

Elephantiasis in the Netherlands, a rare finding and a reason to perform an autopsy

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Forensic Pathology

Elephantiasis in the Netherlands, a rare finding and a reason to perform an autopsy

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ABSTRACT

Introduction: Elephantiasis is endemic around the equator. In Western countries the phenomenon is extremely rare.
Case description: A Dutch 41-year-old severely obese male was found dead in his home. During the external examination, an extensive skin infection was seen on both lower legs. The man was treated for erysipelas two years prior and did not visit the general practitioner again. Clinical autopsy showed there was a serious stasis dermatitis of both lower legs with a superimposed erysipelas which led to a sepsis and multiple organ failure. The erysipelas presented itself as elephantiasis.

Discussion: Elephantiasis is most often caused by filariasis. In our case, the deceased had not been abroad and had predisposing factors for elephantiasis nostras verrucosa (ENV), such as obesity and an objectified erysipelas infection two years prior. The chronic skin infection caused sepsis and multiple organ failure. The position in which the deceased was found may have contributed to the death.

Conclusion: Elephantiasis is a highly uncommon phenomenon in Western countries. A chronic erysipelas can cause non-filarial elephantiasis and if left untreated, can cause death.

Introduction

The term elephantiasis, or elephant skin, refers to an enlarged body part as a result of chronic lymphedema. Filariasis, a tropical disease, is the most common cause of elephantiasis worldwide. Filariasis is called a disease of the poor and it is a severe public health problem. Filariasis is endemic in more than 80 countries. Currently, around 120 million people are infected, of which 43 million have swelling of the limbs and/or breasts. Another 120 million people are at risk of getting the infection. The Global Program for the Elimination of Lymphatic Filariasis (GPELF) was established in 2000. Worldwide there are more than 60 million filariasis programmes in 34 countries around the equator [1].

An infection with filariasis originates after a human individual is infected by the filarial parasite, which is transported by mosquitoes. Filariasis is caused by three different roundworms: *W. Bancrofti*, *B. Malayi* and *B. Timori*, which are found in tropical and subtropical areas [2,3]. The worms nestle in the lymph vessels, causing them to malfunction. The infection can be asymptomatic, acute or chronic. An asymptomatic infection causes damage to the lymph system, but has no symptoms. A chronic infection causes lymphedema or elephantiasis of the limbs. There is usually involvement of the genitals. Acute episodes often

occur beside the chronic situations because the protective skin layer is disrupted. Elephantiasis is treated with medications such as albendazol or ivermectine [4]. Filariasis is treated with diethylcarbamazine (DEC). DEC can be taken in combination with albendazole. The combination has shown good long-term results [1].

Other forms of elephantiasis are seen in breast cancer (lymphedema of the arm), Leishmania (disseminated spots on the body) and Lymphogranuloma venereum (lymphoma of the groin). Non-filarial elephantiasis of the legs is called elephantiasis nostras verrucosa (ENV) and results from the lymphatic blockage of the lymphatic system. The fibrosis can be caused by recurrent bacterial skin infections or by non-infectious causes. Characteristics are non-pitting edema, hyperkeratosis of the dermis, papillomatosis and verrucous lesions, causing the appearance of an elephants leg [2]. Stasis dermatitis is usually caused by chronic venous insufficiency [5]. Obesity is a predisposing factor, because it reduces lymph drainage and aggravates venous insufficiency [2]. The first stage is characterised by edema with a shiny and erythematous appearance. Afterwards dry, scaly spots or spots with ulceration, crusts and moisture discharge arise [6]. The skin is damaged, which causes a higher risk of erysipelas or cellulitis [7]. The skin infections are caused by beta-hemolytic streptococci of which group A is the most common. An infection with *Staphylococcus Aureus*

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is less common [8]. Eventually, the area becomes chronically colonised by multiple species of bacteria and/or fungi.

