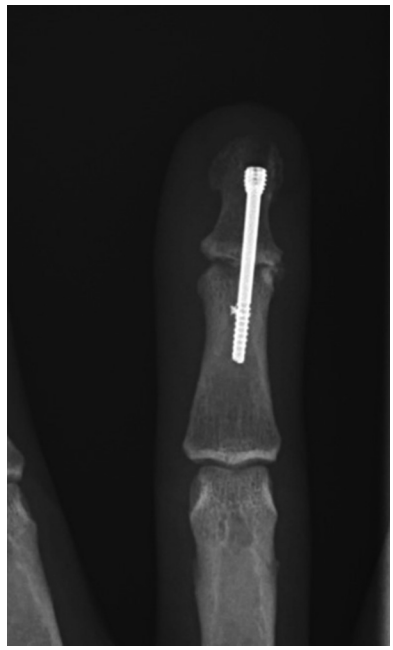

Figure 2. Radiograph of DIP fusion 6 weeks after surgery.

lymphocytic infiltrates (Fig. 5). Because the pathology report referenced possible foreign material and inflammation, the patient was evaluated for a nickel allergy to determine if she was allergic to the headless screw implant.

The composition of the compression screw was found to contain small amounts of nickel; so, the patient was sent to the dermatology department for patch testing to assess if a nickel allergy was present. The results were negative. The initial biopsy slides were evaluated again by a dermatopathologist, who reported a differential diagnosis of cutaneous T-cell lymphoma. Cutaneous T-cell lymphoma is an extremely rare form of lymphoma, and because of its unusual presentation, the dermatopathologist rebiopsied the area. The punch biopsy was 5 mm in size, 1 mm larger than the previous biopsy. The second biopsy, evaluated by a dermatopathologist, yielded the diagnosis of chilblains. After confirming the diagnosis of chilblains, the dermatopathologist recommended the application of 0.1% triamcinolone acetonide ointment topically and wearing a silicone finger sleeve at night to keep the finger warm. This treatment regimen provided symptomatic improvement 1 year after the surgery (Fig. 6).

Discussion

Chilblains are a well-described condition that often manifest as cold-induced cutaneous lesions on the acral surfaces of the body.[2] The lesions are often described as itchy or painful and most commonly occur on the ears, hands, toes, or other distal areas of the legs.[1] Chilblains can be idiopathic (primary) or secondary due to an underlying condition, most commonly systemic lupus erythematosus. A typical lesion lasts between 1 and 2 weeks, and its pathophysiology is thought to be due to vascular constriction, but the precise pathogenesis of chilblains lupus is not fully understood. Severe cases of chilblains have been reported to present with blistering and ulceration; however, this was not present in this patient.[2]

Chilblain is thought to be caused by the dysfunction of neurovascular responses to temperature changes. The resulting vasoconstriction-induced hypoxemia of distal surfaces causes a reactive inflammatory reaction. Due to the neurovascular nature of the disease, poor circulation is a predisposing factor for the development of chilblains. Some studies have shown an increased prevalence of chilblains in patients with pre-existing Raynaud disease.[3]

Several unique factors make this case an atypical presentation of chilblains. First, the patient resides in South Carolina, which has relatively mild winters, making this rare. However, the humidity of the climate likely acted as a precipitating factor. Additionally, the patient did not have lesions bilaterally, which is how chilblains usually present. The patient later admitted to prolonged postoperative icing of the operative digit early after the surgery. This prolonged icing may have incited a predisposition for developing a chilblain reaction in the operative digit. This case was also an unusual presentation, in that the papule appeared over the surgical scar of the previous DIP fusion.[4] This complicated the differential diagnosis because no other lesions were present on the other digits. The differentials included nickel allergy, infection, and nonunion. Nonunion was ruled out radiographically, and there were no clinical signs of infection, as determined using the biopsy. This case ultimately required 2 biopsies with 3 surgical pathologists' opinions. There are no reports that we are aware of at this time that have detailed hand surgery, followed by chilblains lupus isolated to the region of the surgery.

