Disseminated-cutaneous sporotrichosis in an immunocompetent adult

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INTRODUCTION

Classically known for causing 'rose gardeners' disease', *Sporothrix schenkii* is a dimorphic fungus ubiquitous in plant matter.[1] Sporotrichosis infection typically follows minor cutaneous trauma, causing localized lymphocutaneous (up to 95% of cases) or fixed-cutaneous (up to 30%) forms. A rare disseminated-cutaneous form occurs in up to 8%, primarily in immunodeficient patients.[1,2]

CASE REPORT

A 37-year-old woman with a history of transposition of the great arteries surgically repaired via atrial switch presented with a 2-month history of progressive, enlarging tender, erythematous nodules on the trunk, elbows, and legs (Fig 1). She first noted a single, approximately 4-cm, nodule over the posterior aspect of her right elbow. Over the subsequent weeks, similar painful nodules appeared over the medial aspect of the left elbow, abdomen, low back, and legs. She also reported progressive arthralgias involving her left wrist, elbow, knee, and ankle. She denied constitutional symptoms such as fever, weight loss, or night sweats. Skin biopsies demonstrated necrotizing and suppurative granulomatous panniculitis (Fig 2), prompting a broad workup for infectious and rheumatologic diseases. Laboratory testing was negative for tuberculosis, *Histoplasma*, *Cryptococcus*, *Bartonella*, *Brucella*, *Coxiella*, *Tropheryma whipplei*, *Blastomycosis*, and *Coccidioides*; antinuclear, antineutrophil cytoplasmic, rheumatoid factor, anti-cyclic citrullinated peptide, anti-Ro, anti-La, and anti-dsDNA antibodies; and syphilis, hepatitis C, hepatitis B, and HIV. Chest computed tomography (CT) revealed a sub-centimeter pulmonary nodule without lymphadenopathy. Universal microbial DNA testing did not detect bacterial, fungal, or mycobacterial DNA. Nine special stains for microorganisms were all negative for bacterial, fungal, and acid-fast organisms. Absolute lymphocyte count, lymphocyte subset panel, serum free light chains, and serum protein electrophoresis were normal.

After 2 months of incubation, fungal tissue culture grew *S schenckii*. A diagnosis of disseminated-cutaneous sporotrichosis was established, and oral itraconazole 200 mg twice daily was initiated, after which she reported rapid improvement of her joint pain and skin lesions. At one point during her treatment course, the patient briefly self-discontinued the itraconazole, and the lesions recurred and worsened. She was reinitiated on therapy with a plan for a prolonged course and until full resolution of lesions. The diagnosis of disseminated-cutaneous sporotrichosis raised concern for underlying immunodeficiency and/or endovascular disease, particularly given her previous cardiac surgery. Accordingly, transthoracic echocardiography and a follow-up chest CT scan were ordered. Abdominal ultrasound was also ordered to assess for signs of liver disease. Unfortunately, the

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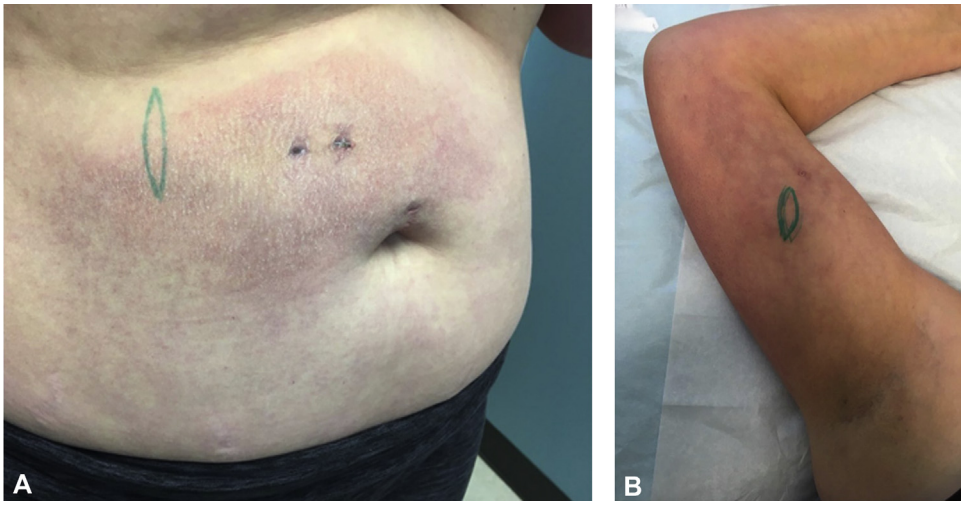

Fig 1. Disseminated-cutaneous sporotrichosis. Firm-to-hard, exquisitely tender subcutaneous nodules over the abdomen (**A**), posterolateral arms (**B**) and legs, and lower back. Some nodules were associated with overlying erythematous scaly plaques (**A**) and livedo racemosa-type skin changes (**B**).

