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Case Report

Lower limb lymphedema mimicking as elephantiasis in intravenous drug user

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ABSTRACT

India been the population of over a billion, it is estimated that 62.5 million people use alcohol, 8.75 million use cannabis, two million use opiates, and 0.6 million use sedatives or hypnotics. Intravenous (IV) drug abuse is fairly spreading throughout the country. Rapid urbanization and migration to metro cities have led to disturbance in the social life render an individual more vulnerable to stresses and strain of modern life and with easy availability of drugs, sharing needle, peer pressure are also among the commonest reason for exponential rise of intravenous drug user (IVDU) especially among the young generation. IV drug abuse is one of most serious and social evil, prevailing in our society that leads to number of infections from local cutaneous infections to dreadful like human immuno-deficiency virus (HIV) which in turn not only increase the morbidity but mortality as well. Here we have patient who had history of IV drug injection to lower limb that led to chronic lower limb swelling which on further investigation turns out to be grade IV lymphedema.

Keywords: Unilateral lymphedema, IV drug abuse, Elephantiasis

INTRODUCTION

Drug abuse is one of most dangerous emerging health issue around globe and almost every country is struck by this. India too is caught in this vicious circle and number of drugs addicted have been increasing day by day. Changing cultural value, increasing socio-economic stress and peer pressure is one of the most common trigger factors among young generation. Official figure released by United Nations (UN) report in 2009 calculate around one million heroin addicts in India and unofficially figure may be as high as five million.[1,2]

There are various type of systemic infection and cutaneous infection which is related to iv drug abuse. Systemic infections include human immuno-deficiency virus (HIV), hepatitis C virus (HCV), hepatitis B virus (HBV), infective endocarditis (IE), community-acquired pneumonia (CAP) including aspiration pneumonia, and a greater incidence of pulmonary tuberculosis compared with the general population, septic pulmonary emboli can seed from injection-site infections and tricuspid valve endocarditis and cutaneous includes ulcer, granulomatous dermatitis, vasculitis, pyoderma gangrenosum and hyperpigmentation at injection sites. Various other, manifestation includes pruritus, sooting tattoos, atrophic scars secondary to skin popping, skin ulcers, puffy hand syndrome and limb odema.[3-5]

Here we had patient that present with unilateral lower limb edema with history of iv drug injection in lower limb that later diagnosed with lymphedema. Lymphedema is defined as localized form of tissue swelling resulting from impaired lymphatic drainage and excessive retention of lymphatic fluid in the interstitial compartment. It is of two types primary and secondary. Defect in developmental

lymphatic vascular anomalies leads to primary lymphedema secondary lymphedema is acquired and arises as a result of an underlying systemic disease, trauma, or surgery.[6]

In its initially stage, lymphedema often remain undifferentiated from general edema and frequently missed, routine therapy with diuretic often unable to resolve the swelling. During initially stages skin is soft, pitting is clear and limb elevation may give a temporary relief to patient but as it progresses swelling is replaced by fibrosis of soft tissue and later skin harden pitting become ceases.[10]

Primary lymphedema is marked by hyperplasia, hypoplasia or aplasia of lymphatic vessels, various other condition that leads to development abnormality of lymphatics includes Klippel Trenaunay, Weber syndrome, and Turner syndrome.[7,8]

Secondary lymphedema develops due to damage or dysfunction of the normally functioning lymphatic system such as various oncological surgical procedures as axillary node dissection in breast cancer, radiation, parasite infection like filariasis is one the leading cause of lymphedema.[9]

There are various pressures at interstitium and capillary levels have been associated with occurrence of lymphedema, these includes capillary pressure, negative interstitial pressure, interstitial fluid colloid osmotic pressure, and plasma colloid osmotic pressure. Variation in any of the four can lead to lymphedema. Additionally, alteration of either extrinsic or intrinsic propulsion mechanisms (e.g., fibrosis impeding muscle movement) or of lymphatic structures (e.g., radiation-fibrosed nodes or absent nodes and vessels) can impede lymph flow, compromise fluid egress, and result in lymphedema.[11]

Lymphedema can be divided in four stages- stage 0: patient is at risk of developing lymphedema e.g. patient with ca breast; stage 1 (spontaneous): reversible stage and had pitting edema and swelling often respond to elevation of limb; stage 2 (spontaneously irreversible): had underlying fibrosis and swelling don't respond to elevation; and stage 3 (lymphostatic elephantiasis): during this stage, papillomas may form, infections/cellulitis may occur, and the skin becomes dry.[12]