Lymph stasis can be managed by applying compression stockings and mechanical massages. Elastic bandage compression is reported to be an effective treatment. Diuretics and systemic antibiotics can be used to reduce edema. Antibiotics can be given to treat the infection. To treat the hyperkeratotic plaques, topical keratolytics or systemic retinoids might be necessary. When using retinoids, monitoring of serum lipids and liver function is needed. In advanced stages the outcome of treatment of ENV is poor [9]. The expected survival for a patient with ENV is based on the severity of lymphedema, underlying diseases, and other contributing factors. Early diagnosis and early intervention to disrupt the vicious cycle will render a better outcome. There are no articles found with exact frequency numbers of ENV. Worldwide there are a couple of case reports published stating that elephantiasis after recurrent erysipelas is very rare [10,11]. This is the first case observed (and described) in the Netherlands.

Case report

A 41-year-old Caucasian man was found dead in his home by his brother. He was lying naked on his stomach in the bathroom.

During the external investigation, performed by a forensic physician and forensic researchers, the following findings were observed:

- There were no signs of burglary or a struggle.
- He was lying in the prone position with his face against the floor and his hands beside his head (arms stretched out to the side).
- He was severely obese, estimated length 1.90 m and weight 200 kg (estimated BMI 55 kg/m²).
- There were petechia on the nose, on the eyelids and above the right eyebrow.
- The tongue protruded from the mouth. There were no abrasions of the tongue.
- The livor mortis disappeared completely on thumb pressure and the rigor mortis was in the beginning stage.
- No bruises or abrasions were present.
- There was no indication that fractures were present.
- In the skin folds between the abdomen and the groin, skin abnormalities fitting with skin infections (erysipelas) and a rough, slightly cobblestoned skin were found.
- Both lower legs were swollen and showed an extremely rough, cobblestoned skin, starting from the knee up to the toes. At the proximal part of the lower legs a white circular debris (width 5 cm) was seen. The right lower leg showed epidermolysis, with wet areas with greenish mash, haemorrhagic areas and blisters. The left lower leg showed, proximal from the knee, multiple, little, dark-red to black coloured maculae. The spots did not disappear on pressure and they presented as purpura or petechia (Photos 1–4).



Photo 2. Right foot.



Photo 3. Detail right calf.



Photo 4. Detail left calf/ankle.



Photo 1. Lower legs and feet. The green arrows show the crusts. The yellow arrow shows the fungal colonisation.

- On the left hip and buttock there was a region showing multiple, little (0.1 cm), dark-red to black coloured maculae. The spots did not disappear on pressure and they presented as purpura or petechia (Photo 5).

According to the family, the deceased had not been abroad. Until recently he had been going to his place of work. He rarely visited the general practitioner or other healthcare providers. The general practitioner last saw the deceased two years ago because of an erysipelas infection of both the lower legs. There had been no contact afterwards with the general practitioner and it was unknown whether the treatment was successful.

Because there were no signs of burglary, no signs of a struggle and no signs of violence on the body, a criminal offence was not thought to be

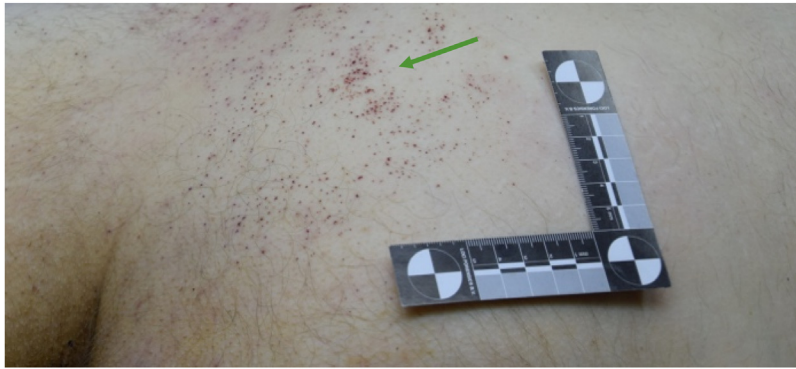


Photo 5. Left hip/buttock. The green arrow shows the petechia.

probable. Because the deceased had attended work recently and had not mentioned any medical problems, a natural cause of death was not found to be probable. Because of the extreme obesity and the prone position in which the body was found, the cause of death was thought (by the forensic physician) to be positional asphyxiation possibly after the man lost his balance. We thought he may not have been able to stand up from this position because of his weight. The petechia, visible in the face and occurring after asphyxia, support this hypothesis. The passing was registered as an unnatural death.

