Case 1-2013: Filarial Lymphoedema of Upper and Lower limbs

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Case presentation:

Twenty two-year-old house wife from Sharkeya, Egypt presented to the outpatient clinic of Tropical Medicine Department with fatigue and bilateral heaviness and swelling in her lower limbs as well as her left upper limb that started at the dorsum of her feet and progressed gradually to affect both limbs asymmetrically (the left lower limb was swollen more than the right) (Figure 1) up to the knee and then appeared in the left upper limb starting from the hand and gradually progressing to reach level of the elbow. The patient had normal general examination.

Examination of the affected limbs revealed normal color, temperature and hair distribution. No visible or dilated veins were noted. The examination revealed no ulceration or any skin lesions in the affected limbs. The oedema was partially pitting with spongy sensation (stage 2). The circumferences of the affected limbs were measured to observe the response to therapy. The oedema level was up to knee in both lower limbs and up to elbow in the upper limb (grade 1). The pulsation of dorsalis pedis, posterior tibial, popliteal, and femoral arteries in both lower limbs were intact as well as radial, ulnar, brachial and axillary arteries pulsations in the affected upper limb. There were no palpable inguinal or axillary lymph nodes. The patient was admitted to Tropical Medicine Department, Zagazig University Hospitals for evaluation of her condition. The patient received broad spectrum antibiotic and diuretic therapy and was advised to elevate her lower limbs and wear an elastic band over the affected upper limb. The previous measures gave minimal improvement of the swelling. The patient's routine laboratory investigations were normal. The patient performed Doppler evaluation for the venous systems in the affected limbs which was normal. The nocturnal peripheral blood film was negative for *W. bancrofti* microfilaria. The lymphiscintigraphy for lower limbs revealed patent lymphatics.

Differential diagnosis of lymphoedema:

1- Primary congenital lymphoedema.

2- Secondary to injury of lymphatics by dissection, irradiation or obstruction by malignancy.

3- Long standing untreated venous insufficiency.

4- Filariasis.

5- Podoconiosis

6- Leprosy

7- Mycetoma pedis.

Staging of lymphoedema: [1, 2]

According to the consistency and oedema response to elevation of the limb:

- **Stage 0 (latent)**: The lymphatic vessels have sustained some damage which is not yet apparent. Transport capacity is still sufficient for the amount of lymph being removed. Lymphedema is not present.
- **Stage 1 (spontaneously reversible)**: Tissue is still at the "non-pitting" stage: when pressed by the fingertips, the tissue bounces back without any indentation. Usually upon waking in the morning, the limb or affected area is normal or almost normal in size.
- **Stage 2 (spontaneously irreversible)**: The tissue now has a spongy consistency and is considered "pitting": when pressed by the fingertips, the affected area indents and holds the indentation. Fibrosis found in stage 2 lymphedema marks the beginning of the hardening of the limbs and increasing size.
- **Stage 3 (lymphostatic elephantiasis)**: At this stage, the swelling is irreversible and usually the limb(s) or affected area is very large. The tissue is hard (fibrotic) and unresponsive; some patients consider undergoing reconstructive surgery, called "debulking". This remains controversial, however, since the risks may outweigh the

benefits, and the further damage done to the lymphatic system may in fact make the lymphedema worse.

Grading for lymphoedema: [1, 2]

According to the extent of involvement in the affected body parts.

- **Grade 1** (mild edema): Lymphedema involves the distal parts such as a forearm and hand or a lower leg and foot. The difference in circumference is less than 4 cm, and other tissue changes are not yet present.
- **Grade 2** (moderate edema): Lymphedema involves an entire limb or corresponding quadrant of the trunk. Difference in circumference is more than 4 but less than 6 cm. Tissue changes, such as pitting, are apparent. The patient may experience erysipelas.
- **Grade 3a** (severe edema): Lymphedema is present in one limb and its associated trunk quadrant. The difference in circumference is greater than 6 centimeters. Significant skin alterations,
  suchas cornification or keratosis, cysts and/or f istulae, are present. Additionally, the patient may experience repeated attacks of erysipelas.
- **Grade 3b** (massive edema): The same symptoms as grade 3a, except two or more extremities are affected.
- **Grade 4** (gigantic edema): Also known as elephantiasis, in this stage of lymphedema, the affected extremities are huge due to almost complete blockage of the lymph channels. Elephantiasis may also affect the head and face.

