GUINEA WORM INFESTATION PRESENTING AS A SOFT TISSUE SWELLING

N. N. Murali

1General Surgeon, Department of General Surgery, Azeedia Medical College, Kollam, Kerala.

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PRESENTATION OF THE CASE

Guinea worm infection is one of the most easily preventable parasitic disease, but it is nevertheless a common cause of disability in rural areas of Africa, south-west Asia, and India. Infection occurs when drinking water is infested with infected cyclops, a microcrustacean. Worms up to 70-80 cm in length develop in the subcutaneous tissues of the feet or legs and larvae are liberated to renew the cycle when an infected individual, steps into a well or pond from which others draw drinking water. Infection is markedly seasonal due to the influence of the climate on the types of water source and the developmental cycle of the parasite. Chemical treatment of water bodies with temephos is an effective and safe way of controlling transmission. Treatment consists of taking out of each emerging worm and giving certain drugs which reduce the pain and pruritis. It is important to take out the whole worm intact without breaking the worm.

Dracunculiasis (commonly known as guinea-worm disease) is a crippling parasitic disease caused by Dracunculus medinensis - a long, thread-like worm. It is transmitted exclusively when people drink stagnant water contaminated with parasite-infected water fleas.1 Dracunculiasis is rarely fatal, but infected people become non-functional for weeks. It affects people in rural, deprived and isolated communities who depend mainly on open surface water sources such as ponds for drinking water.

A 20-year-old male presented with a swelling over right scapular region for the past 2 months. It was insidious in onset associated with occasional pain and itching. It doesn't cause any pain or numbness over right upper limb. Local examination showed a hemispherical swelling of size 5x4 cm, non-tender, well defined margins, firm in consistency and mobile. Systemic examination was within normal limits. Blood investigations and X-ray right shoulder were unremarkable. After routine work up, patient was taken up for excision swelling right scapular region making a differential diagnosis of sebaceous cyst or lipoma. An elliptical incision was placed over the swelling and incision deepened. During dissection, the live worm came out of the tissues and the whole tissues along with the worm was removed in toto. Haemostasis attained against high vascularity and wound sutured in layers. Postoperative period was uneventful. Specimen sent for histopathology. Macroscopically, grey white tissue measuring 1 cm in diameter and a linear piece of worm measuring 7 cm in length also seen with suspicious hook end. Microscopically fibrocollagenous and fatty tissue with focal collection of inflammatory cells- consistent with nematelmiths infestation possibly Dracunculus medinensis.

CLINICAL DIAGNOSIS

Guinea-worm disease is caused by the parasitic worm Dracunculus medinensis or “Guinea-worm”. This worm is the largest of the tissue parasite affecting humans. The adult female, which carries about 3 million embryos, can measure 600 to 800 mm in length and 2 mm in diameter.

History of Discovery-
Dracunculiasis is documented as early as the 15th century BC in Egypt.2 A description of the disease is found in the “Turin Papyrus,” which describes the ancient myth of the Egyptian sun god. This documentation was recently supported by the pathological examination of an Egyptian mummy in which a calcified worm was identified as Dracunculus medinensis.3 Similar documentation, dating back to 11th century AD, has also been found in Persia and Israel. It wasn't until the 18th century that such Swedish naturalist Carolus Linnaeus first suggested that the "fiery serpents" plaguing Middle Eastern countries were a type of parasitic worm. Finally, in 1870, Alexei P. Fedchenko discovered the life cycle of the Guinea Worm and the intermediate host—the Cyclops. Beginning in the early 1920's, world-wide eradication programs were first implemented which focused on water sanitation in an effort to eliminate the Cyclops and therefore dracunculiasis from public water supplies. These efforts have continued throughout the 20th and 21st centuries. In 1980, one of the goals set for the International Water Supply and Sanitation Decade (1981-1990), by the World Health Organization in collaboration with the United Nations, was the eradication of dracunculiasis. Finally, as states on the website for the
Center for Disease Control, “In May 1991 the Forty-fourth World Health Assembly endorsed the goal of eradicating dracunculiasis (guinea worm disease) by the end of 1995 (resolution WHA44.5).” This goal has been reached in numerous areas of the world, including Pakistan (last case reported in 1993), Iran (last case reported in the mid-1970’s), and India (last case reported in 1997). All current endemic areas are in sub-Saharan Africa.

PATHOLOGICAL DISCUSSION
Pathogenesis and Life Cycle
About a year after infection, a painful blister forms – 90% of the time on the lower leg – and one or more worms emerge accompanied by a burning sensation. To soothe the burning pain, patients often immerse the infected part of the body in water. The worm(s) then releases thousands of larvae (baby worms) into the water. These larvae reach the infective stage after being ingested by tiny crustaceans or copepods, also called water fleas. People swallow the infected water fleas when drinking contaminated water. The water fleas are killed in the stomach, but the infective larvae are liberated. They then penetrate the wall of the intestine and migrate through the body. The fertilized female worm (which measures from 60–100 cm long) migrates under the skin tissues until it reaches its exit point, usually at the lower limbs, forming a blister or swelling from which it eventually emerges. The worm takes 10-14 months to emerge after infection.①

![Figure 2](Image)

Figure 2

The local discomfort diminishes with the release of the embryos, but if the worm happens to break during attempted extraction, intense inflammation, with cellulitis and suppurration follows. Secondary bacterial infection is frequent. Sometimes it may lead to tetanus. The disability due to guinea worm disease lasts usually for 1 to 3 months. Sometimes the worm travels to unusual sites such as the pericardium, the spinal canal or the eyes, with serious effects. People become infected with Guinea worm by drinking stagnant water containing copepods (tiny “water fleas”) that carry Guinea worm larvae (immature forms of the worm). As the worm moves downwards, usually to the lower leg, through the subcutaneous tissues, it leads to intense pain localized to its path of travel. The burning sensation experienced by infected people has led to the disease being called “the fiery serpent”. Other symptoms include fever, nausea, and vomiting. Female worms cause allergic reactions during blister formation as they migrate to the skin, causing an intense burning pain. Such allergic reactions produce rashes, nausea, diarrhoea, dizziness, and localized oedema. When the blister bursts, allergic reactions subside, but skin ulcers form, through which the worm can protrude. Only when the worm is removed is healing complete. Death of adult worms in joints can lead to arthritis and paralysis in the spinal cord.

