



Puffy Hand Syndrome: A Case Report

Urgent Message: Incorporating questions regarding the use of intravenous drugs may help ensure the diagnosis of puffy hand syndrome is included in the differential for patients presenting with bilateral hand swelling.

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Abstract

Introduction: Bilateral hand swelling is a common presentation in a variety of conditions, particularly rheumatological disorders. Urgent care (UC) providers can avoid unnecessary testing if they are able to recognize that patients with a history of intravenous and injection drug use (IVDU) can present with this pattern of swelling—termed “puffy hand syndrome.”

Clinical Presentation: A 35-year-old female with a history of IVDU, hepatitis C, and tobacco use presented with redness and swelling of her bilateral hands for greater than 1 year. She reported some mild, generalized stiffness of her hands that got worse with heat but denied focal swelling or pain of any particular joint.

Physical Exam: The patient was afebrile. Examination of her bilateral hands demonstrated diffuse erythema when compared to her forearms without warmth and generalized non-pitting edema. There were stigmata of frequent injections scattered on her upper and lower extremities.

Case Resolution: Based on the duration of her symptoms, lack of history and findings specific for Raynaud’s syndrome, systemic lupus (SLE), scleroderma, or inflammatory arthritis, a presumptive clinical diagnosis was made of puffy hand syndrome related to her known history of IVDU.

Conclusion: Puffy hand syndrome should be suspected



in patients with bilateral non-pitting, painless edema of the hands. Including this diagnosis in the differential can mitigate emergency department (ED) referrals. Patients with suspected puffy hand syndrome can generally be counseled of the likely diagnosis and follow-up with a primary care provider (PCP) or rheumatologist to determine what, if any, confirmatory testing is indicated.

Introduction

Many conditions can present with bilateral hand swelling, particularly rheumatological disorders. The differential diagnosis for this presentation is broad and the finding can be a manifestation of a serious underlying disorder such as heart failure, liver failure, nephrotic syndrome, or infection (eg, cellulitis).¹ It can also be seen after lymph node removal in the axillary region if done bilaterally.² Consideration must also be given for

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Figure 1. Bilateral Diffuse Puffiness Without Pitting

various autoimmune and inflammatory conditions, such as rheumatoid arthritis, polymyalgia rheumatica, remitting seronegative symmetrical synovitis with pitting edema, crystal arthropathies (eg, gout or calcium pyrophosphate deposition disease), scleroderma, mixed connective tissue disease, and SLE.³ This case report describes a 35-year-old female with a history of injection and IVDU who presented with chronic bilateral hand puffiness and swelling.

Case Presentation

A 35-year old female with a history of IVDU, hepatitis C, and tobacco abuse presented with redness and swelling of her hands for greater than 1 year. She reported some mild and generalized stiffness to her hands that got worse with heat but denied swelling or pain to a particular joint. She denied recent trauma, fevers, photosensitive rashes, oral/nasal ulcerations, dry eyes or dry mouth, chest pain, triphasic discoloration to her hands or feet, digital ulcerations, and history of miscarriages.

Physical Exam Findings

The patient was afebrile, and her vitals were normal/unremarkable. She was generally well nourished and in no distress. Her mucocutaneous exam was negative for

telangiectasias, digital pitting or ulcerations, and periungual erythema, and the nailfold capillary exam was normal. Examination of her extremities of her bilateral hands was significant for diffuse erythema without warmth when compared to her forearms and the remainder of her upper extremities. Generalized non-pitting edema restricted to the hands was noted. Incidentally, there were scattered scars on her upper and lower extremities consistent with her stated history of IVDU. Her joint exam showed no focal swelling/effusions, and her range of motion of all joints was grossly normal.

The patient was referred to a rheumatologist in this case who initiated a broad screening rheumatologic work-up antinuclear antibody; ribonucleoprotein; hepatitis C antibody; rheumatoid factor; sedimentation rate; and C-reactive protein. The results of these tests were non-specific and did not suggest a clear autoimmune diagnosis. The patient declined further lab testing given the difficulty of venous access.

Based on the longevity of her symptoms, lack of history, and findings specific for Raynaud's syndrome, SLE, scleroderma, or inflammatory arthritis, a more benign process was believed to be most likely. As such, a presumptive diagnosis of puffy hand syndrome due to IVDU was made.

Discussion

Puffy hand syndrome associated with IVDU is a clinical diagnosis of exclusion based on a suggestive history and after consideration of other etiologies that may be progressive without treatment (eg, SLE, inflammatory arthritis, etc.). It typically occurs in patients with an extensive history of IVDU and is likely underdiagnosed.¹ While the differential for swollen hands is broad, the history, duration of symptoms, and clinical exam often strongly suggests the diagnosis. Incorporating questions regarding IVDU is critical for determining the likelihood of puffy hand syndrome.

Pathophysiology and Risk Factors

The suggested pathogenesis of puffy hand syndrome is that repeated trauma from venipuncture and injection of caustic substances can produce vascular and dermal sclerosis, which over time obstructs venous return.¹ It is also hypothesized that the history of injection of toxic substances can damage and impair lymphatic drainage of the hands as well.^{4,5} Repeated, subcutaneous and skin infections from non-sterile injection practices may also play a role in compromising the upper extremity's lymphatic system and impede lymphatic drainage. In patients who undergo diagnostic testing with puffy hand syndrome, lymphangiograms typically show the presence of collateralization due to destruction of deep channels of venous drainage. Additionally, skin biopsies show extensive fibrosis.⁶ In 1 study, findings of musculoskeletal ultrasound in puffy hand syndrome suggested the only expected abnormal finding to be diffuse, subcutaneous edema.⁵

Diagnosis

The diagnosis of puffy hand syndrome is made after excluding other possible etiologies such as rheumatoid arthritis, polymyalgia rheumatic, remitting seronegative symmetrical synovitis with pitting edema, crystal arthropathies such as gout or calcium pyrophosphate deposition disease, or autoimmune connective tissue diseases such as scleroderma, mixed connective tissue disease, and lupus.³

Urgent Care Management

Management involves a similar approach that is used in lymphedema treatment, including long term use of low-stretch bandages and elastic compression gloves.⁷ The patient should stop use of IV drugs permanently. Referral to physical therapy can also help with lymphedema management.¹ Puffy hand syndrome can persist even after a person stops injecting drugs. One case

described a patient with IVDU who stopped drug use and 1 year later developed his initial episode. Three years later, he developed his second episode.⁸

Case Resolution

In this case, the rheumatologist believed that given the unremarkable and non-specific rheumatologic screening work-up, a diagnosis of puffy hand syndrome was most likely.

Ethics Statement

The patient provided verbal consent for publication of this case.

Takeaway Points

- An underappreciated complication of IVDU is puffy hand syndrome. It should be suspected in patients with bilateral non pitting, painless edema with substantial history of IVDU.
- Incorporating questions regarding the use of intravenous drugs may help to ensure the diagnosis is included in the differential.
- Recognition of this syndrome can prevent patients from undergoing unnecessary interventions and treatments.
- Treatment of puffy hand syndrome involves use of low stretch bandages and elastic compression gloves, as well as counseling and support to prevent ongoing IVDU. ■

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