Photoclinic

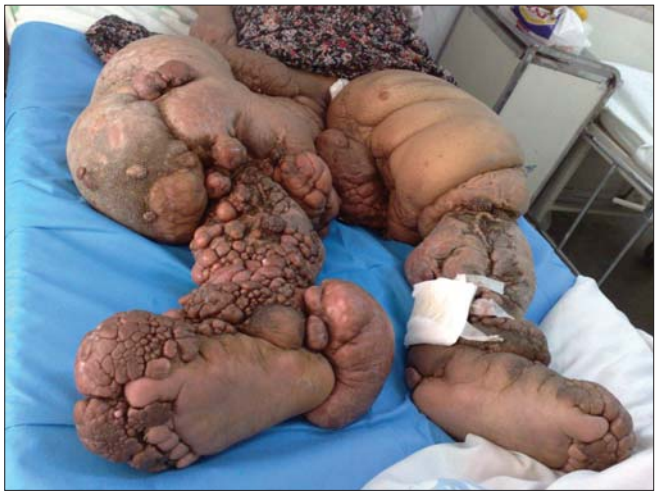

Figure 1. Lower extremity disfigurement and swelling

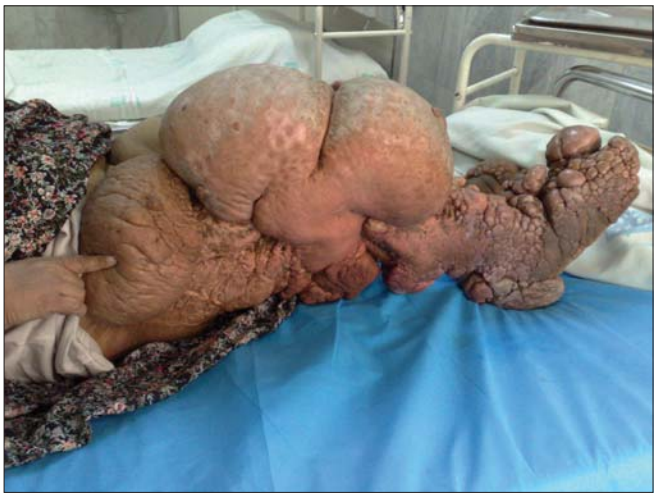

Figure 2. The patient's right lower extremity

Figure 3. The patient's left lower extremity

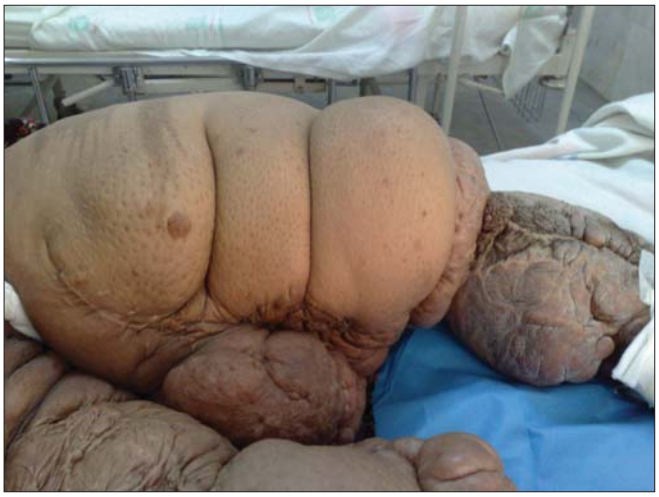

Figure 4. Peau d'orange on the patient's left thigh

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A 50-year-old woman presented to our clinic complaining of bilateral disfiguring of lower extremity by swelling and masses. She had been well until 30 years of age when she first noticed a feeling of heaviness in her legs and a need for progressively larger shoe sizes. A pitting edema had developed over the dorsum of her feet and progressed to non-pitting edema involving both her lower extremities, always more pronounced on the right side. She had experienced multiple bouts of cellulitis and skin ulcerations leaving scars on her lower extremities (Figures 1, 2, 3). The patient's travel history was negative and she could not recall any history of lower limb trauma or bare-foot walking. The patient did not report any family history of limb swellings. On physical examination, severe trophic skin changes including lichenification, acanthosis and verruciform papulonodules creating a cobblestone appearance were evident. The patient's left thigh exhibited non-pitting edema with a peau d'orange appearance (Figure 4). The remainder of the patient's skin and her eyelashes and hair were normal on examination. Extensive paraclinical work-up ruled out heart, liver, kidney and thyroid disease as well as filariasis and venous obstruction.

Anahita Sadeghi MD•[1], Behrouz Navabakhsh MD[1], Mohsen Sfandbod MD[2]
Author's affiliations: [1]Department of Internal Medicine, Shariati Hospital, Tehran University of Medical Sciences, Tehran, Iran, [2]Department of Hematology Oncology, Shariati Hospital, Tehran University of Medical Sciences, Tehran, Iran.
•**Corresponding author and reprints:** Anahita Sadeghi MD, Department of Internal Medicine, Tehran University of Medical Sciences, Shariati Hospital, N. Kargar St., Tehran 14114-13137, Tehran, Iran. Tel: +98-9121246555, E-mail: anahita825@gmail.com
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What is your diagnosis?
See the next page

Photoclinic Diagnosis: **Elephantiasis Nostras Verrucosa secondary to Lymphedema Praecox**

Elephantiasis is the disfiguring enlargement of a body part secondary to lymphedema. Elephantiasis nostras ("nostras" meaning "from our region") was originally coined in 1969 to differentiate the chronic lymphedema caused by temperate zone bacteria from elephantiasis tropica caused by the tropical Wuchereria species (filariasis). Later on, the term Elephantiasis Nostras Verrucosa (ENV) has been used to include all the non-filarial causes of lymphedema, for all of which recurrent bacterial skin infections are a common pathogenetic denominator.[1,2]

Lymphedema is the interstitial accumulation of protein-rich fluid secondary to the disruption of regional lymph drainage caused by developmental (primary) or acquired (secondary) abnormalities of the lymphatic system. Primary lymphedemas are conventionally classified into 3 groups based on their age of onset: congenital (onset before 2 years of age), praecox (onset between 2 and 35 years of age), and tarda (onset after 35 years of age) with lymphedema praecox being the most common type. The typical case of lymphedema praecox (Meige's disease) is the development of unilateral lower extremity lymphedema in a female patient around puberty and its indolent progression over time. Most congenital and tarda patients have bilateral involvement. Although clinical bilateral involvement occurs in only 30% of praecox patients, subclinical bilateral pathology has been observed in 70% of praecox cases.[3]

Rezaei, et al. have refuted the earlier assertions of a possible association between mutations in FOXC2 (a member of the Forkhead family of transcription factors) and non-syndromic lymphedema praecox.[4] FOXC2 mutations are known to cause lymphedema-distichiasis (LD) syndrome (a rare genetic cause of syndromic primary lymphedema with AD inheritance). The lower limb edema observed in LD syndrome is clinically indistinguishable from that of Meige's disease and distichiasis–a condition of having an extra row of eyelashes arising from the Meibomian glands- is difficult to detect on physical examination, unless several family members are examined. Careful phenotyping for distichiasis is therefore recommended in all cases of post-pubertal primary lymphedemas.[4]

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