An under-recognized non-filarial cause of lower extremity lymphedema

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A 14-year-old boy presents with bilateral lower extremity elephantiasis with lichenification of the overlying skin and a mossy appearance of the toes. The lesions began 5 years ago and were accompanied by edema and pruritus. The boy is from the Guraghe zone of Ethiopia and because of poor socioeconomic conditions has been barefoot throughout his entire life. He works in the fields and has no significant medical history.
Causes of lower extremity lymphedema

- Deep vein thrombosis
- Hypoalbuminemia
- Renal failure
- Lipedema
- Postoperative complications
- Baker cyst
- Neurofibromatosis
- Sclerema neonatorum
- Idiopathic edema
- Congestive heart failure
- Venous insufficiency
- Cellulitis
Differential diagnosis

- Lymphedema secondary to systemic disease (cardiac, renal)
- Vascular causes
  - Lymphangioma
  - Mycetoma
  - Kaposi sarcoma
  - Leprotic lymphedema
- Elephantiasis verrucosa nostra
- Filarial elephantiasis
- PODOCONIOSIS
What is podoconiosis?

- Podoconiosis is a non-infectious, non-filarial, geochemical lymphedema
- Derived from Greek terms *podos* and *konos*, meaning foot and dust, respectively
- Elephantiasis dates back to second millennium BC. Until 1924, all forms of elephantiasis were considered infectious until the 1960’s and 1970’s, when non-filarial elephantiasis in Ethiopia termed podoconiosis
- Affects farmers mainly in tropical parts of Africa and is caused by long term barefoot exposure to red clay soil of volcanic origin
Epidemiology

- At least 4 million people globally with podoconiosis
- Affects people in more than 20 countries
  - Tropical Africa
  - Central and South America
  - Northern India
Countries where podoconiosis is endemic or has been described
Geographical distribution of podoconiosis in Ethiopia, results from historical maps and recent surveys conducted on podoconiosis.
Prevalence

- In endemic areas, podoconiosis is more prevalent than HIV/AIDS, TB, malaria, or filarial elephantiasis.
- Ethiopia: 1 million podoconiosis patients with prevalence of 2.8-7.4%.
- Common among barefoot farmers and caused by inorganic particles (silicon, aluminum, iron) in red clay soils triggering an inflammatory process.
- Average age of onset: 20-30 years of age.
- Possible genetic component.
- Patients develop recurrent bacterial and fungal superinfections with multiple episodes of acute lymphadenitis.
- Annual economic cost > 16 million dollars per year.
- Social burden and loss of productivity.
Pathogenesis

- Inorganic particle induced inflammatory response with possible genetic susceptibility
- Mineral particles are taken up through the skin into macrophages in the lower limb lymphatics and induce an inflammatory response in the lymphatic vessels, causing fibrosis and obstruction of the vessel lumen
- A study by Price and Pitwell about the mineral content of the lymph nodes in barefoot people with and without elephantiasis of the legs confirmed presence of elements including silicon, aluminum, and iron in all barefoot people, with slightly higher amount in those with elephantiasis
- Endemic areas were free of filariasis; footwear had a protective effect; birefringent silica particles were found in the lymph node macrophages; and the dermal content of various mineral elements was consistent with local soils
- Biopsies from inguinal and femoral lymph nodes of affected individuals have shown the presence of birefringent particles and foreign body granuloma
Clinical Presentation

- Early on presents with burning and itching sensation, often following a long day in the fields
- Some associated fevers and tender lymphadenopathy
- Usually unilateral and early symptoms continue for years, may progress to involve both legs
Three early stages

✧ 1. Leg swelling

✧ 2. Lichenification

✧ 3. Mossy foot
Three forms of lymphedema

1. Soft and pitting – responds to elevation

2. Sclerotic and fibrotic leathery elephantiasis

3. Mixed elephantiasis – grossly swollen, non pitting edema, no sclerosis, no response to elevation
Other features

- Fibrous nodules
- Interdigital maceration
- Cellulitis
- Fusion of toes
- Scarring and depigmentation
Histopathology

- Histopathological examination of lymph nodes revealed birefringent minerals such as kaolinite, quartz, hematite, geothite, and gibbsite.

- Histopathology of lymphadematous skin reveals epidermal hyperkeratosis, acanthosis, and hypergranulosis.
Work up and diagnosis

✧ History of progression of swelling
  ✧ Foot is site of first symptoms (vs filariasis, leg is first site)
  ✧ Swelling bilateral, asymmetric, below knees, rarely involves groin (vs filariasis)
  ✧ No sensory loss, neurotrophic ulcers, or thickened nerves (lepromatous leprosy)

✧ Family history of the disease

✧ Exclude other causes of lymphedema
  ✧ Peripheral blood for microfilariae
  ✧ Systemic causes of lymphedema
Management

- EDUCATION, EDUCATION, EDUCATION!
  - Public health policies to improve awareness and provide education to the at risk populations
Management

- Foot hygiene
  - Wash daily with antiseptic solution for 10-15 minutes then rinse with clean water
- Footwear
- Emollients
  - Aquaphor ointment, vaseline QHS
  - Applying topical antibacterial and antifungal creams for prevention of superinfection
- Drysol to prevent moisture and maceration
- Compression and elevation
- Osteopathic manipulative medicine – lymphatic treatments
Surgical management

- Nodulectomy

  - Caution as poor wound healing
Raising awareness

- In 2011, the World Health Organization included podoconiosis in its list of neglected tropical diseases.

- The Ethiopian Ministry of Health included podoconiosis in the National Master Plan for NTDs and in modules for the upgrading of Health Extension Workers. In addition, initiatives including Footwork - the International Podoconiosis Initiative, and the Ethiopian National Podoconiosis Action Network have been formed with vision to eliminate podoconiosis.
References

- Fasil Tekola-Ayele, Wendemagegn Enbiale Yeshanehe. *Podoconiosis: Tropical Lymphedema of the Lower Legs*


Thank you