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A rare case of post traumatic charcot neuroarthropathy with lymphostatic elephantiasis

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Abstract

Introduction: Charcot neuroarthropathy is a chronic and progressive condition affecting the bones, joints, and soft tissues, mostly associated with diabetes mellitus. Lymphedema, at its utmost stage, called lymphostatic elephantiasis, is irreversible and associated with very large limbs. In developed countries, malignancy and the consequences of its treatment represent the main cause of lymphedema.

Patients and methods: We present the case of a 43 year-old man who suffered a severe road traffic accident, with disrupted left hemi-pelvis with lumbosacral plexus lesion. Thirteen years after he exhibited an unique association of post traumatic severe Charcot neuroarthropathy with lymphostatic elephantiasis.

Results: After a multidisciplinary team approach and following patient's desire, he was submitted to a below-knee amputation.

Conclusions: Charcot neuroarthropathy with lymphostatic elephantiasis poses a great challenge to the surgeon. The stasis signs of the distal leg skin precluded filleting for stump closure, leading us to graft it with healthy skin. Retrospectively, grafting revealed a bad option. Five month later and under a customized compression garment, the patient has not achieved complete epithelialization, with continuous lymphorrhea through the mesh spaces. The patient is waiting for a gastric bypass and after will be probably a candidate for a lymphedema physiological procedure.

Keywords

charcot; foot; neuroarthropathy; lymphedema; elefantiasis

Background

Charcot Neuroarthropathy (CN) is a chronic and progressive condition affecting the bones, joints, and soft tissues, characterized by joint subluxations, dislocations, bone loss, pathologic fractures, fixed deformities, and ulceration. Although CN can occur at any joint, foot and ankle weight bearing joints are most commonly affected. The hallmark deformity is midfoot collapse, known as the “rocker-bottom” foot [1].

Eventhough pathophysiology remains unknown, two main theories exist and they present

peripheral neuropathy as an underlying triggering condition. The neurotraumatic theory states that CN is caused by unperceived and repetitive trauma to an insensate foot. Under these circumstances, the bone and soft tissues respond with an unregulated inflammatory process which enhances osteoclast activity and excessive bone turnover. The neurovascular theory states that CN occur secondary to a hypervascular state that exists due to vaso-autonomic neuropathy. The increased blood flow enhances capillary leakage leading to increased compartimental pressure and tissue isquemia. Moreover, it causes the minerals to be washed off and also stimulates the osteoclasts. In fact, resting blood flow in patients with diabetic CN may be five times normal values and patients with diabetes mellitus and peripheral arterial disease have lower chances of developing CN [2,3]. CN is usually associated with diabetes mellitus, with prevalence ranging from 0.4 to 13% or even more, as this condition often goes undiagnosed. However, any condition that causes sensory or autonomic neuropathy can lead to this disease [4].

Lymphedema, at its utmost stage, called lymphostatic elephantiasis, is irreversible and associated with very large limbs. The most common cause of secondary lymphedema worldwide is filariasis. In developed countries, malignancy and the consequences of its treatment represent the main cause. Obstruction can arise from primary or metastatic cancer, lymph node dissection or radiotherapy induced scarring. Obesity, trauma and congestive heart failure are other important etiologies [5].

Case Presentation

Fourty-three-year-old man who suffered a severe road traffic accident thirteen years before in the United Kingdom, with disrupted left hemi-pelvis with lumbosacral plexus lesion, ruptured bladder, and fractured right femur, tibia and fibula. Later developed left lower limb deep venous thrombosis, progressive bilateral edema more pronounced on the left, left foot and leg ulcerations, left foot deformity and recurrent episodes of erysipela under prophylactic penicillin.

Physical Examination: On the left with significant enlargement of the leg and foot, drop and inverted foot with mid and hind foot deformity and collapse of the longitudinal and transverse arches, scars of previous ulcers and grafting, and digital and interdigital ulceration; erythematous and thickened skin with cobble-stoned and hyperkeratotic areas; on palpation with nonpitting edema with below-knee anesthesia (except ankle).

On the right with lower limb shortening due to multiple fractures; edema of the leg with erythematous and thickened skin *peau d'orange* like (figure 1 and 2).

Complementary exams

Electromyography (only left lower limb) – denervation potential at rest of the posterior leg compartment; anterior compartment without muscle resistance; external compartment not studied for lack of patient collaboration.

Lymphoscintigraphy – pronounced bilateral delay of lymphatic progression in the lower limbs, with an obstruction in the metatarsal region on the left and superficial collateral circulation.

