CASE REPORT

An unusual cause of an ankle mass

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SUMMARY

We report the case of a patient who presented with a 7-year history of a mass over the medial aspect of his right ankle, which had been gradually increasing in size. He had given up his occupation as a bus driver due to decreased movement of his ankle. An initial diagnosis of endemic syphilis was made after treponemal antibody and treponema pallidum particle agglutination tests were positive. However, following surgical debulking, cultures grew *Fusarium solani* and the diagnosis was changed to eumycetoma. He received prolonged treatment with antifungal agents and at 18 months follow-up remains well.

BACKGROUND

Mycetoma is a chronic and mutilating fungal and bacterial infection of tropical and subtropical countries. It may be caused by true fungi or by filamentous bacteria. Disease begins following inoculation into the subcutaneous tissue and may spread to involve the skin and deep structures and result in tissue destruction and deformity. Early diagnosis, antibiotics for the bacterial type and surgical excision with clear margins and azole therapy for the fungal type result in the best outcomes.

CASE PRESENTATION

A 42-year-old man presented with a mass of approximately 15 cm diameter involving the posteromedial aspect of his right ankle and hind foot (figure 1A, B). He recalled it being present for 7 years, gradually increasing in size. At presentation, the mass was firm with some fluctuant areas. It was not inflamed but there was some increased pigmentation of the overlying skin. The examination was otherwise unremarkable. There was no evidence of discharging sinuses and the patient denied noting sinuses previously. He had no significant medical history and was taking no regular medications. He was born and brought up in Abidjan in the Ivory Coast and had little rural exposure. He had moved to the UK in 1990, aged 28, and had only once returned to Africa. He had been working as a bus driver, but had been forced to stop due to the decreased movement of his ankle and inability to wear enclosed shoes.

INVESTIGATIONS

Routine blood tests were normal and an HIV test was negative. MRI revealed an inhomogenous infiltrative soft tissue lesion with abnormal bone signal (figure 2). This was felt to be a reactive bone oedema. An Ultrasound-guided biopsy revealed no bacteria growth and was acid-fast bacillus negative. No mycobacteria or fungi were cultured after

6 weeks' incubation. Histology showed a chronic fibrosing inflammatory condition with abundant plasma cells and a venulitis, but excluded sarcoma as a possible diagnosis. Treponemal antibody and treponema pallidum particle agglutination (TPPA) were both positive. There was no history of genital ulceration and a sexually transmitted infection screen was negative but a diagnosis of endemic syphilis was considered on the basis of serology alone and the patient was treated with intramuscular benzathine penicillin G and referred for surgery. This diagnosis was made on the basis of serology alone despite clinical features not suggestive of endemic treponemal disease.

DIFFERENTIAL DIAGNOSIS

- Bone tuberculosis
- Bone tumour

TREATMENT

A 12×12 cm firm lobulated mass was excised through an extensile posteromedial approach, however, due to intradermal infiltration, the

Figure 1 Photographs showing the mass over the medial aspect of the right ankle: (A) posterior view; (B) medial view.

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Figure 2 MRI demonstrating the mass.

procedure was considered debulking rather than eradication (figure 3). Plaster cast immobilisation and a pneumatic walker boot were utilised for 1 month postoperation.

OUTCOME AND FOLLOW-UP

Postoperative biopsy revealed an acute suppurative inflammatory response with formation of microabscesses centred on clumps of basophilic granular material. Fungal stains showed septate budding hyphae. A Gram stain for actinomyces was negative as were the treponemal stains. As such, the diagnosis was revised to eumycetoma and treatment with itraconazole (200 mg twice daily) was started. Ten weeks after surgery the wound started to deteriorate with the appearance of suspicious spherical bleb-like elevations. Fungal cultures ultimately confirmed the presence of *Fusarium solani* that was resistant to itraconazole, and therefore treatment was switched to voriconazole (400 mg once daily) on the 80th day postoperatively. The wound quickly began to improve. At 6 months following surgery the patient had returned to driving a bus. A repeat MRI at 6 months

demonstrated a significant improvement, however, therapy was continued for 18 months postoperation. He tolerated the voriconazole therapy well and did not suffer adverse effects. At last follow-up, 18 months from surgery, he remains well with no recurrence of the mass clinically or radiologically. He has had MRI scans at 6 months and 18 months following the surgery. He will remain under surveillance at 6 monthly intervals, with MRI scans annually.

