Clinical vignette: Puffy hand syndrome: a complication of arterial injection of illicit drugs resulting in chronic limb edema

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Puffy Hand Syndrome: A Complication of Arterial Injection of Illicit Drugs Resulting in Chronic Limb Edema

CASE PRESENTATION

A 39 year old male with history of hepatitis C, rheumatoid arthritis and history of endocarditis presented with 3 days of bilateral hand swelling, pain and tingling after injecting heroin and methamphetamine into both his radial arteries. His hands felt tight and he had difficulty moving his hands. His only other symptom was feeling chilled. He denied fever. The patient reported using heroin, methamphetamine and cocaine with last use 1 day prior to admission. Vital Signs on admission: afibrile, blood pressure of 107/56, heart rate 87, respiratory rate 16. Physical exam revealed a male in no acute distress. Pertinent findings included bilateral swelling distal to the wrists with erythema of the knuckles and no cyanosis of the digits. Swelling was greater on the right. There were no areas of fluctuance appreciated. Laboratory data demonstrated no leukocytosis. ESR was 22 and CRP was 1.1. Urine Drug Screen was positive for opiates. Bilateral Hand X Ray had findings of soft tissue swelling greatest in lateral aspect.

HOSPITAL COURSE

A diagnosis of puffy hand syndrome was made. This was the most likely diagnosis given lack of leukocytosis, systemic symptoms and minimally elevated inflammatory markers. The patient's history and exam was also consistent with findings described in the literature.

Initial treatment for his condition included management of lymphedema with warm compress and elevation. Given his presentation and lack of signs of infection, we did not feel that this was cellulitis. However, the consequences of not addressing a possible deep or fascial space infection could result in severe complications requiring surgical debridement.1 Also, we did not have the capability to perform a lymphangiogram. He was treated with IV antibiotics in the hospital and was discharged with oral antibiotics for a 10 day duration. Patient's blood cultures drawn on admission remained negative.

Of note, he returned to the ED a week later with a similar presentation after arterial heroin injection and was discharged from the ED without admission or medications. Months later he was admitted for an abscess to his wrist requiring surgical debridement, discharged, and was subsequently lost to follow-up.

DISCUSSION

Puffy Hand Syndrome was first described in a letter to the New England Journal of Medicine in 1965, “Puffy-Hand Sign of Drug Addiction” was noted by a physician performing physical exams on prison inmates.1 He noted that in this non-infectious phenomenon, which resulted in a lymphedema similar to that seen after mastectomy, veins and tendons on the dorsum of both hands could not be seen.1 He also noted that the skin was thin and smooth, without a pitting quality.2 The changes are typically found bilaterally and physical exam also shows signs of intravenous drug use.3 A study performed at methadone substitution centers found that risk factors for puffy hand syndrome are injection into the hands, female gender, and non-use of a tourniquet.4 Buprenorphine injection was not found to be associated with development of puffy hand syndrome.5 Past local infections (e.g. abscess, cellulitis) may have contributed, but determining a temporal relationship was difficult.2

The pathophysiology was later described by Nevisier in study of intravenous drug users through biopsies of skin and subcutaneous tissue, venograms and lymphangiograms, to be destruction of the lymphatics leading to fibrosis of the subcutaneous tissue.3,6 This is likely due to soluble compounds in heroin that damage lymphatic vessels such as quinine.1

Lymphangiography is performed by subcutaneous patent blue dye injection in the dorsum of the hand.6 If the dye is absorbed by the lymphatics, the lymphatic vessels can be cannulated for injection of contrast dye to define the lymphatic system anatomy.7 A normal study shows deep lymphatic outlines. In Puffy Hand Syndrome, only superficial lymphatic vessels are identified with extensive collateralization.4

The diagnosis can be made clinically by exam findings of non-pitting edema from the proximal fingers to the wrist that is unaffected by elevation.2 There is also an inability to see the dorsal veins and extensor tendons.8 This case highlights bilateral upper extremity edema of a non-infectious cause due to lymphatic sclerosis after injection of intravenous drugs. This is a chronic condition that is treated by cessation of intravenous drug use and lymphedema specialists. The chronic edema may not improve after cessation of intravenous drug use. Although this is not a primary infectious process it has been recommended to treat cellulitis to prevent a fascial space infection.9

Risk Factors for Puffy Hand Syndrome:

- Female gender
- Injection Drug Use
- Dorsal Hand Vein Injection
- Lack of Tourniquet Use

Physical Exam Findings

- Non-pitting edema from proximal fingers to wrist
- Obscured dorsal veins and extensor tendons
- Skin is thin and smooth
- Pathophysiology
  - Lymphatic destruction from intravenous drug use
  - Fibrosis of subcutaneous tissue

