JOURNAL OF FORENSIC SCIENCES

J Forensic Sci, November 2009, Vol. 54, No. 6
doi: 10.1111/j.1556-4029.2009.01146.x
Available online at: interscience.wiley.com

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Unexpected Death in Elephantiasis Due to an Abnormal Life-Style

ABSTRACT: A 22-year-old man was found dead after he had continued to sit on a reclining chair for 2 years. He had consumed an unbalanced diet, kept wearing the same pair of socks and never washed himself for the term. His skin of bilateral crura developed into elephantiasis with severely festered ulcers on its surface. At autopsy, subcutaneous edema was significant in his lower limbs, and chronic circulatory disturbance of lymphoducts and veins was observed histologically. There were no crucial findings to account for chronic edema in the lower limbs. It has been reported that maintaining a seated posture obstructs both lymphoducts and veins because of bending the groin, decreases their return flow by inducing muscular atrophy, and causes subcutaneous edema in the lower limbs. Oligotrophia and dirt on his limbs might have exacerbated the chronic edema in elephantiasis. We concluded that a long-term abnormal life-style had caused fatal elephantiasis.

KEYWORDS: forensic science, forensic pathology, chronic lymph edema, chronic venous insufficiency, elephantiasis, sepsis

Elephantiasis is characterized by cutaneous changes such as hyperkeratotic, verrucous, and papillomatous lesions like an elephant's skin, and most cases of elephantiasis occur in the lower limbs (1). The central notion for the pathogenesis of elephantiasis is chronic lymph edema. Lymph edema is commonly a result of lymphatic obstruction which leads to impaired drainage and accumulation of interstitial fluid (1). A long-term sitting posture is reported to cause chronic lymph edema in the lower limbs (2,3). It has been pointed out that keeping a seated posture would obstruct the lymph vessels due to bending of the groin and decrease the return flow of lymph by inducing muscular atrophy, finally resulting in the outbreak of lymph edema (3). Chronic lymph edema produces an increase in the number of fibroblasts and keratinocytes in the edematous tissue, leading to elephantiasis on the surface of the skin (4,5). Lymphatic filarial infection is also commonly known as the cause of elephantiasis in tropical countries (6). In addition, any surgical extirpations of lymph nodes, traumatic disruptions, or neoplastic disorders can cause elephantiasis as a consequence of disturbance of lymphatic pathways (1). A long-term venous insufficiency has been described to cause elephantiasis-like dermal symptoms via chronic congestive edema (1,7). Elephantiasis in a schizophrenic patient was reported previously by Hirano et al. (3). We report here on an atypical autopsy case of elephantiasis in the lower limbs after a long-term abnormal life-style, and discuss the cause of death. To the best of our knowledge, no similar fatal case of elephantiasis has been reported.

Case Report

A 22-year-old man was found dead sitting on a reclining chair. He was unclothed except for a filthy jacket and socks and his whole body was very dirty. According to his family, he had stayed indoors at home with them for about 10 years. Three years before death, he had been burned at the tip of his right toe with fireworks, and left the toe untreated. The burnt part of the toe gradually became so painful that finally ambulation became very difficult. Since then, he began to live an abnormal life-style on a reclining chair all day long. He continued to maintain a sitting posture on the chair, eat irregular meals of an unbalanced diet, sleep, excrete, and wear the same pair of socks, and never bathed for about 2 years. He had not received any physical examination or medical treatment, though the exudative fluid had blotted from his lower limbs for a half year before death. His appetite had been lost for a week before death. His family discovered him dead in his usual position sitting on the reclining chair.