X-ray & MRI – pronounced structural and anatomical relationship changes of the left tarsal bones, particularly the hindfoot, with osteolysis; marked edema of subcutaneous tissue and muscles with signs

Venous ultrasound – left deep venous thrombosis completely recanalized; normal permeability and valvular competence of the remaining deep and superficial venous system bilaterally.

After a multidisciplinary team approach and following patient's desire, he was submitted to a below-knee amputation and skin grafting of the stump.

Discussion

This patient shows post traumatic stage III lymphedema (or lymphostatic elephantiasis - International Society of Lymphology classification) with pronounced Charcot neuroarthropathy (stage 1 and type 2-3 according to Eichenholtz and Brodsky classification respectively) [6]. This is a very rare association, also described in Fabry disease, for instance [7].

The patient was under antidepressants and had class II obesity due to the extreme lower limb weight and the severe impairment of daily activities and consequent sedentary lifestyle.

Charcot neuroarthropathy with lymphostatic elephantiasis poses a great challenge to the surgeon. Surgery in CN is usually reserved for patients in whom conservative treatment has failed, for treating complications such as deformity, joint instability, fractures, infection, and ulceration or high risk to ulceration associated with bony deformities or contractures. Charcot-related ulceration significantly increases amputation risk and so every attempt should be made in order to prevent this complication. Exostectomy of bony prominence, osteotomy, arthrodesis and Achilles tendon lengthening are some of the advocated procedures [6,8]. In this setting, the extensive distortion prevented any of these limb salvage procedures.

Neuropathy, primarily caused by the plexus lesion, was also worsened by the extreme lymphedema, which contributed to the reduction of foot sensitivity and consequent unperceived and repetitive trauma. Stage III lymphedema is ameanable to ablative operations, such as the Charles procedure, fraught of wound-healing problems and recurrence, or physiological procedures, such as vascularized lymph node transfer [9]. At this stage, lymphovenous anastomosis is not feasible due to the deterioration of the lymph capillaries [10]. Again, debulking an insensate, distorted and drop foot did not seem reasonable, with no chances of improving patient's quality of life status. Vascularized lymph node transfer before weight loss would limit the potential of this procedure. He underwent a below-knee amputation, with an expected good ambulatory outcome (figure 4). The stasis signs of the distal leg skin precluded filleting for stump closure, leading us to graft it with healthy skin. Retrospectively, grafting a stump in a limb with pronounced delay of lymphatic progression revealed a bad option. Five month later and under a customized compression garment, the patient has not achieved complete epithelialization, with continuous lymphorrhea through the mesh spaces. Posterior leg skin, which was not ulcerated, should have been used. Despite strictly adhering to the recommendations of the dietitian, the patient is unable to lose weight, certainly because of the difficulty in practicing physical exercise. Consequently, he is now waiting to undergo a gastric bypass. Being one of the etiologies of lymphedema, the resolution of obesity will probably improve the lymphatic drainage of the lower limbs. After weight stabilization the patient will certainly be a candidate for a lymphedema physiological procedure, such as vascularized submental lymph node transfer to the groin or to the stump.