CASE REPORT

A 56-year-old male shopkeeper by occupation and reformed IV drug user presented to GGSMCH, Faridkot outpatient department (OPD) with unilateral right lower limb edema and overlying skin changes from last twenty years which was insidious in onset and progressive, initially edema involved only foot and ankle, from last 10 days swelling progresses and extends to thigh and scrotum. he used to inject drug in the foot 20 years ago. Edema was pitting in nature and with over hyperpigmentation of overlying skin. There was no history of pain, redness, fever, itching. No history of trauma, no history of breathlessness and abdominal distension.

Patient had past history of left lower limb above knee amputation 10 years ago post trauma. He was hepatitis C positive from last 10 years but not took treatment.

Patient was calm, conscious oriented to time, place and person, his vitals on presentation are blood pressure (BP) 110/70 mmHg, pulse rate 78/min, respiratory rate 18/min, saturation 98% on room air and afebrile. On general physical examination pallor and clubbing present. Respiratory and cardiovascular was normal on examination. On per abdomen examination liver was palpable with irregular surface and span of 16 cm and shifting dullness present.

On local examination of right lower limb pitting edema was present extending from foot up to scrotum and overlying skin hyperpigmentation was noted. there was no tenderness, warmth and erythema.

Routine investigations were done total leucocyte count (TLC)-wnl, haemoglobin (Hb)- 9.2 g/dl, platelet count-30000, TSP-7.5 g/dl, albumin-2.6 g/dl rest was with in normal limits. His APRI score was 3.0 (severe fibrosis) and Fib4 -11.3 (advanced fibrosis).

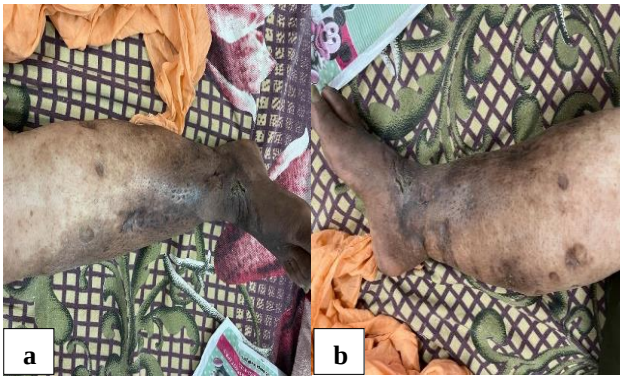

Figure 1 (a-b): Lymphedema with hyperpigmentation.

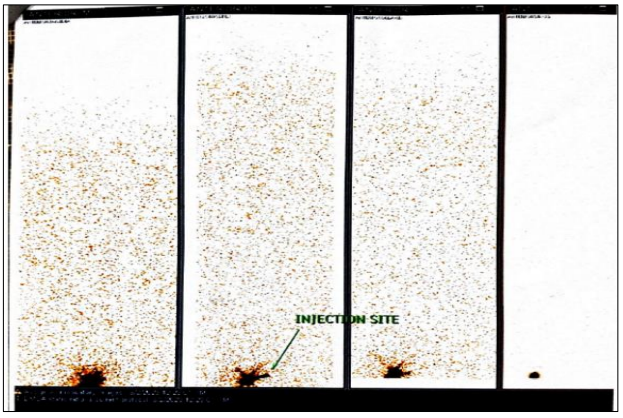

Figure 2: Lymphoscintigraphy of lower limb.

Ultrasound whole abdomen done suggestive of hepatomegaly with ascites. Liver elastography was suggestive of cirrhosis. Upper GI endoscopy was grading I and II single column varices and portal hypertensive gastropathy. On right lower limb venous there was no features of deep venous thrombosis (DVT) and arterial doppler was also normal. Right lower limb lymphoscintigraphy was done suggestive of grade IV lymphedema.

Patient was managed conservatively with iv antibiotics and diuretics and with limb elevation and scrotal support.

