A Note from History: Elephantiasis

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The first recorded cases of elephantiasis date back to before Biblical time. Celsus (30 BC-50 AD) is credited with the first use of the term “elephantiasis” (similar to the hide of elephants; see Fig. 1) for this condition, which was also known as satyriasis, leontiasis, and sardocele. Others, including Haley Ben Abbas (930-994) and Avicenna (980-1037), accepted the term elephantiasis and set the tone for its usage during the next thousand years [1].

The cause of elephantiasis was unknown, but its endemic occurrence in Asia and Africa was recognized for centuries and was known to be associated with leprosy. It has been speculated that the soldiers of Alexander the Great who returned from India spread it to Southern Europe and Northern Africa, that the Crusaders brought it to Western and Northern Europe, and that it was later introduced to the Americas through the slave trade from Africa [1].

Although the contagious nature of leprosy was recognized in 1246 by a monk, Anglicus, the organism that causes leprosy was not identified until 1873 when Gerhard Hansen (1841-1912), a physician who worked at the hospital for lepers in Bergen, Norway, demonstrated “rod-like bodies,” ie, bacilli, in leprous lymph nodes [2]. This was 10 years before Robert Koch discovered the tubercle bacilli.

Another condition associated with elephantiasis is filariasis, which is endemic in India, Southern Asia, Egypt, and Central and South America and had long been suspected as an infectious disease. Its first description in association with elephantiasis can be found in the Ebers Papyrus, written in 1550 BC. Patrick Mason (1844-1922), a Scottish parasitologist who worked in Taiwan, reported in 1877 that the lymphedema of elephantiasis is caused by embryos of a parasite and transmitted by the mosquito, *Culex fatigans* [3]. The parasite that causes filariasis was named *Wuchereria bancrofti* in honor of Otto Wuchereria (1820-1873) who, in 1866, first described the embryo form of a worm [4], that was identified in 1876 by Joseph Bancroft (1836-1894) in cases of filarzious disease [5].

Fig. 1. Elephantiasis of the leg and foot. Note the thickened, elephant's hide-like appearance of the skin. [Fig. 189, page 628, from “A System of Surgery” by Samuel D. Gross, published by Blanchard and Lea, Philadelphia, in 1862; see Ref 16].

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As observations of elephantiasis accumulated, Dunglison's Medical Dictionary, published in 1874, in Philadelphia, categorized elephantiasis according to geographic types, such as Indian, Arabian, Greek, Italian, Barbadian, etc [6].

Despite general acceptance that elephantiasis had an infectious origin, its association with conditions other than leprosy and filariasis began to appear in medical publications. In 1892, William Milroy (1855-1942), a native of New York City who was Professor of Medicine and Hygiene at Omaha Medical College in Nebraska, reported the occurrence of congenital hereditary lymphedema in six generations of an afflicted family. The disease is known today as Milroy disease [7].

By the turn of the twentieth century, trauma was recognized as another cause of lymphedema [8]. In 1921, William Halstead (1852-1922) of Baltimore, the surgeon who introduced mastectomy with axillary node dissection at Johns Hopkins Hospital, reported the first cases of surgically induced elephantiasis [9].

James Ewing (1866-1943) of New York City indicated as early as 1919 that leprosy and filariasis can be readily separated from other causes of elephantiasis. He stated that whatever the cause of elephantiasis, "It seems to exhibit a general predisposition often combined with the effect of recurring inflammation" [10].

In 1938, a landmark paper by Telford and Simmons [11] made it clear, for the first time, that elephantiasis and lymphedema are the same disease. Telford and Simmons classified lymphedema as caused by infectious agents, trauma, congenital or genetic defects, and surgical treatment. Although chronic lymphedema, regardless of its etiology, had been suspected as a precursor of cancer for centuries, the first cases of lymphangiosarcoma in lymphedematous extremities were reported in 1906 and 1918 [12,13].

The definitive paper on malignant transformation of chronically lymphedematous tissues to lymphangiosarcoma was published in 1948 by my teacher, Dr. Stewart, in collaboration with Dr. Treves from the Memorial Hospital for Cancer and Allied Diseases in New York City [14]. This report, based on observations of six patients, appeared in the first issue of a new medical journal, Cancer, of the American Cancer Society. The described malignant transformation is known today as Stewart-Treves syndrome.

There have been many attempts to treat lymphedema through the ages, including the application of leeches, incisions, and bloodletting [15]. In 1862, Samuel Gross (1805-1884), Professor of Surgery at Jefferson Medical College in Philadelphia, wrote: "The edematous parts [should] be occasionally scarified, kept at rest in an elevated position, and well bandaged, strapped, or mechanically compressed"; he concluded that: "The treatment of elephantiasis is, at best, very unsatisfactory" [16].

Regrettably, little progress has been made from Samuel Gross’ time in the treatment of lymphedema. It is hoped that eradication of leprosy and filarial infection and advances in surgery, irradiation, and oncology may reduce the occurrence of lymphedema, as well as chronic lymphedema-associated angiosarcoma, since there is still no effective therapy for these conditions.

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