Autopsy Findings

The autopsy was conducted on the day after death. The victim was 163 cm tall and 52 kg in weight with oligotrophia. The surface of the whole skin was adhered with a lot of dirt. The bilateral lower limbs were edematous with brown-colored hyperkeratotic, verrucous, and papillomatous lesions which were so-called "elephantiasis" on both crura. There were severely festered ulcers with necrosis that extended to the bilateral tarsal and gluteal regions and posterior surfaces of the thighs (Fig. 1). There were no findings of surgical disturbance, traumatic injury, neoplastic lesion, filarial infected disruption, or obstruction of lymphatic pathways. Pathological findings of congestive heart disruptions were not observed in the victim. Histologically, the subcutaneous tissue of lower limbs consisted of fibrotic edema with expressions of fibroblasts and keratinocytes (Fig. 2). The lymph duct in the edematous tissue was dilated, and the veins were also dilated with thickened basement membranes, perivascular infiltration of inflammatory cells, and deposition of hemosiderin. In addition, the dermal changes of hyperkeratosis were observed on the elephantiasis regions of the crura (Fig. 2). Severe suppurative necrosis was observed in the subcutaneous tissue around the festered ulcers. Intravascular aggregations of neutrophils were present in all the primary organs

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Received 26 Aug. 2008; and in revised form 9 Oct. 2008; accepted 1 Nov. 2008.

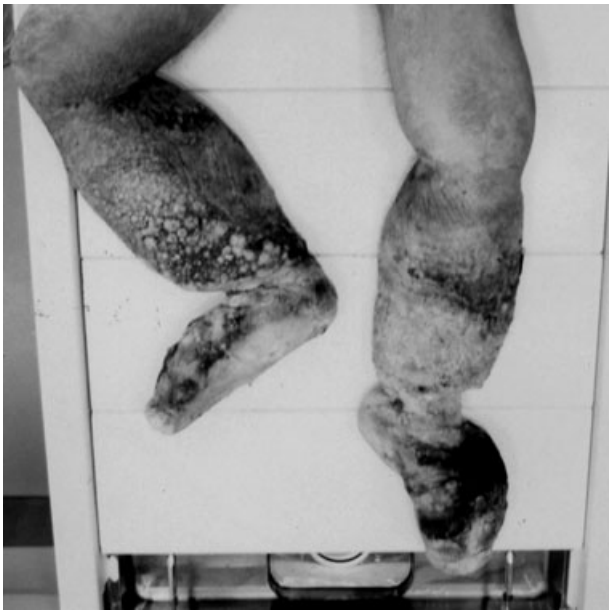

FIG. 1—*The bilateral lower limbs were edematous with brown-colored hyperkerakotic, verrucous, and papillomatous lesions which were so-called elephantiasis on both crura. The exudative fluid had blotted from infected and necrotic ulcers on the surface of the elephantiasis.*

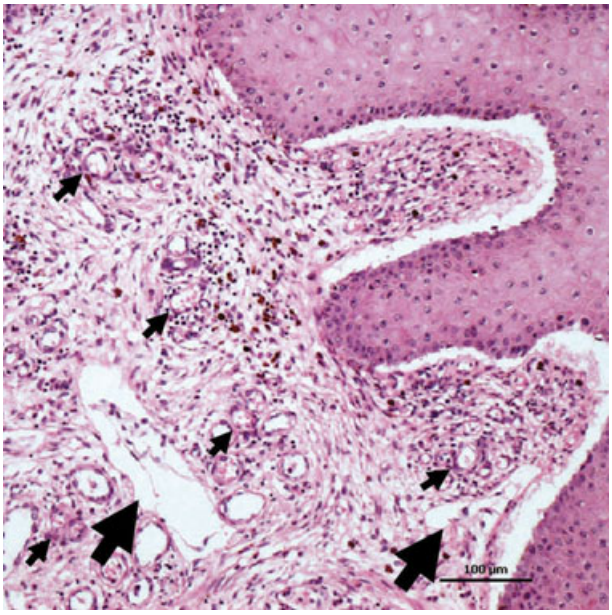

FIG. 2—*Histologic examination of elephantiasis showed the dermal changes of hyperkeratosis. The subcutaneous tissue of the lower limbs consisted of fibrotic edema with some expressions of fibroblasts and keratinocytes. The lymph ducts in the edematous tissue were dilated (large arrow). The veins were also dilated with thickened basement membranes, perivascular infiltration of inflammatory cells, and deposition of hemosiderin (small arrow). Hematoxylin and eosin staining. Bar 100 µm.*

such as brain, lung, heart, kidney, and liver. These findings implied that the victim suffered from systemic sepsis before death.