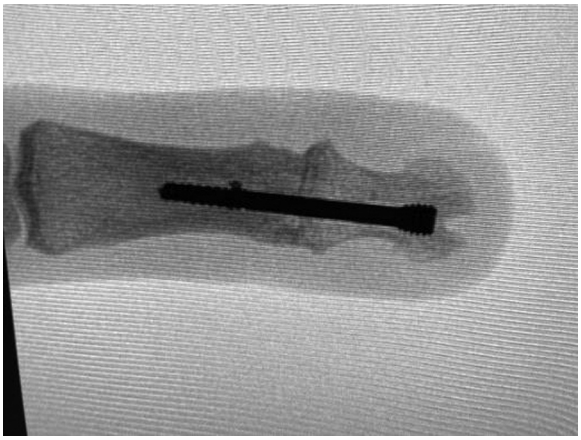

Figure 3. Anteroposterior fluoroscopic image 12 weeks after surgery.

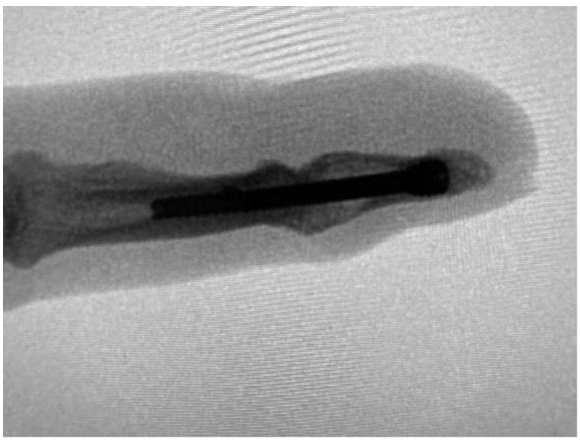

Figure 4. Lateral fluoroscopic image 12 weeks after surgery.

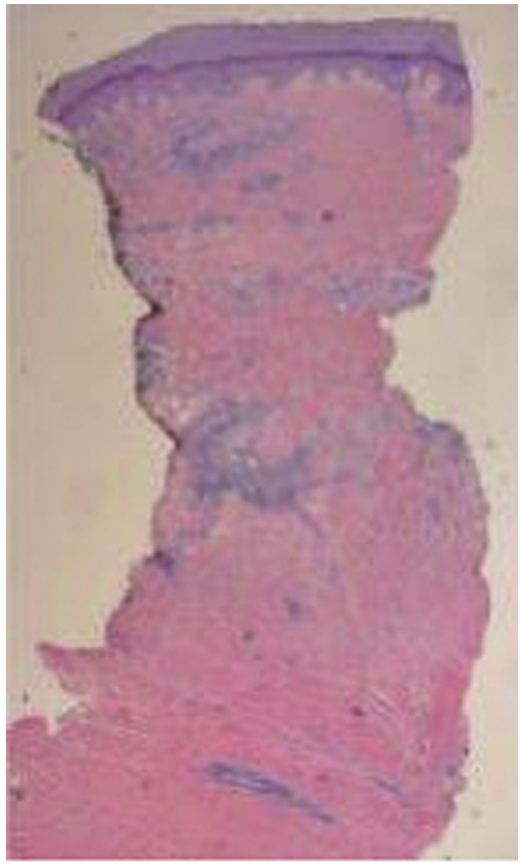

Figure 5. Hematoxylin and eosin staining of a 4-mm biopsy punch over the lesion. The microscopic description includes dermal collagen, with mild fibrosis, and extensive small lymphocytic infiltrates. Pathology reported a foreign body reaction.