In consultation with the family, a clinical autopsy was performed.

The results of the autopsy: the heart (830 gr) showed concentric myocardial hypertrophy of the left ventricle with signs of hypoperfusion. There was extensive peripheral edema and stowage of blood in the lungs, liver, spleen and kidneys, fitting with cardiac decompensation. The kidneys showed necrosis of the proximal tubule cells.

The spleen was enlarged and showed a disorganized parenchyma with abnormal white and red pulpa. The adrenal glands were of normal size and showed a normal cortex and medulla. The bone marrow showed a left shift of the white blood cells, fitting with a severe acute inflammation.

Macroscopically, the skin of both lower legs showed extreme swelling of the lower legs up to the toes with a cobble stone effect. After cleaning of the body, several infiltrations of the skin were seen. Microscopically extensive scar formation with chronic inflammation infiltrates, fitting with stasis dermatitis with an acute inflammation with necrosis of the dermal collagen (Photos 6–8) was seen. The inflammation reached focally into the deep dermis.

The additional microbiological examination of a skin biopsy of both lower legs and a smear test of the skin lesions of the hip showed the presence of *Staphylococcus Aureus* and haemolytic *Streptococci* group G.

In conclusion, there is a serious stasis dermatitis of both lower legs with a superimposed erysipelas which led to a sepsis and multiple organ failure (in particular of the heart, lungs and kidneys).

Discussion

The rough cobblestoned skin as observed in the deceased can be labelled as elephantiasis. Microscopically the lymphatic stasis was confirmed. The elephantiasis was in a stage of chronic inflammation, ulceration and colonisation with bacteria and supposedly fungi. The man had not been abroad which means that filariasis could be ruled out. The

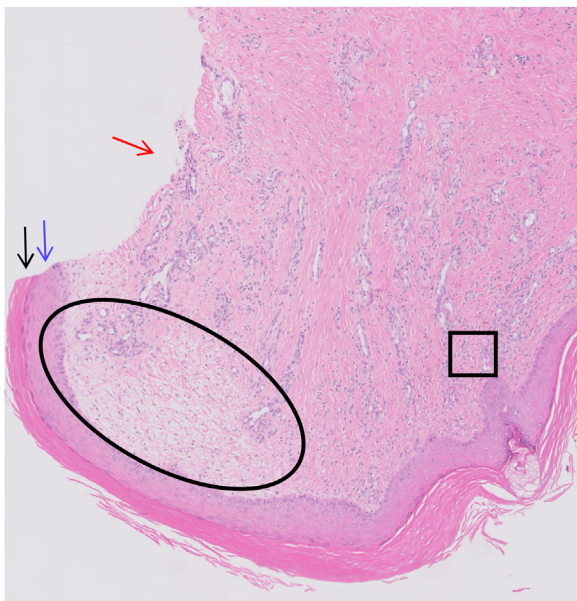


Photo 6. Overview stasis dermatitis.
Black arrow: stratum corneum.
Blue arrow: epidermis.
Red arrow: dermis.
Black oval: edema.
Black square: detail [Photo 7](#).