Location -
Subcutaneous tissues, generally in such a part of body which regularly comes in contact with cold water like hands, legs, feet etc. ⑤

Clinical Features -
Symptoms include pain in joints, nausea, fever, pruritus, blisters, ulcers, eosinophilia, and secondary infections.⑥ In
the best-case scenario, after extraction of the worm, the patient may only be disabled for 2-4 weeks. However, more often than not, the patient remains incapacitated for several months, due to the presence of multiple worms, the sensitivity of areas of the body which worms migrate to and through (such as the soles of the feet) and serious secondary bacterial infections.² The is no way to guarantee immunity. People in remote, rural communities are most commonly affected by Guinea worm disease. These patients most often do not have access to medical care. Ulcers may therefore take many weeks (8 weeks average) to heal and often become infected, causing complications such as locked joints or even permanent crippling.² Each time a worm emerges, persons are often unable to work and resume daily activities for an average of 3 months. This usually occurs during planting or harvesting season, resulting in heavy crop losses. Parents who have active Guinea worm disease cannot care for their children. They also cannot tend or harvest or crops, which leads to financial problems for the entire family.

**Figure 3**

*Incubation Period -

*In Cyclops -

After the Cyclops swallows the embryonic parasite, it takes approximately two weeks for the parasite to puncture the digestive tract, make its way to the abdomen, and develop into infectious larvae. 1-3 weeks after the larvae reaches the infectious stage, the Cyclops, which can no longer swim, sinks to the bottom of the water supply and dies.³

*In Humans-

Symptoms of the disease start to appear one year after a human drinks water containing an infected Cyclops, when the full-grown female worm starts to make its way through the body.⁶

**DIFFERENTIAL DIAGNOSIS**

Sebaceous cyst, lipoma, neurofibroma.

**Epidemiology**

The worm was present in tropical Africa, the Middle East in Arabia, Iraq, Iran, and in Pakistan and India. In India, it was seen in the dry areas in Rajasthan, Gujarat, Madhya Pradesh, Andhra, Maharashtra, Tamil Nadu and Karnataka. About 50 million people were estimated to be infected with the worm. The infection has been eradicated from India and all of South East Asia region by 2000.¹ Ten years after reporting its last GWD case, Chad reported 10 confirmed GWD cases in 2010. It has subsequently reported 9–14 cases each year from 2011 through 2015. In a highly unusual pattern for GWD, the vast majority of cases have occurred alone in separate villages, unconnected to other cases by person, place, or time. Few cases have been linked to each other or have shared common water sources. Guinea worm infections in dogs in Chad were confirmed in 2012. Since then, the number of dogs infected with Guinea worms has increased annually and far surpassed the number of human cases: 21 infections reported in 2012, 54 in 2013, 113 in 2014, and 483 in 2015.⁹ GW infections in both humans and dogs tend to cluster around the Chari River. The worms infecting dogs are genetically indistinguishable from the worms infecting humans in Chad. However, the current pattern and volume of GWD transmission in dogs in Chad has never been documented before in other countries or within Chad prior to 2012. The working theory for the unusual circumstances in Chad is that dogs and the occasional human are getting infected by eating an aquatic animal that is itself infected with Guinea worm (called a paratenic host).

**Figure 4**

**FINAL DIAGNOSIS**

Diagnosis is evident when the tip of the worm projects from the base of the ulcer. By bathing the ulcer with water, the worm can be induced to release the embryos, which can be examined under the microscope. Calcified worms can be seen by radiography. An intradermal test with guinea worm antigen elicits positive response. No serological test is available.

**Management**

There is no vaccine or drug developed to prevent or treat this disease. Until recently, the only effective treatment for dracunculiasis was the time honoured one of laboriously
rolling out each emerging worm onto a small stick every day accompanied by antibiotics to prevent secondary bacterial complications. However, in the late 1960s, three compounds were tested and reported to have a marked effect on the emerging adult female worms. These are niridazole (25 mg/kg/day daily for 10 days), thiabendazole (50 mg/kg for 3 days), metronidazole (400mg for an adult for 10-20 days). Prevention strategies include: Guinea worm case detection with the continuous surveillance, Preventing transmission from each worm by treatment, cleaning and bandaging regularly the affected skin-area until the worm is completely expelled from the body, making drinking water safe: provision and maintenance of safe drinking water supply on priority in GW endemic villages. By use of temephos, Filtering water of open water bodies before drinking in endemic areas through fine mesh (size 100 micrometers), or double layered cloth strainers to remove Cyclops, increasing awareness among endemic communities by health promotion and behaviour change.

Even though, guinea worm infestation is not common in India, occasional cases have been reported. Usage of pond water as a source for domestic purposes has become the main cause for its non-eradication. Better diagnosis and adequate treatment including proper antibiotics and intact removal of the worm is necessary for its complete control. Awareness to the public about the incidence of guinea worm is of public health interest because of the complications like involvement of joints and the pericardium which can be the cause of permanent ailments.

REFERENCES