DISCUSSION

Mycetoma is a chronic, localised, slowly progressive granulomatous infection of soft tissue. It may be caused by true fungi (eumycetoma) or by filamentous bacteria (actinomycetoma). The disease most probably begins following inoculation into the subcutaneous tissue following minor trauma, although many patients have no history of trauma. It may spread to involve the skin and deep structures, and result in tissue destruction and deformity. Although usually found in the foot, other sites can be affected as well.¹ It is likely that the patient in this case acquired the infection while living in the Ivory Coast, decades earlier. While he only reports a 7-year history, we suspect he may not have noted a much smaller lesion, as it would have been asymptomatic and not caused him any significant functional impairment.

The earliest descriptions of this disease process are found in ancient Indian Sanskrit texts, which refer to a mass that is described as ‘anthill foot’.² However, the first case was reported in 1832 in India. It was subsequently named Madura foot in 1846 by Colebrook after it was noted in field workers in the Madurai district of Tamil Nadu.³

Mycetoma is endemic in tropical and subtropical countries between latitudes 30°N and 15°S, with highest numbers reported from Mexico, India and Sudan.⁴ More rarely, cases have also been reported from temperate countries.⁵ Most cases are found in men aged between 11 and 40.⁴

Many species have been identified to cause mycetoma. Those of fungal origin tend to occur in hot, arid regions with

Figure 3 Photograph showing the mass that was excised.

prolonged sunshine. Bacterial mycetoma, however, occurs in hot regions with high rainfall.⁶ Bacterial mycetoma is more common, however, the commonest causative organism overall is *Madurella mycetomatis*, a fungus.⁴ The *Fusarium* species, isolated in this case, is rare. The prevalence of the different aetiological agents varies between countries. The incubation time ranges from weeks to years.⁷ After a traumatic inoculation, the pathogens grow and survive through the production of grains. These grains are found within abscesses surrounded by granulation tissue.⁸ The disease manifests initially with painless subcutaneous nodules that gradually increase in size and are then accompanied by sinus formation.⁹

Tissue biopsy or microbiological culture will usually provide the diagnosis, however, for certain fastidious organisms this is not always possible. In these cases, MRI can be useful in making the diagnosis. The 'dot-in-circle sign' is highly specific for mycetoma. This is the appearance of fungal grains as low signal intensity within hyper-intense lesions representing the surrounding granulomata.^{10 11} Furthermore, a radiological scoring system has been developed in Sudan, called the 'Mycetoma Skin, Muscle, Bone Grading System' (MSMBS). The score is based on MRI appearances of the skin, subcutaneous tissue, muscle and bone. The typical appearance of actinomycetoma is evidence of soft tissue micro abscesses with periosteal bone reaction and reactive sclerosis. The eumycetoma, however, shows evidence of bone destruction and cavitation.¹²

Bacterial and fungal mycetomas differ in their response to treatment. Actinomycetoma responds well to drug therapy. Combined therapy of cotrimoxazole plus an aminoglycoside is thought to be best. Eumycetoma is more difficult to cure, and a combination of medical and surgical treatment is the gold standard.¹³ Pharmacological treatment with ketoconazole or itraconazole for 18–24 months is recommended as well as surgical excision or debulking. Long courses of ketoconazole therapy are more likely to be associated with adverse effects in comparison with other azoles. Newer antifungals including voriconazole and posaconazole may be more effective, however, their high cost and limited availability in endemic areas preclude their use. There is also limited evidence for their use in treating eumycetomas. In Khartoum, Sudan, where mycetoma is more common, patients receive preoperative antifungal treatment for 6 months to a year, to allow encapsulation and localisation of the lesion. This is followed by wide local excision followed by repeated debridements, if necessary, to reduce the lesion size. Amputation is indicated for massive disease with bony destruction, severe secondary bacterial infection that is refractory to antibiotic therapy, massive disease that does not respond to medical therapy and patient preference. In a study of 1544 patients with eumycetoma, predictors of cure included longer medical and surgical treatment, combined medical and surgical treatment,

and first occurrence of mycetoma. Lesions greater than 5 cm and long disease duration were associated with amputation.¹⁴

Learning points

- ▶ Mycetoma can be difficult to diagnose but should be considered as a differential diagnosis of any foot and ankle lump in a patient from the subtropics.
- ▶ Eradication surgery can be possible with small lesions but for massive lesions, repeated debulking surgical excisions and prolonged antifungal treatment are necessary.
- ▶ Serial MRI may aid decision-making over treatment duration although a cure may never be possible.
- ▶ Amputation remains an option for recurrent intractable disease.

Competing interests None.

Patient consent Obtained.

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