Massive Unilateral Lower Limb Lymphedema in a 42 Year Old Brazilian Man

Vitorino Modesto dos SANTOS1, Lister Arruda Modesto dos SANTOS2, Antônio Augusto Dall’Agnol MODESTO3, Milena de Oliveira AMUI4

1Catholic University (UCB) and Armed Forces Hospital (HFA), Internal Medicine, Brasília-DF, Brazil
2State Workers Hospital (HSE), Surgery, São Paulo-SP, Brazil
3Community Medicine, Family and Community Medicine, São Paulo-SP, Brazil
4Uniube, Internal Medicine, Uberaba-MG, Brazil

This 42-year-old Brazilian man of low socioeconomic status came for a clinical evaluation because of malaise, fever (39°C) and shivering associated with pain in the left lower limb. He stated that Fifteen years ago he underwent surgical procedures on the left thigh and leg, which supposedly gave origin to repeated episodes of dermatolymphangioadenitis (DLA) for about ten years. Benzathine penicillin (1.200.000 units at 5-week intervals) contributed to control the frequency of DLA. Relevant epidemiological data included a previous period living with bare feet in Brazilian forest areas, and in other South American regions. His father worked as a collector of latex from the Hevea brasiliensis tree and suffered chronic edema of the legs, allegedly due to tropical filariasis. The patient had no previous cancer or venous thrombosis, nor cardiac, hepatic, renal, or thyroidal disorders. Physical examination revealed an accentuated lymphedema with circumferential measures many times greater in the left lower limb as compared with the uninvolved right extremity (Figure 1A). Testicles and scrotum were normal, and mildly enlarged elastic lymph nodes were palpated in the left groin. The skin was thickened and with a peau d’orange appearance on the calf (Figure 1B). An extensive surgical scar, as well as two very deep skin folds were evident on the affected extremity. It was worthy of note that cobblestone papules, hyperkeratosis, lichenification, mossy and warty changes were absent. The patient was not obese, and the results of laboratory routine tests were unremarkable. Investigation concerning the differential diagnoses of unilateral elephantiasis affecting his lower extremity included thrombophilic tendency, microbiology, histopathology, and serologic studies.

What is your diagnosis?

A. Elephantiasis nostras verrucosa
B. Lymphatic filariasis
C. Podoconiosis
D. Primary lymphedema
E. Secondary lymphedema
ANSWER to PHOTO QUIZ

Secondary lymphedema (post trauma and post surgery)

Discussion

The patient developed progressive fibrolymphedema in his lower left extremity within 6 months of the surgical and orthopedic procedures undertaken to correct multiples fractures secondary to a severe motorcycle accident that involved his femur, patella, and the proximal end of the tibia. He reported that, the lymphedema presented a moderate grade approximately 5 years after surgery, and the left lower limb gradually increased in volume following repeated episodes of infections. In this patient, chronic secondary lymphedema was associated with cellulitis, erysipelas, and recurrent DLA, and these played a role in the development of elephantiasis in the affected extremity. Microfilaraemia was evaluated by microscopic examination of night blood films, while the Elisa method was utilized for determination of anti-filarial antibodies, and both results were negative. In samples of tissue biopsies, neither microorganisms nor micro particles of silica were found. Although dilated lymphatics were seen, pseudoepitheliomatous hyperplasia and accentuated fibrosis were absent, contributing to the exclusion of rule out the hypothesis of elephantiasis nostra verrucosa. Pathogenesis of this lymphedema secondary to trauma and soft tissue surgery includes changes in the lymphatics of the lower extremity due to an inflammatory response related to local cytokines, the action of microorganisms, and self-antigens. The longstanding inflammatory reaction elicited dilatations and obstructions of the lymphatic spaces, and enlargement of the lymph nodes in the left groin. Elephantiasis nostras verrucosa is an uncommon chronically progressive sequel of lymphedema, which includes cobblestoned papules, plaques, verrucous changes, accentuated fibrosis and hyperkeratosis, and conspicuous deformities. This condition more often affects the lower limbs. Associated factors are: obesity, chronic venous insufficiency, congestive heart failure, radiation, surgery, trauma, malignancy, scleroderma, lymphangioma, and recurrent infections (cellulites, erysipelas, and DLA) in the lower extremity. This condition may develop in patients with filarial lymphedema and primary or secondary lymphedema. Surgical treatment has limited utility in more advanced cases, but shaving and debridement may be useful. Conservative management includes compressive stockings, mechanical massages, and diuretics. Early prevention of local infections contributes to better quality of life and a more favorable outcome. Lymphatic filariasis (tropical elephantiasis) is an endemic parasitic disease affecting around 120 million people in 81 countries, with 30% of individuals showing severe disabling consequences. Filarial parasites that most commonly cause this condition are Wuchereria bancrofti, W. malayi, W. pacifica, Brugia malayi, and B. timori. Recurrent infections are common, as are cellulitis, erysipelas and DLA. Manifestations of bancroftian filariasis include lymphedema, hydrocele and scrotal elephantiasis. Microfilaraemia has been evaluated by microscopic examination of night blood films (between 22:00 and 02:00 hours), and anti-filarial antibodies can be determined by the specific ELISA test. Doppler ultrasonography, lymphoscintigraphy, and digital tonometry of the affected extremity are useful tools for an accurate evaluation of the lymphedema that is caused by filariasis. Preventive measures can be based on health education of the population at risk, and administration of single doses of albendazole associated with ivermectin or diethylcarbamazine, for the interruption of transmission in endemic regions.

Podoconiosis (non-filarial geochemical elephantiasis) is a chronic entity characterized by lymphedema on the lower extremities of individuals who have had contact of bare feet with red clay soil of volcanic origin. The majority of cases are from Africa, Central America and India. Typical features include bilateral lymphedema in the lower extremities and recurrent DLA, changes that are due to an inflammatory response, followed by fibrosis and occlusion of the lymphatic vessels of the lower limbs. This is secondary to the absorption of microparticles of silica and aluminium silicate derived from alkaline volcanic rocks through the skin of the bare feet. Preventive measures involve footwear and good daily foot hygiene in zones with risk of the disease. Treatment includes leg elevation, compression therapy, and surgical management of late lesions. Primary lymphedema is a sporadic or familial condition associated with congenital defects in the lymphatic circulation, mainly affecting the lower extremities. There are three modalities of this condition: 1) congenital or Milroy disease (appearing within the first year of life); 2) praecox or Meige disease (appearing from puberty to the age of 35); and 3) tarda (after the age of 35). Genetic characteristics and lymphoscintigraphy imaging can establish these diagnoses. Primary lymphedemas, differing from the secondary types of lymphedema, present with no previous trauma or infection. However, the occurrences of repeated episodes of cellulitis, erysipelas and DLA are very similar. Treatment includes leg elevation, compression therapy, use of diuretics, and surgical procedures.

References