## Case Report

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# Isolated renal mucormycosis: a rarity

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#### **ABSTRACT**

Mucormycosis is a rare, opportunistic, highly aggressive, fungal infection primarily noted in immunocompromised individuals. Isolated involvement of any organ is rare in general. Isolated renal involvement is unusual, with only a few cases reported in literature to date. We present a rare case of isolated renal mucormycosis successfully managed with systemic anti-fungal therapy and nephron sparing surgery.

Keywords: Isolated renal mucormycosis, Immunocompromised host, Anti-fungal therapy

#### INTRODUCTION

Mucormycosis (MM), previously known as zygomycosis, are rare infections caused by fungus of the order Mucorales, which are characterized by the production of aseptate hyphae and asexual spores. These infections are opportunistic and are mainly noted in individuals with compromised immunity although of late few cases in immunocompetent individuals have also been reported. <sup>2-4</sup>

#### **CASE REPORT**

A 36 years old male presented to us with the complaints of bilateral ureteric colic associated with passage of whitish flakes in urine for 3 months. He had no history of fever, haematuria, lithuria, frequency of micturition, dysuria or any other urological complaint. The patient was diagnosed with psoriasis three years prior for which he was given a course of methotrexate. Four months back he had taken corticosteroid injections for the purpose of bodybuilding being a Kabaddi player. Three months previously he was diagnosed as suffering from pulmonary tuberculosis and was advised six months course of anti-tubercular therapy (ATT) (ethambutol, isoniazid, rifampicin and pyrazinamide).

Examination of the patient was unremarkable. He underwent a detailed laboratory and radiological evaluation. The ESR was 36 mm but other hematological and biochemical tests were normal. Urine cytology was negative for malignant cells. Montoux test was negative suggestive of an immunocompromised state. HIV test and workup for genito-urinary tuberculosis was negative. Chest X-ray and non-contrast CT-scan chest showed evidence of bilateral consolidation and cavitation suggestive of pulmonary tuberculosis. Contrast enhanced CT abdomen showed a 62x37x69mm encapsulated, exophytic mass arising from the posteroinferior aspect of right kidney with mixed fatty tissue (-44 to -56 HU) and enhancing solid component (27-46 HU) suggestive of angiomyolipoma, (Figure 1) left kidney appeared normal. MR Urography showed an exophytic mass lesion involving inferior pole of right kidney with evidence of macroscopic fat consistent with angiomyolipoma, left kidney showed a wedge-shaped lesion in inferior pole suggestive of an infarct. Histopathological examination of urinary cast showed fungal hyphae suggestive of mucormycosis. Fungal culture of urinary demonstrated no growth. Fine needle aspiration cytology of the right renal mass was done since it was suspected to be of inflammatory origin but was inconclusive. ENT examination ruled out a rhinosporidial involvement.

With a presumptive diagnosis of renal mucormycosis, the patient was started on liposomal amphotericin-B in a cumulative dose 2650 mg over four weeks. Patient was taken up for surgery two weeks after completing the full course of antifungal therapy with a plan to perform partial/total nephrectomy depending on the operative findings. Intra-operatively a 7x6 cm exophytic mass on posterolateral aspect of lower pole of right kidney was noted which could be enucleated in toto (Figure 2). Post-operative recovery was uneventful. On histopathological examination, a well encapsulated mass with aseptate fungal hyphae suggestive of mucormycosis and central necrosis was seen (Figure 3).

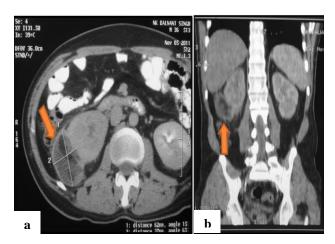


Figure 1 (a and b): Cross sectional images of patient showing 62x37x69 mm encapsulated, exophytic mass arising from posteroinferior aspect of right kidney.

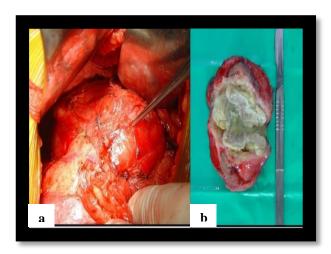


Figure 2 (a and b): Intra-operative findings; exophytic mass in posterolateral aspect of lower pole of right kidney being pointed by forceps, excised mass with central necrotic material.

On post-operative follow-up, patient was asymptomatic and had gained 12kg weight. X-ray chest after completion of ATT showed clear lung fields. Repeat contrast enhanced CT-scan abdomen showed no evidence of residual/ recurrent disease (Figure 4).

