

Unusual cutaneous manifestations of dracunculiasis: Two rare case reports

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Abstract

A nematode parasite, *Dracunculus medinensis*, causes dracunculiasis. Despite being non-fatal, this condition causes significant morbidity. Dracunculiasis is considered an eradicated disease in India since 1999. We report two cases that document the unusual linear morphea-like morphology of the calcified *D. medinensis* and the rare periorbital location of the worm. These cases presented a rare and a diagnostic challenge, considering the eradicated status of dracunculiasis.

Key words: Dracunculiasis, morphea, periorbital

Introduction

Dracunculiasis, also called guinea worm disease, is caused by *Dracunculus medinensis* which belongs to the nematode superfamily Dracunculoidea of the order Spirurida which are tissue parasites.¹

In 1986, 20 Asian and African countries recorded 3.5 million cases. In 2002, 13 African countries reported >55,000 cases.

The global incidence of new cases of dracunculiasis has decreased to 25 in 2016, mostly confined to three countries such as Chad, Sudan and Ethiopia.³

In India, the last reported case was in July 1993, though subsequently, three more cases were reported from parts of Rajasthan.⁴ The goal is global eradication of this disease by 2020.⁵

The disease is not fatal but its complications may cause considerable disability in acute and chronic stages.

The cases presented here show that though considered an eradicated disease, occasional guinea worm cases with unusual cutaneous manifestations may occur and require a high index of clinical suspicion.

Case Report

The first case was a 45-year-old female housewife, from Punjab complaining of mild swelling, itching and discomfort around the right ankle for five years. There was no history of trauma or associated pain.

On examination, we noted a serpiginous swelling with intermittent break-up over the medial aspect of her right ankle and Achilles tendon. Overlying skin showed brownish-black hyperpigmentation with the pulled up appearance and puckering at a few places [Figure 1].

On palpation, along the lesional length, non-tender, thick, indurated cord-like swelling was appreciated. The patient denied biopsy but consented to a local ultrasound examination. Differentials considered were morphea, resolved thrombophlebitis and fibromatosis.

On ultrasound biomicroscopy [Figure 2] with a 50 MHz frequency probe, the dermis was hypochoic in echotexture and increased in thickness [red line]. A few small calcifications (purple arrow) were present in the superficial region of the subcutaneous tissue.

On high-frequency ultrasonography with 7–15 MHz multifrequency probes with color and spectral Doppler

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Figure 1: A serpiginous swelling with intermittent break-up with brownish-black hyperpigmentation and the pulled-up appearance and puckering at few places

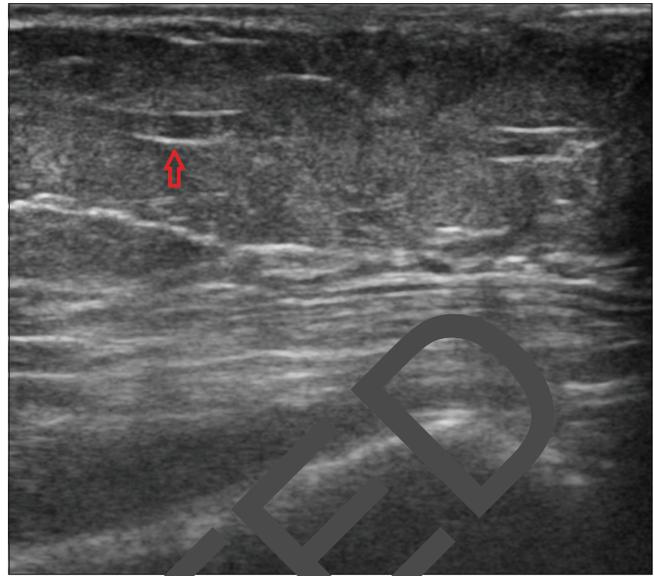


Figure 3: A static linear cord-like structure (which is the worm, measures 1.5–2 mm in diameter [red arrow])

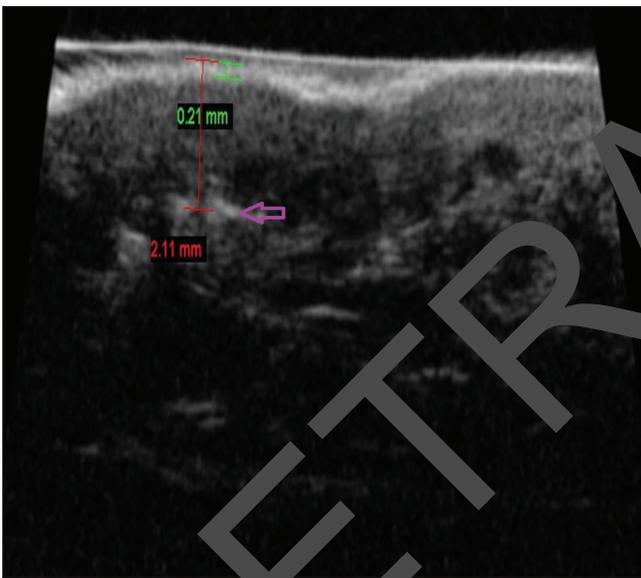


Figure 2: The region is hypoechoic and has increased in thickness (red line). A few small calcifications (purple arrows) are noted in the superficial region of the subcutaneous tissue.

