

Olgu Sunumu

Mycetoma of the Upper Extremity; A Reason For Above Elbow Amputation

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Abstract

We report a case of Mycetoma of the forearm and hand in a 52 year-old man caused by *Streptomyces* spp. Because of the extensive involvement of all compartments that rendered the extremity functionless, an above elbow amputation was done. In the early stages of the disease the treatment with antibiotics is curative. We report this case of extensive actinomycotic mycetoma of the hand for its rarity and we encourage clinicians to maintain a high index of suspicion in cases of chronic granulomatous infection of the upper extremity to prevent the catastrophic complications that may result in limb amputation.

Key words: Mycetoma, amputation, actinomycosis

Introduction

Mycetoma is a chronic, local, subcutaneous granulomatous and slowly progressive inflammatory disease caused by either fungi (eumycetoma) or bacteria (actinomycetoma). The presentations of this rare condition include tumefaction, multiple discharging sinuses, and grain-filled sinuses. The differential diagnosis includes other subcutaneous infectious, neovascular proliferation, and soft tissue tumors (1,2). They are not uncommon in endemic regions, an early diagnosis may improve the result and lead to initiation of appropriate treatment that may prevent more serious complications such as an amputation (1). Investigations are complicated by the fact that most patients tend to present late, by lack of clinical signs, or health care facilities, or because of patients' scare of amputation (3,4,5). The foot is affected most often in mycetoma. The lesions are seen usually on the dorsal aspect of the foot.

The hand is the second common region (6). The granules in the discharge are aggregates of microcolonies of the organism (7). Once established, mycetoma enlarges and spreads through fascial planes leading to deformity and disability; amputation may be required if the condition is allowed to advance (8). Eumycetoma tends to remain encapsulated and runs an indolent course over many years, whereas actinomycetoma tends to be locally aggressive, and often involves bone at an early stage and has a tendency to infect secondary sites (9).

Case Report

A 52 year-old man was referred to our outpatient clinic with a diagnosis of chronic swelling on the whole right forearm and the hand. His past history included an accidental injury to the right thumb as he was working in a garden in Saudi Arabia 22 years before. At the beginning of the illness, he developed a single firm, painless nodule on the hand which increased in size over months and on the surface of which vesicles gradually appeared. Then, the vesicles burst and multiple sinus tracts were formed which began discharging fluid containing small granules. The sinus tracts healed and re-occurred intermittently, discharging a sero-purulent yellowish-gray fluid. Within 12 years, there was extensive involvement of the volar and the dorsal surface of the hand and forearm. Most of his wrist and hand joint movements were restricted with deformations. He has been treated surgically and medically for the past twenty years in different centers in Saudi Arabia and Turkey. However no reports were found about his previous treatment and he denied

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a history of intravenous drug abuse or systemic illness. Physical examinations revealed an extensively swollen right forearm and hand with multiple discharging sinuses and foul smelling (Figure 1). Laboratory investigations revealed a normal leukocyte count, anemia (Hb 7.9 g/dL), a high ESR (120 mm/h) and high C-reactive protein (CRP: 13.3). Radiologic examination of the hand and forearm showed osteolytic lesions, periosteal reaction and bone destruction in the phalangeal, carpal, metacarpal and forearm bones with massive soft tissue swelling (Figure 2).



Fig. 1. A gross view of the diseased upper extremity.



Fig. 2. AP view of the right forearm and hand showing massive soft tissue mass, periosteal reaction and bone destruction.

MRI of the hand and forearm revealed diffuse multiple small discrete spherical hyperintense lesions. In the center of some of these lesions, there was a tiny hypointense focus, resulting in the dot-in-circle sign (Figure 3).

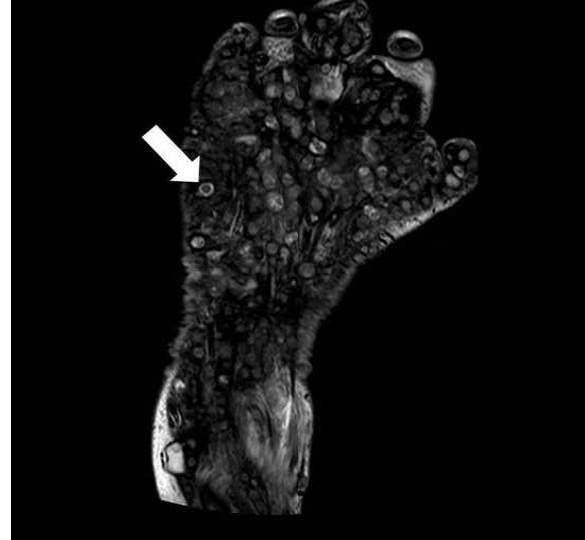


Fig. 3. MRI image showing multiple dot-in-circle lesions which represent the colonies of the organism.