DISCUSSION

The most probable diagnosis of this condition is filariasis. Filarial disease is endemic in Egypt in some villages of Nile Delta governorates where it is transmitted by *Culex pipiens* female. [3] The prevalence of filariasis in Egypt is 10-50 case/ 100000 populations. The prevalence of asymptomatic microfilaraemia is higher. [4]

Filariasis is considered endemic in tropical and subtropical regions of Asia, Africa, Central and South America, and Pacific Island nations, with more than 120 million people infected and one billion people at risk for infection. [4] In communities where lymphatic filariasis is endemic, as many as 10% of women can be afflicted with swollen limbs, and 50% of men can suffer from mutilating genital symptoms. [5]

The most important landscape elements associated with high prevalence of filariasis in Egyptian countryside are water, clay soil and different vegetation. Knowing this association not only helps mapping of the high prevalence areas but also helps predicting high risk of transmission. [3]

Filariasis is usually diagnosed by identifying microfilariae on Giemsa stained, thin and thick blood film smears, using the "gold standard" known as the finger prick test. The finger prick test draws blood from the capillaries of the finger tip; larger veins can be used for blood extraction, but strict windows of the time of day must be observed. Blood must be drawn at appropriate times, which reflect the feeding activities of the vector insects. Most cases of elephantiasis are amicrofilaremic in a condition called (occult filariasis). [6]

In conditions where microfilaria can't be seen in blood film, various concentration methods are applied: membrane filter, Knott's concentration method, and sedimentation technique. Polymerase chain reaction (PCR) and antigenic assays, which detect circulating filarial antigens, are also available for making the diagnosis. The latter are particularly useful in amicrofilaraemic cases. Spot tests for antigen are far more sensitive, and allow the test to be done any time, rather in the late hours. [7]

Lymph node aspirate and chylus fluid may also yield microfilariae. Medical imaging, such as CT or MRI, may reveal "filarial dance sign" in chylus fluid; X-ray tests can show calcified adult worms in lymphatics. The DEC provocation test is performed to obtain satisfying numbers of parasites in daytime samples. [6, 7]

A panoramic look up on this case can help you exclude most causes of lympoedema. The primary congenital type rarely presents late in the 3rd decade of life (lymphoedema tarda). [8] However, the normal lymph flow excludes this possibility. The long standing venous insufficiency was excluded by the Doppler study the possibility of cancer is excluded by absence of lymphadenopathy, however further investigations and follow up will be necessary later to exclude the possibility of hidden malignancy. The injury of the lymphatic vessels isn't supported by history of surgery or fractures in the affected limbs. Podoconisis is a type of lymphoedema that is caused by continuous

exposure to irritant soil in genetically predisposed individuals that occurs exclusively in the lower limbs. The mycetoma pedis is associated with multiple sinus formation on the skin of the affected limb with sulpher granules-like discharge, so it was excluded because of the healthy skin overlaying oedema. Leprosy is excluded because the sensations in the affected parts were preserved and the absence of the leprosy disfiguring lesions elsewhere.

The recommended treatment for filariasis is albendazole (a broad-spectrumthis patient is anthelmintic) combined with ivermectin. A combination of diethylcarbamazine and albendazole is also effective. All of these treatments are microfilaricides; they have no effect on the adult worms. [9]

In 2003, the common antibiotic doxycycline was suggested for treating elephantiasis. [10]Filarial parasites have symbiotic bacteria in the genus *Wolbachia*, which live inside the worm and seem to play a major role in both its reproduction and the development of the disease. Clinical trials in June 2005 by the Liverpool School of Tropical Medicine reported an eight-week course almost completely eliminated microfilaraemia. [11]

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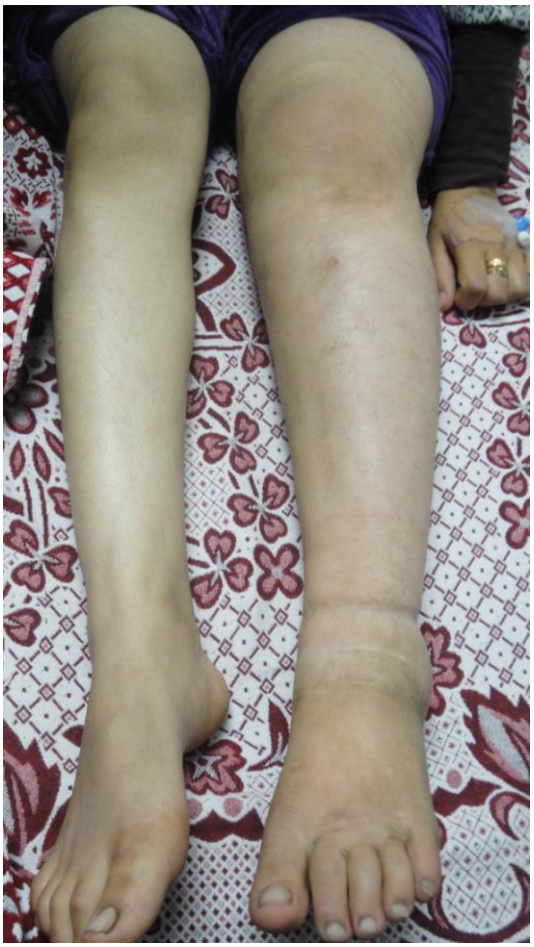

Figure 1: Asymmetrical swelling of the lower limbs (more swelling is noticed in the left side).