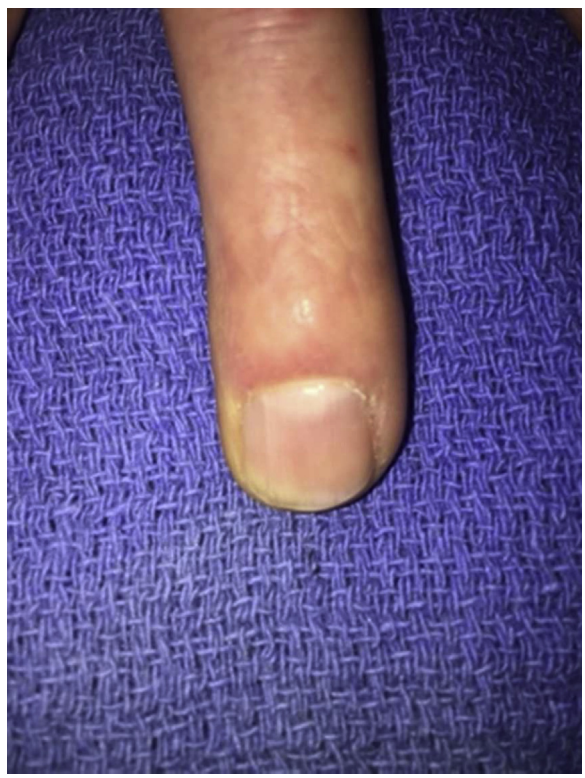

Figure 6. Patient's right middle finger 12 months after surgery.

No drug therapy was initially given to help reduce the size, pain, or itching of the lesion. There are recommended lifestyle changes that normally bring symptom relief to patients with chilblains. Topical corticosteroids, calcium-channel blockers, and vasodilators have been reported in the literature, with modest symptom relief.[5] With keeping the hands dry and warm, the patient reported reduction in the size and eventual disappearance of the lesions. Since their disappearance, the patient has reported exacerbations when exposed to cold. Figure 7 shows exacerbation after 6 hours of cold exposure to air conditioning. The patient was later started on amlodipine, which provided moderate symptom relief. Topical steroid cream and silicone compression finger sleeves also provided symptom relief during chilblain flare reactions.

It is important for hand surgeons to include chilblains lupus as a differential diagnosis for patients who present with raised, red, plaques on their hands. It is especially important for physicians who practice in humid climates to acknowledge the possibility of chilblains. Even more reason for consideration would be if the patient initially presents with these new painful lesions in the winter. Although a cold therapy system was not used in this case, it has been implicated in causing chilblains in some patients following its use during orthopedic procedures.[6] This case also emphasizes the importance of a thorough history and awareness of the seasonal onset of lesions.

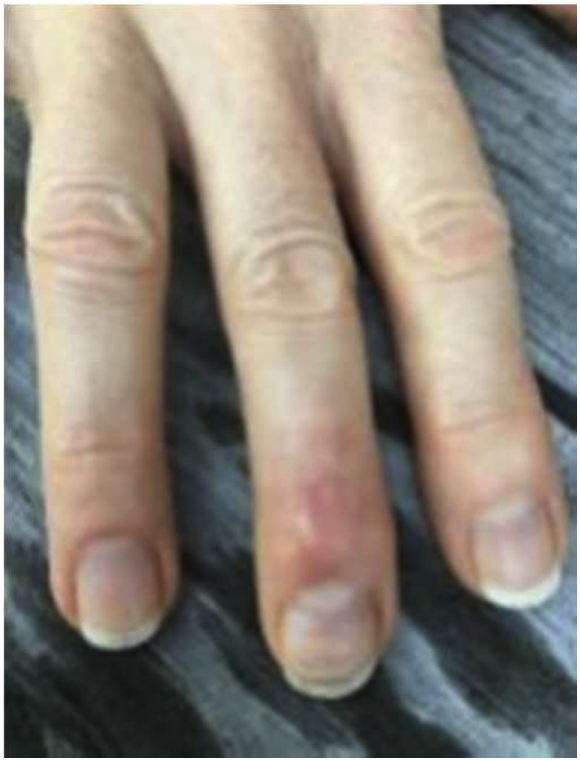

Figure 7. Cold exacerbation of 6 hours in air conditioning, resulting in reappearance of lesion.

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