DISCUSSION

Lower extremity edema is frequently encountered in clinical practice. The challenge is to correctly identify the etiology of edema, and hence correctly manage the cause. In most of the case it presents as bilateral pedal edema while unilateral edema is rarely encountered in clinical practice and needs evaluation to look for the cause. In most of the case unilateral swelling is caused by unilateral DVT, ruptured baker's cyst, ruptured leg muscle, compartment syndrome, intramuscular hematoma, infection, superficial vein thrombosis, mass/tumor, fracture and in most of the chronic case it includes primary venous disease, post-thrombotic syndrome, iliac vein compression, lymphedema, vascular malformation, static foot disorders, radiation, hypertrophy.[13]

Unilateral lower extremity edema below the knee commonly results from deep venous thrombosis, venous insufficiency, or lymphedema. Lower extremity edema may have various causes. Heart failure, liver cirrhosis, nephrotic syndrome, and hypothyroidism are among the more common causes of bilateral lower extremity edema. Unilateral lower extremity edema below the knee commonly results from DVT, venous insufficiency, or lymphedema, although one third of lymphedema cases may also be unilateral.[18]

Generally, the leg edema is of two types: venous edema and lymphedema. In venous edema there is excessive protein-poor interstitial fluid from increased capillary pressure that can't be accommodated by lymphatic vessels while excessive protein-rich fluid within skin hinders the normal functioning of lymphatics. A third type, lipidemia, is more accurately considered a form of fat maldistribution rather than true edema. Puffy hand syndrome can present during or after bouts of intravenous drug abuse with intermittent non-pitting edema and similarly edema occur in lower limb that may be unilateral or bilateral depending upon the site of injection.[14,15]

Foot involvement is less common yet remains possible if injections are frequently administered in the surrounding area of vein. Both venous and lymphatic involvement is speculated. Often the continuous use of iv drug injection leads to sclerosis of underlying subcutaneous tissue leads to lymphatic blockage and other related complication. Lymphatic destruction and obstruction have also been theorized as modes of pathogenesis for puffy hand syndrome.[15,16]

There are various local complications that occur at the site or in the area of injection. That includes acute complications occurring within a few hours to 48–72 hours after injection and delayed complications. Acute complications include, Injection marks, cutaneous infections, necrotizing fasciitis, necrotizing ulcers. Thrombophlebitis and delayed includes hyperpigmentation, shooting tattoos, chronic venous insufficiency and ulcers. Cutaneous nodules, panniculitis, sclerosis and ulcers and lymphedema.[17]

It is really a diagnostic challenge for physician and often for diagnosis we start with appropriate laboratory investigation such as complete blood count, liver function test (LFT), renal function test (RFT) urine analysis thyroid profile, serum albumin. D-dimer level may give the initially hints regarding DVT though duplex ultrasound (DUS) is the first investigation of choice in case of swollen lower limbs without a clear cause. With sensitivity and specificity rate >90% for DVT, venous reflux, and non-vascular etiologies, swelling can be readily and robustly evaluated.[13,19-21]

With invasive procedure like venography with intravascular ultrasound (IVUS) is not only useful for assessment but also help in guiding for endovascular treatment of abdominal/pelvis venous etiologies of lower limb edema. Although this is an invasive procedure lesion within venous system such stenosis, compression can be assessed with sensitivity >85%.[21]

Although lymphedema is usually a clinical diagnosis, imaging techniques can be used for confirmation or planning for surgical intervention. Direct contrast lymphangiography has largely been abandoned for the evaluation of a swollen extremity due to its technical complexity and complications. Currently, lymphoscintigraphy is the gold standard testing that use Tc-99m radiolabeled sulphur colloid which is injected in the intra-dermally in the second and third web spaces of the feet with serial imaging using a scintigraphic camera for the assessment of lymph movement from the feet to the chest.[22,23]

Treatment can be divided in to intensive phase, maintenance phase and decongestive therapy. The intensive phase, which aims to reduce swelling and normalize the tissue pressure, consists of education and skin care, MLD, multilayer short stretch bandages, and lymphedema exercises. After the volume of the limb is stabilized, the patient enters the maintenance phase in which she/he continues to skin care, self-drainage, compression garments, and exercises. Complete decongestive therapy should be performed by educated and mastered lymphedema therapists or lymphedema specialists.[24,25]

CONCLUSION

Easy accessibility of iv drugs and regular injection practice are likely to cause number of infections and one of them is lymphedema that in turn increase the morbidity and mortality which increases the burden on health system. So it is a high time to start the educational campaigns that should be directed toward patients who struggle with intravenous drug use. Although infections in IDUs can be challenging to manage, they can be satisfying to look after with the right approach.

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