This was at first misinterpreted as dilated lymphatic, however actually this was representing the discharging sinuses. Scanning of technetium bone scan showed increased uptake in the bone and soft tissue of the hand and no sign of metastasis in any other region. Macroscopic examination revealed extensive involvement of all tissues with multiple sinusoidal structures containing multiple small spherical yellow color granules (sulphur granules). Microscopic examination revealed actinomycetic colonization in the bony lamellae (Figure 4). Due to the chronic and the extensive involvement of the upper extremity, we performed an above elbow amputation. The abnormal laboratory findings returned to normal levels within 6 weeks after the operation.

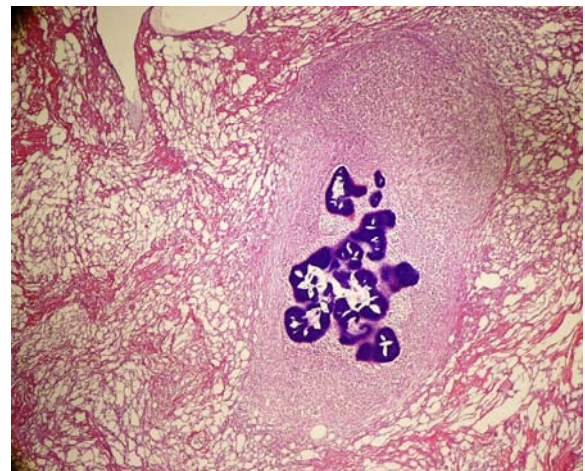


Fig. 4. Micrograph showing Streptomyces spp. grain surrounded and infiltrated by polymorphous inflammatory cells.

Discussion

Because of rarity of mycetoma in the upper extremity and in our region, the current case was misdiagnosed as a case of elephantiasis. Blood cultures for microfilaria and the cultures obtained from the surface of the sinuses were negative. The diagnosis was made only after biopsy which revealed the characteristic sulphur granules which is strongly characteristic of actinomycosis. The multiple circular lesions seen on MRI were first misinterpreted as dilated lymphatic vessels. These circular lesions are characteristic for actinomycosis. The dot-in-circle sign was first described by Sarris et al. (10) as a unique pathological feature of mycetoma.

It is seen as a tiny hypointense focus within high-intensity spherical lesions. The small central hypointense foci represent the fungal balls or grains, while the surrounding high-signal intensity foci represent the inflammatory granulomata. In general, a combined therapy is the standard approach in treating this condition (6). For chronic, resistant and extensive involvement, like the one we reported, the above elbow amputation seemed to appear a reasonable approach. A giant size hand and forearm with extensive involvement in an adult is very rare. The patient's history revealed that he was presented with findings of a simple infection when he was 30 years of age. At that time he was managed with surgical debridement without adjunctive long-term chemotherapy. Perhaps if proper diagnosis and long-term chemotherapy had been administered, he might not have presented with such extensive involvement. In advanced and neglected cases of actinomycetoma, amputation seems to be inevitable. Therefore, we encourage clinicians to maintain a high index of suspicion in cases of chronic, granulomatous infection of the hand. We recommend a close collaboration with the department of radiology and a multidisciplinary approach to the chronic granulomatous infection of the upper extremity.

Üst Ekstremitede Miçetoma; Dirsek Üstü Amputasyon Sebebi

Özet

Biz burada Streptomyces spp. tarafından ön kol ve elde oluşturulmuş 52 yaşındaki miçetoma vakasını

sunduk. Tüm kompartmanların yaygın tutulumu nedeniyle ekstremitte işlevsiz hale gelmiş ve dirsek üstü amputasyon yapılmıştır. Hastalığın erken evrelerinde antibiyotik küratiftir. Biz bu olguyu miçetomanın elde nadir görülmesi ve kronik granülatöz enfeksiyon varlığında, amputasyon gibi katastrofik komplikasyonların engellenmesi için klinisyenin yüksek oranda miçetomadan şüphelenmesi gerekliliğini belirtmek için sunduk.

Anahtar kelimeler: Miçetoma, amputasyon, aktinomikozis

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