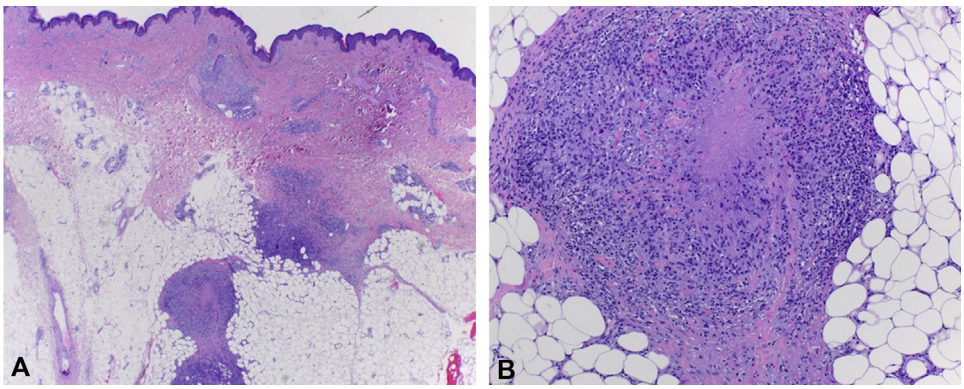

Fig 2. Granulomatous panniculitis demonstrated by hematoxylin-eosin staining. Histopathologic findings from punch biopsy at (Original magnification: **A**, ×2; **B**, ×200.) Biopsies demonstrated dermal and subcutaneous infiltrates of lymphocytes and histiocytes palisaded around zones of necrosis. Fite, Periodic acid—Schiff—diastase, and Brown-Brenn stains were negative. No detectable fungal organisms were present.

patient was unable to complete these follow-up studies during the COVID-19 pandemic.

DISCUSSION

We report a rare case of disseminated-cutaneous sporotrichosis in a patient without known immunodeficiency. Initially, a broad differential diagnosis of this nodular dermatitis was considered, including infections (bacterial, fungal, and mycobacterial), inflammatory disease (panniculitis, Sweet syndrome, granuloma annulare, sarcoidosis, vasculitis, lupus profundus), and malignancy (panniculitis-like T-cell lymphoma). The diagnosis was confirmed by fungal tissue culture.

Diagnosis of sporotrichosis is challenging due to its variable clinical presentation, the paucity of causative organisms present on biopsy, and the organism's slow-growing nature.[1] Arthralgia was reported by 53 of 178 patients in one case series, of which only 5 had radiographic evidence of arthritis.[3] Osteoarticular sporotrichosis, leading to bony destruction requiring surgical intervention, is rare.[4] Reactive arthritis in sporotrichosis is typically polyarticular, migratory, and rapidly improves with antifungal therapy, consistent with our patient's presentation.[5]

The variable morphology of lesions in disseminated-cutaneous sporotrichosis includes

ulcerated nodules with framed borders or crusted, verrucous, erythematous scaly, papulo-pustular, or infiltrative plaques.[5] Mucosal lesions occur in one-third of disseminated-cutaneous cases.[2] This polymorphic presentation is unique in comparison with the classic 'sporotrichoid' appearance in the most common lymphocutaneous form of sporotrichosis; that presentation has the typical appearance of a noduloulcerative lesion (sporotrichotic chancre) developing at the inoculation site, from which a string of similar cold nodules develop along the proximal lymphatics.[4]

Fungal culture is the gold standard for diagnosis but is time-intensive, and a minority of cases are culture-negative (5%).[3] The typical histopathologic finding of suppurative granulomatous disease is supportive but nonspecific, and *Sporothrix* infection may result in sarcoidal or tuberculoid patterns.[1] Yeast forms are visualized in only 5% to 10% of the cases, and asteroid bodies—radiating, eosinophilic structures—in 20% to 66% of cases.[1,2] Immunohistochemistry, molecular detection (PCR), sporotrichin-based skin testing, and serum antibody detection have high sensitivity and specificity but are not always commercially available.[1]

Disseminated-cutaneous sporotrichosis is typically only seen in immunodeficient patients. While this patient had a relevant exposure (gardening), it is unclear why this immunocompetent patient developed disseminated disease. Dissemination may occur after pulmonary inoculation, and while a single small pulmonary nodule was identified on this patient's chest CT, the singularity, absence of lymphadenopathy, and the patient's lack of respiratory symptoms make this less likely. Dissemination in immunocompetent hosts has been linked to cat scratches, which cause multifocal repeat inoculations.[1] This patient owns dogs, which are rarely linked to zoonotic transmission despite being a disease reservoir.[1,2] The patient is at risk for endocarditis after repair of congenital heart disease. She declined echocardiography and blood cultures, making this diagnosis difficult to exclude; however, *Sporothrix*-associated endocarditis is extremely rare, and her rapid improvement with therapy argues against this. There is evidence that cardiac surgery during infancy might impair immune function or immunologic maturation due to disruption of thymic development.[6] As this case importantly demonstrates, disseminated-cutaneous sporotrichosis can occur in patients who are not classically immunosuppressed.

Conflicts of interest

None declared.

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