Tables

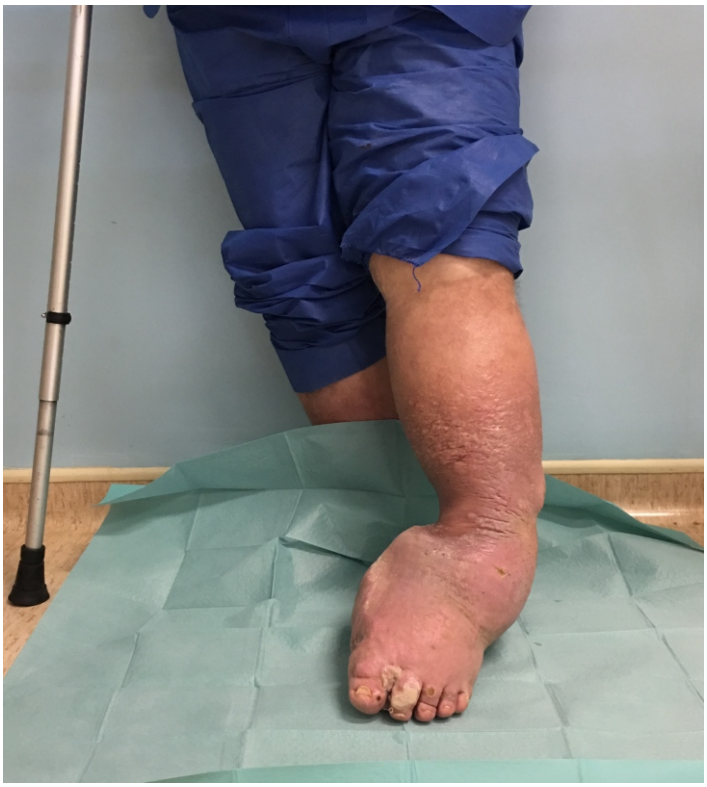

Figure 1

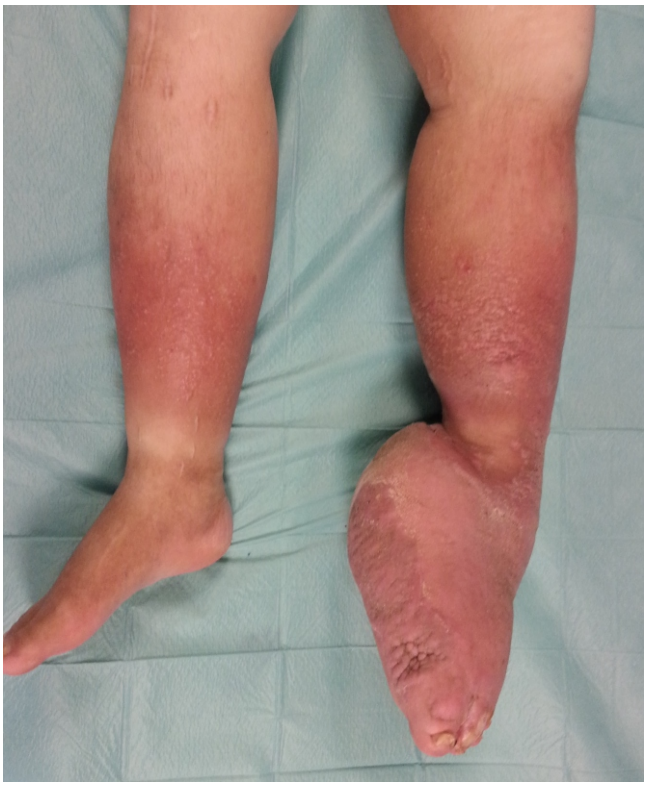

Figure 2

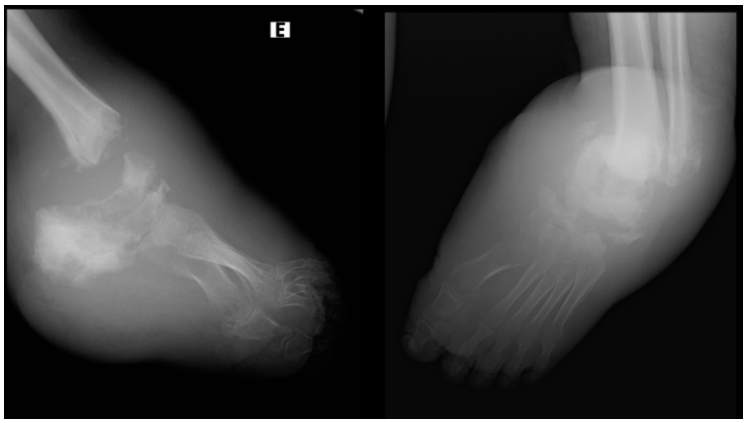

Figure 3

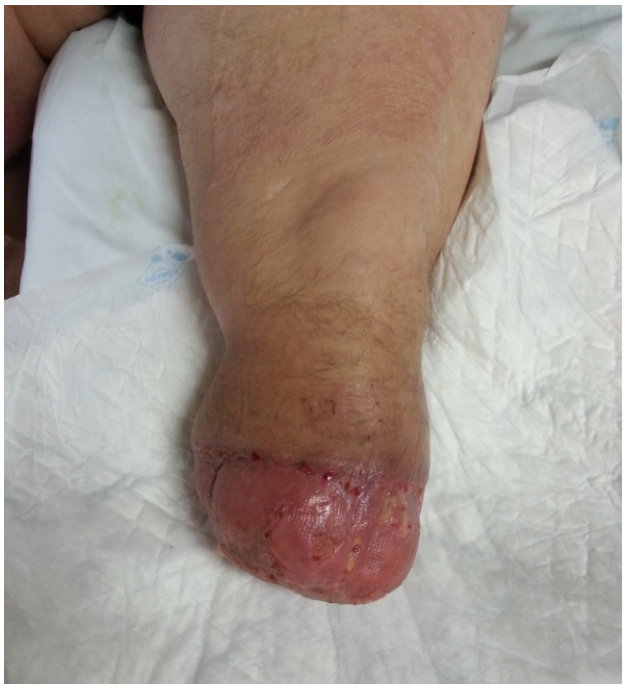

Figure 4

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