

Mucormycosis of Forearm Masquerading as Soft Tissue Sarcoma

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Abstract

Case Report

Mucormycosis is a rare invasive and life-threatening fungal infection usually affecting immunocompromised individuals caused by zygomycetes fungi. Incidence of mucormycosis in immunocompetent individuals is quite rare and is usually attributed to minor trauma. Most common clinical presentations include rhinocerebral and pulmonary forms followed by cutaneous form of mucormycosis where it usually presents as indurated erythematous plaques, targetoid and purpuric lesions. We present a case of mucormycosis in a 34-year-old immunocompetent male presenting with forearm masquerading as soft tissue sarcoma for its rarity in site and clinical presentation.

Keywords: Mucormycosis, forearm swelling, soft tissue sarcoma.

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INTRODUCTION

Mucormycosis is a rare but life-threatening infection, caused by fungi of the Mucorales order which belong to the class zygomycetes [1]. Zygomycetes organisms are unique among filamentous fungi because of their high capacity to cause devastating disease even in individuals with no underlying condition [2]. Trauma is the most common predisposing factor leading to mucormycosis in immunocompetent patients [3]. We present a case of cutaneous mucormycosis in a 34 year old immunocompetent male which is a rare manifestation of an aggressive fungal infection where early diagnosis and treatment are vital for better outcome.

CASE REPORT

A 34 years old male, welder by occupation presented with a swelling in the right forearm for about 2 months. A history of antecedent trauma was present after which the swelling appeared which was initially small in size and gradually progressed to reach the present one. A small ulceration developed over the surface of the swelling around 2 weeks later. There was no pain associated with the swelling. No other associated co-morbidities were present. The patient initially sought treatment from an alternate practitioner who wrapped the swelling in a bandage of herbal combinations.

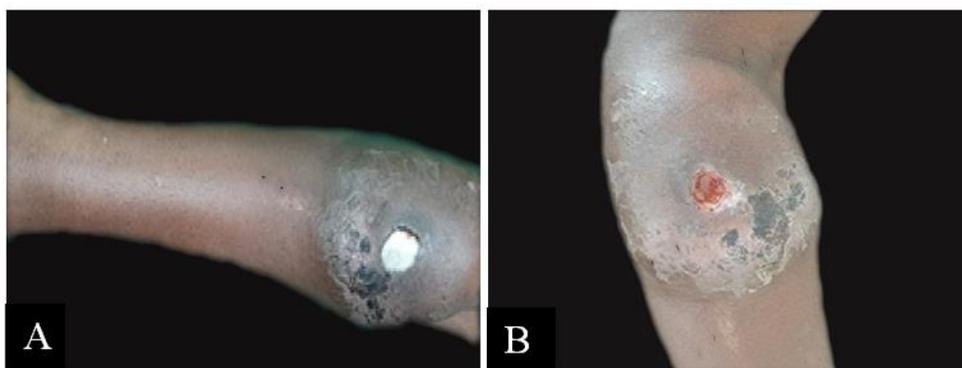
On examination, the patient was conscious, afebrile and his systemic examination was within normal limits. On local examination, a well-defined hard swelling of 8 x 7 cm was present on the pronator aspect of the right forearm, just below the elbow with an ulcer measuring 1.5 x 1.5 x 0.8 cm present over the surface. The skin over the swelling was erythematous. The swelling was fixed, non-mobile and there was no restriction in the movements of associated elbow. There was no enlargement of the regional lymph nodes or loss of sensation.

To proceed with the investigations, CBC was done which was normal. X-ray of the affected extremity showed no sign of osteolysis or a periosteal reaction. MRI of right forearm was also done which showed a well-defined, unencapsulated lesion of approximately 7.5 x 7 x 1.6 cms, which appeared to be iso to hypointense in T1 with associated skin ulceration and significant perilesional edema – likely a soft tissue sarcoma.

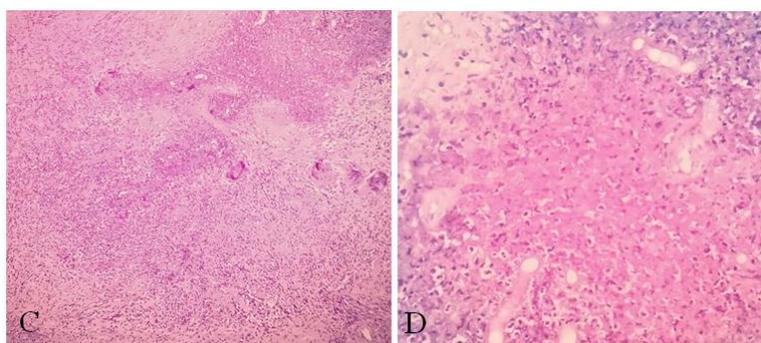
An incision biopsy was done, and we received four fragments of grey white to grey brown soft tissue bits altogether measuring 1.5 cms. Microscopic examination with hematoxylin and eosin revealed a granulomatous lesion with foci of necrosis comprising of few broad branching hyphae without septations,