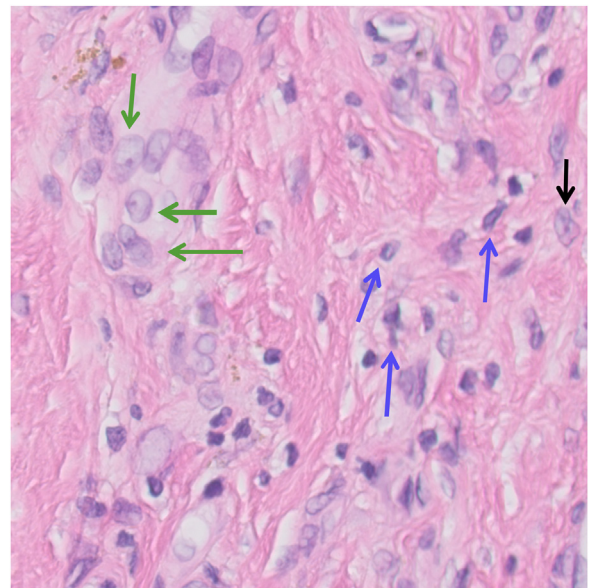


Photo 7. Detail stasis dermatitis.
Black arrow: macrophage.
Blue arrows: lymphocytes.
Green arrows: endothelial cell nuclei.

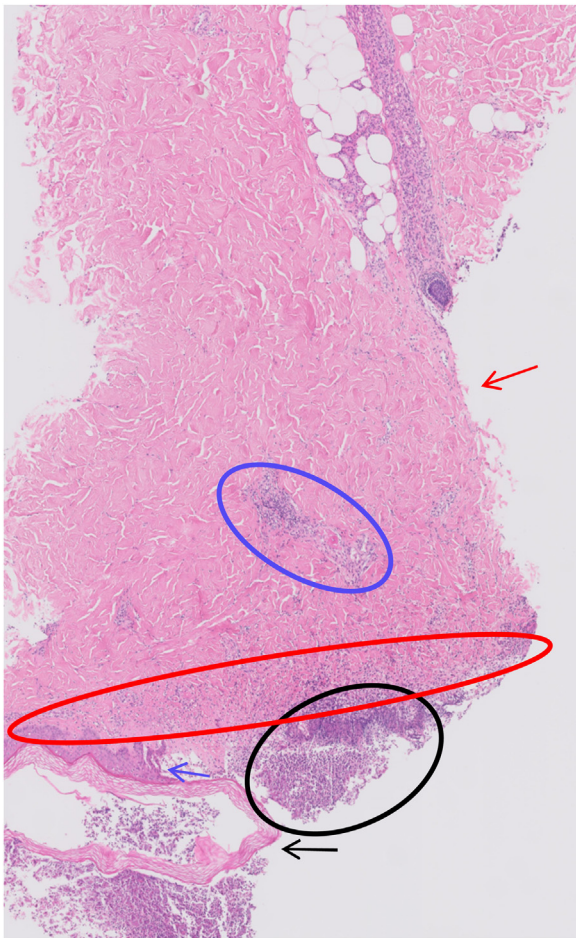


Photo 8. Overview erysipelas and pustel.
Black oval: purulent inflammation.
Blue oval: dermal perivascular lymphocytic infiltrate.
Red oval: dermal interstitial inflammation.
Black arrow: stratum corneum.
Blue arrow: epidermis.
Red arrow: dermis.

man was obese and had an erysipelas infection of his legs which was treated two years prior. No medical check had been done since the treatment of the infection. The microbiological examination of a skin biopsy and a smear test of the skin lesions of both lower legs showed the presence of *Staphylococcus Aureus* and haemolytic *Streptococci* group G, which points at a bacterial infection of the lower legs disseminating to the hip (visible via the petechia). The fungi were not objectified

microscopically. It is not clear whether the infection was treated adequately two years before and whether the symptoms had disappeared in the meanwhile. Because of the severity of the elephantiasis and the infection, it most probably had not. The conclusion of this case is that the man died of ENV caused by a chronic erysipelas infection and obesity, complicated by a sepsis and multiple organ failure. The hypoperfusion of the heart, cardiac decomposition and necrosis of the proximal tubule cells in the kidneys fit with this conclusion. The obesity of the deceased and the prone position in which he was found could have contributed to death.

Conclusion

Elephantiasis is most often caused by filariasis, a tropical disease. Non-filarial elephantiasis can be the result of a chronic erysipelas infection which can lead to sepsis, multiple organ failure and death if not treated in time. Although non-filarial elephantiasis is highly uncommon in Western countries, it does occur and should be considered as a cause of death in an otherwise unexplained death.

Declaration of Competing Interest

None.

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