Discussion

In this case, the histological findings such as edema of the interstitial space, fibrosis of subcutaneous tissue, and thickness of the basement membrane along with enlargement of lymphoducts (Fig. 2) indicated the presence of chronic lymph edema in the process of elephantiasis. Lymph edema of lower limbs commonly develops as a consequence of any surgical, traumatic, neoplastic, filarial infected disruptions, or obstructions of lymphatic pathways (1,6). At autopsy, such pathological findings could not be observed in the victim, but the chronic lymph edema was confirmed.

In addition to the lymph vessels, pathological changes of venation such as dilatation of venous cavity and subcutaneous deposition of hemosiderin around the veins were also observed in the victim (Fig. 2). These findings indicate the presence of chronic venous congestion in the lower limbs. Ronald et al. demonstrated that the continuous elevation of venous pressure would induce the inflammatory cell collections on the venous wall through the expression of endothelial cell adhesion molecules such as vascular cell adhesion molecules and intercellular adhesion molecule (ICAM)-1 (8). The accumulated inflammatory cells injured the venous wall, deteriorated the venous function, and finally led to venous insufficiency (1). The fibrous thickness of the venous wall, perivascular infiltration of inflammatory cells, and the chronic venous congestions in the victim could be characterized as the pathological findings of chronic venous insufficiency via disturbance of venous structure. In many clinical cases, chronic elevation of venous pressure is commonly caused not only by the intrinsic obstruction of venous cavity due to either venous thrombosis, surgical disturbance, or neoplasia but also by venous stasis resulting from chronic heart disease (9,10). However, such pathological findings related to venous insufficiency were not observed at autopsy. We theorized that both elevation of interstitial pressure derived from lymph stasis and the extrinsic obstruction of the venous cavity would cause chronic venous dysfunction, and finally lead to chronic venous insufficiency in the lower limbs of the victim. Moreover, in chronic venous insufficiency the inflammatory cells aggregating around the vessels are reported to disrupt the integrity of the microvascular barrier and lead to extravasation of fluid into the interstitial space (8).

Chronic lymph stasis often increases keratinocytes and hyperkeratosis in the edematous tissues (2). On the contrary, in the case of chronic venous insufficiency, there was no hyperkeratosis but brown-colored dermal pigmentation derived from hemosiderin deposition in the chronic edematous lesion (1). In this case, both hyperkeratosis and brown-colored dermal pigmentation were observed in addition to chronic edema in the subcutaneous tissue (Fig. 2). These pathological findings also support venous insufficiency as the cause of elephantiasis in the victim.

It is pointed out that "a long-term malnutrition" and "a secondary infection in the region of elephantiasis" in addition to "a long-term seated posture" and "a lack of muscular movement in the limbs" would increase interstitial fluid retention in elephantiasis of a schizophrenic patient (3). In this case, the long-term unbalanced diet and wearing the same socks made the victim oligotrophic and dirty in his limbs. These abnormal habits could have deteriorated his chronic edema to form elephantiasis.

As for the elephantiasis resulting from chronic lymph edema, progress is usually benign and rarely causes death of the host (1). However, in the patients suffering from chronic venous dysfunction, microvascular blood flow has been suggested to be compromised, and as a consequence, tissue hypoxia would arise and induce severe cellular injury, necrosis, and ulceration (4). Indeed, the elephantiasis-like symptom such as lipodermasclerosis resulted from chronic venous insufficiency, and became fatal if complicated by necrotizing phlegmon or ulcerative infection (4). We believe

that severely infected and necrotic ulcers in the dorsum pedis or in the posterior region of the femora caused systemic sepsis and resulted in death.

In conclusion, an atypical case of elephantiasis was the subject of this report. The lesion was primarily caused by chronic edema in his limbs and ended in fatal sepsis. A long-term abnormal life-style was considered to play a pivotal role in this unfortunate scenario.

Acknowledgment

The authors would like to acknowledge Mrs. M. Ejima for technical assistance.

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