suggesting a fungal origin- Mucormycosis. There was

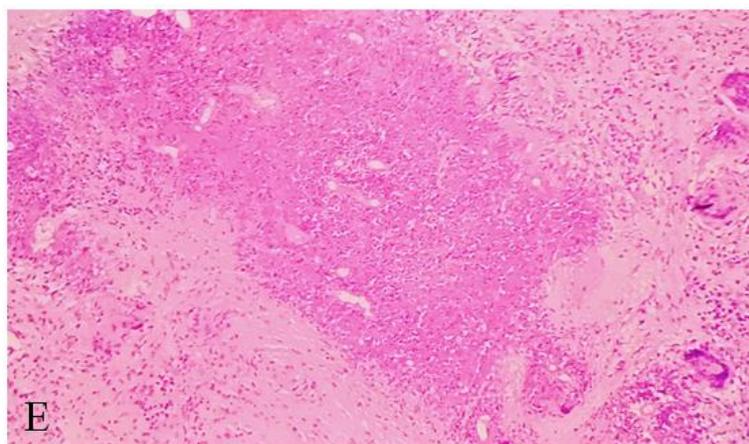
no evidence of dysplasia or carcinomatous change.



A & B - show clinical picture of the swelling in the pronator aspect of the forearm associated with an ulcer



C - 10x H&E shows a granulomatous lesion with foci of necrosis consisting of inflammatory cells and Langhans type of giant cell. **D** - 40x H&E shows necroinflammatory tissue consisting of fungus with broad branching hyphae



E - 10x H&E shows a granulomatous lesion with foci of necrosis consisting of inflammatory cells, Langhans type of giant cell and fungus with broad branching hyphae

DISCUSSION

Mucormycosis is an invasive fungal infection that could appear following trauma [3]. Other risk factors include diabetes, high dose of corticosteroids, neutropenia, iron overload and prematurity [4]. Though trauma can cause acceleration of the infective process in a healthy individual, it does more so when the person is immunocompromised, where the infection might extend into deep layers and cause disseminated infection [3]. The literatures states that a trivial trauma like the ones caused by needle prick for self-assessment of blood glucose by glucometer and thorn prick has resulted in

fatal disseminated disease [5, 6]. As with cutaneous mucormycosis, sporadic cases have been linked with contaminated bandages and adhesive dressings [7]. Trauma and contact with herbal combinations which possibly were contaminated could have put the patient under the risk of infection in our case.

The mean age of presentation is 38 years and is more common among males for reasons unknown [2]. Though the cutaneous form of the infection can affect any part of the body, upper and lower extremities are the most commonly involved sites as in our case [3].

The other common reported sites of infection include sinus (39%), pulmonary (24%) and skin (19%) [7].

Cutaneous mucormycosis can be classified into localized, deep and disseminated depending on the extent of involvement.² When only the skin or subcutaneous tissue is involved, it is called localized cutaneous mucormycosis, whereas involvement of muscle, tendon or bone warrants the use of term deep cutaneous mucormycosis and disseminated is used when there is involvement of non-contagious organs [2].

Clinical presentation of cutaneous mucormycosis greatly varies. The clinical course can be fulminating resulting in gangrene and hematogenous dissemination [5] or may also present as a gradual, slowly progressive lesion [8]. Necrotizing fasciitis and gangrene are other forms of presentation of the disease [3], where the patient initially presents with induration, pustules or necrotizing ulcer which may rapidly progress to abscess or necrosis of the underlying cutaneous or subcutaneous tissue [9]. In our case, the patient presented with a swelling in the upper extremity associated with an ulcer.

Diagnosis is mainly by histopathology and molecular diagnostic tests that are developed recently. Histopathology shows thick, hyaline, nonseptated and bifurcated hyphae along with necrosis and inflammatory cell infiltrate. Newer molecular diagnostic tests with real-time PCR target 18S ribosomal DNA and are highly specific with no cross reactivity with other filamentous fungi.

A multidisciplinary approach including surgical debridement and antifungal treatment with lipid-based amphotericin B is recommended for best outcome.

The mortality rate of cutaneous mucormycosis ranges from 4% in localized form to 94% in disseminated disease [7]. In a study by Roden *et al.*, 44% of cutaneous infection were complicated by deep extension or disseminated infection [10].

As the signs and symptoms in most of the cases are highly non-specific and radiologically mimics soft tissue sarcoma, an increased awareness and a suspicious eye is required for the early diagnosis and effective management of a case of mucormycosis.

CONCLUSION

Considering the fact that zygomycetes are capable of extending rapidly along the tissue planes, especially so when the individual has other associated co-morbidities, a high index of suspicion is required for prompt diagnosis and effective treatment of a patient

with an uncommon clinical finding as in our case, because delay in diagnosis and antifungal treatment can raise the mortality.

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