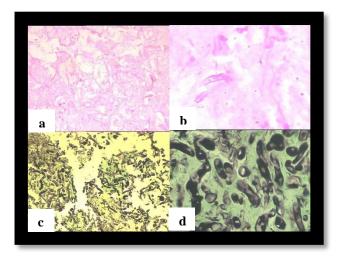


Figure 3 (a-d): H and E stain slides along with Grocott's methenamine stained slides (10X and 40X images) showing aseptate broad ribbon shaped hyphae in the background of inflammation and necrosis.

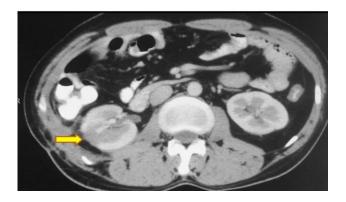


Figure 4: Follow up contrast enhanced CT-scan showing no evidence of residual/recurrent disease.

### **DISCUSSION**

Mucormycosis is an opportunistic fungal infection primarily caused by group Mucorales, most common isolate of which being *Rhizopus oryzae* 44%, followed by *Rhizopus microsporus* 22%, *Mucor circinelloides* 9.5% etc.<sup>3</sup> These fungus are ubiquitous in nature and the spores of them gain access into body via respiratory system, gastrointestinal tract or breach in the skin.<sup>5</sup> It is defect in function of macrophages and neutrophils especially when associated with acidosis and hyperglycemia, which help in proliferation of these infections.<sup>6</sup>

The various primary clinical forms of mucormycosis are rhino-cerebral (most common), pulmonary, cutaneous or gastrointestinal. Any of these primary forms can develop into disseminated form due to its high angio-invasive property. In disseminated form which accounts for 9% of total cases; the organ most commonly involved is the lung, with involvement of the kidneys being reported in up to 20% of cases. Isolated organ involvement is a rare

occurrence with only limited cases reported in literature.<sup>4</sup> Renal mucormycosis has been postulated to result from either haematogenous dissemination or via retrograde spread from lower urinary tract infection.<sup>9</sup> Pathologically mucormycosis is characterized by presence of extensive angioinvasion of both small and large arteries by hyphae with resultant vessel thrombosis and subsequent tissue necrosis.<sup>8</sup> Isolated renal mucormycosis usually presents with fever, flank pain, tenderness, gross haematuria, pyuria or renal failure in case of bilateral involvement.<sup>10</sup>

Antemortem diagnosis of MM is difficult due to variable clinical presentation and limitations in achieving a tissue diagnosis. A high index of suspicion and judicious use of radiological and laboratory investigations can help. Renal ultrasound with colour doppler, contrast enhanced CTscan/MRI commonly demonstrates enlargement of the kidneys, multiple low attenuations and non-enhancing areas in the parenchyma, reduced/ absent contrast excretion and perinephric collections. These imaging features has been described by various authors as 'diffuse patchy nephrogram'. Imaging not only helps in pointing towards the diagnosis but also helps in delineating the extension of the lesion. Urine analysis and biopsy can occasionally demonstrate the fungal hyphae which are characteristically wide (3-25 µm in diameters), ribbonlike, aseptate (pauciseptate), thin walled hyphae branching at right angles. It is recommended to perform both periodic acid Schiff (PAS) and Grocott's methenamine silver (GMS) stains as it is the poor staining of hyphae with the latter that often suggest MM.<sup>7</sup>

The mortality of different forms of mucormycosis reaches 75-100% in most series, with survival for isolated renal zygomycosis estimated to be 65%. The recommended management of mucormycosis is combination of aggressive surgical debridement and administration of systemic antifungal along with medical treatment and control of predisposing factors. Amphotericin-B is the first line drug of choice but in recent times Posaconazole as a substitute, especially as salvage therapy, is gaining popularity. Surgery is considered necessary due to the massive amount of tissue necrosis occurring in the disease process that may prevent entry of anti-fungal agents in adequate concentrations.

#### **CONCLUSION**

To conclude the key points of interest in our case are as follows; immunocompromised state predisposed to infection with two different organisms causing pulmonary tuberculosis and renal MM, isolated renal MM and presentation as a renal mass is extremely rare, mass being predominantly exophytic with involvement of perinephric fat, a mistaken diagnosis of angiomyolipoma on CT and MRI was made, microscopic examination of the urinary cast clinched the diagnosis and a course of systemic anti-fungal agent followed by excision of the fungal mass rather than nephrectomy as commonly described produced a satisfactory outcome.

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