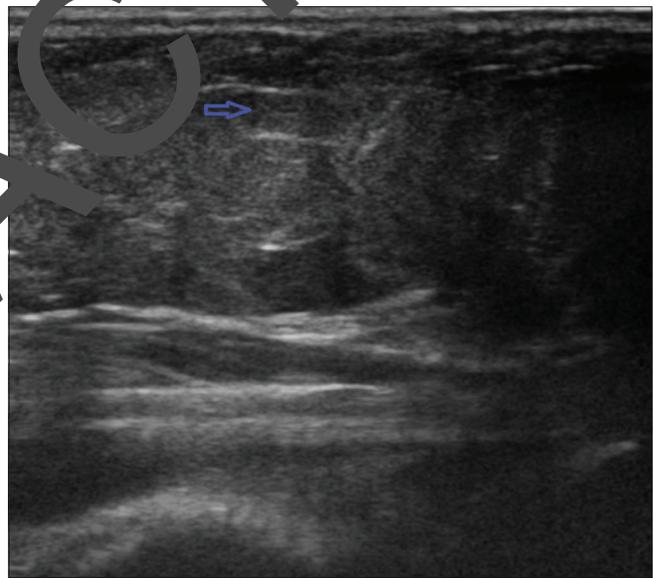


Figure 4: A thin linear anechoic region within the worm is most probably fluid in the worm's gastrointestinal tract (blue arrow)

[Figure 3], we identified a worm having static linear elongated to curvilinear cord-like structure measuring 1.5–2 mm in diameter [red arrow]. No color flow/vascularity was present within this cord-like structure on color and spectral Doppler study. A thin linear anechoic region within the worm was observed possibly representing fluid in the worm's gastrointestinal tract [Figure 4, blue arrow]. The above findings suggest a dead worm with inflammation of the skin and subcutaneous tissue around the ankle joint.

The clinical presentation, along with the findings on ultrasonography, pointed to the diagnosis of dracunculiasis.

Unfortunately, this patient refused to undergo any surgical intervention and was lost to follow up for further management.

The second case was a 40-year-old female housewife, from Maharashtra, who presented with periorbital swelling and redness associated with itching and crawling like sensation in the skin for 15 days. There was no history of traveling, trauma or associated pain.

Examination showed an erythematous, non-indurated periorbital swelling with a worm-like serpiginous outline along the lateral canthus of the right eye [Figure 5, black arrow].

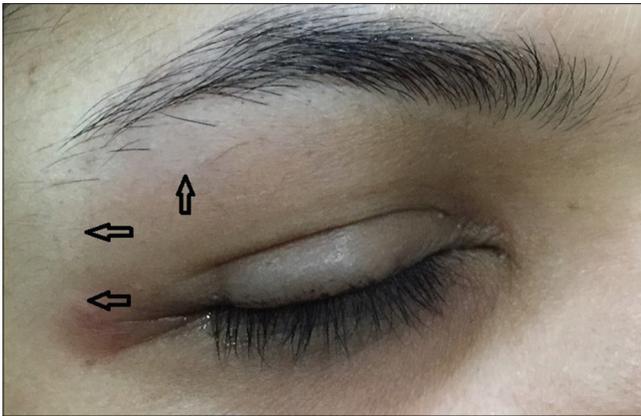


Figure 5: Erythematous, edematous, inflamed non-indurated periorbital swelling with rope-like structure at (black arrow) lateral canthus

Ultrasound biomicroscopy of the skin and subcutaneous tissue in the lateral canthus and supraorbital region of the right eye was performed [Figure 6].

We observed a well-defined, tubular, hypoechoic lesion measuring 1.5 mm in diameter and 4 mm in length with a thin, linear, hyperechoic area within it (the worm, green arrow) in the dermis and the subcutaneous tissue. The thin, hyperechoic line indicates the gastrointestinal system of the worm. We also noted a well-defined, oval, hypoechoic lesion measuring 4 × 3.7 mm (yellow arrow) noted on the right side of the worm and surrounding it in the subcutaneous tissue. The hypoechoic, tubular lesion with branching pattern in the subcutaneous tissue below the worm possibly occurred due to local tissue inflammation.

The patient consented to undergo surgical intervention for further confirmation. During surgery, a moving worm was identified and extracted by forceps [Figure 7].

The extracted worm was identified as *Dracunculiasis medinensis* by the microbiologist.

Discussion

Dracunculiasis, also known as guinea worm disease, is a preventable water-borne parasitic disease. It is transmitted when people drink stagnant water contaminated with copepods that carry guinea worm larvae. Humans are the principal definitive host, and *Cyclops* is the intermediate host. A significant transmission route is drinking unsafe water containing small *Cyclops* infected with the larvae of *D. medinensis*.⁶ Transmission is limited among remote rural settings without a safe portable water supply.

Dracunculiasis was previously considered an exclusively a water-borne anthroponosis. Recent reports of infection by ingestion of paratenic (frogs) or transport (fish) hosts support that dracunculiasis could also be a food-borne zoonosis.⁵

The clinical features of dracunculiasis include mild fever, itchy rash, nausea, vomiting, diarrhea and dizziness. Nearly

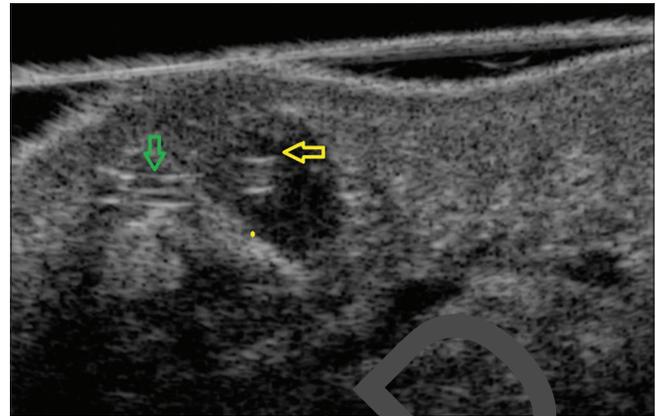


Figure 6: There is a well-defined, tubular, hypoechoic lesion measuring 1.5 mm in diameter and 4 mm in length with a thin, linear, hyperechoic area within it, noted in the dermis and the subcutaneous tissue. This is the worm (green arrow). There is a well-defined, oval, hypoechoic lesion measuring 4 × 3.7 mm (yellow arrow) noted on the right side of the worm and surrounding the worm in the subcutaneous tissue.

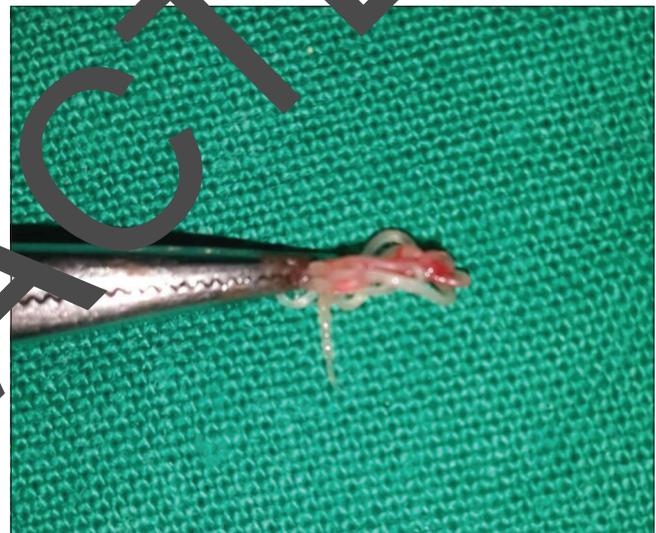


Figure 7: Extracted worm winding around the forceps

one year after infection, the female worm induces a blister on the skin, generally on the distal lower extremity, which ruptures. Acute stage complications include cellulitis, abscesses, septic shock and septic arthritis, while late-stage calcification of worm and joint deformities can occur.⁷

If the worm fails to reach the skin, it disintegrates or becomes calcified which becomes readily appreciable on the x-ray.⁸

In our first case, the calcified worm was present in subcutaneous tissue, whereas the second case presented with unusual periorbital location and mild cellulitis like features. The calcified worm may remain indolent or rarely causes intermittent mild discomfort and itching. Both of our cases presented with discomfort and itching.

Guinea worm calcification may take several forms, ranging from linear elongated to curvilinear to oval shapes. Muscle movement may break up the worm in several places,

leading to elongated, nodular, beaded and fragmented appearance.⁸

The characteristic appearance is long linear, serpiginous or coiled, whorled “chain mail” type of calcification in the soft tissues, mostly in the lower extremity.⁷

In our first case, linear elongated to curvilinear worm was present in the dermis and subcutaneous tissue.

Multiple “rice grain” calcifications oriented along the direction of the muscle fibers are seen in cysticercosis.⁸ *Loa loa* and *Onchocerca volvulus* may calcify but show small, coiled masses of calcification and occasionally may be linear, but never as large or extensive as the guinea worm.⁷

The localization and the characteristic linear elongated and curvilinear appearance of worm with calcification on ultrasound in our patient were diagnostic of calcified guinea worm presenting as linear morphea-like lesion. In the second case, a live worm was extracted after surgical intervention.

Although dracunculiasis has been eradicated from several countries, it might present in an unusual location (periorbital) with acute skin manifestations. The remnant of disease can still be detected in some patients as a calcified worm. A high index of clinical suspicion is required to diagnose these of guinea worm disease.

Conclusion

Although dracunculiasis has been considered as an eradicated worm from several countries, unusual acute skin

manifestations and the ghost of the disease in the form of a calcified worm can still be seen in some patients. A high index of clinical suspicion and collaboration with radiologists is required to diagnose these cases of guinea worm disease.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent.

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Conflicts of interest

There are no conflicts of interest.

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