Isolated Renal Mucormycosis in Immunocompetent Children: A Report of Two Cases

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Abstract

Isolated renal mucormycosis is a rare entity in children. It is potentially fatal when not detected and managed early with antifungal therapy, and surgery as and when needed. We present two immunocompetent children who developed this infection and subsequently succumbed to it. The diagnosis was established postmortem on renal biopsy specimens. We also discuss the 9 cases of isolated renal involvement in children published in literature.

Keywords: Mucormycosis, renal biopsy, zygomycosis

Introduction

Fungal infections are frequently missed and underdiagnosed in critically ill patients leading to significant morbidity and mortality. Among these, candidal infections predominate, followed by Aspergillus and Mucor. In a child with a history suggestive of urinary tract infection, features such as anuria and enlarged kidneys with the loss of vascularity on renal Doppler should alert the clinician and lead to further diagnostics. We present two apparently immunocompetent children who succumbed to a lethal fungal infection of the kidneys, mucormycosis, and suggest a clinical approach to manage such patients.

Case Reports

Case 1

A previously healthy 12-year-old boy presented with high fever, right flank pain, and oliguria of 5 days progressing to advanced renal failure. He underwent eight sessions of hemodialysis and received intravenous (IV) antibiotics. He continued to be febrile and anuric after 18 days when he developed respiratory distress and one episode of seizure and was referred to our hospital. On evaluation, the child (weight: 24 kg) was in respiratory failure and shock, for which intubation, mechanical ventilation, and vasopressors were initiated and he was shifted to Intensive Care Unit (ICU). Workup revealed neutrophilic leukocytosis, deranged renal function, hyperkalemia, hyperlactatemia, and metabolic acidosis. He was started on antimicrobials (piperacillin-tazobactam, levofloxacin, and metronidazole), antiepileptic phenytoin, and continued on hemodialysis. Ultrasound imaging of the abdomen revealed hepatosplenomegaly, moderate ascites, and bilateral bulky kidneys with heterogeneous echotexture and a suspicious abscess in the lower pole of the left kidney. A computed tomography (CT) of the head was normal. Serology for HIV, hepatitis B surface antigen (HBsAg), and hepatitis C virus (HCV) was negative. Renal Doppler showed no flow in both main renal arteries and noncontrast CT of the kidneys showed multiple hypodensities in both kidneys [Figure 1a] though renal vessels could not be commented upon (IV contrast was not used in view of deranged renal function). With a suspicion of fungal pyelonephritis, the patient was started on conventional amphotericin B. On day 4, he deteriorated further, and he succumbed to his illness on day 6 of his hospital stay. A postmortem kidney biopsy showed completely necrosed renal cortical parenchyma on light microscopy, with a few fungal elements with acute angle and broad-based hyphae, a picture suggestive of renal mucormycosis [Figure 1b].

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More recently, Nayagam et al. reported three cases with isolated renal involvement, of whom two were immunocompetent children, both of whom improved with surgery and antifungal therapy.\(^\text{[2]}\) More recently, Nayagam et al. reported an 18-month-old immunocompetent child with unilateral renal involvement, who improved after nephrectomy and had stable renal function.\(^\text{[3]}\) Previously described children with bilateral involvement [Table 1] have uniformly succumbed to their illness. The children described here are apparently immunocompetent and both developed bilateral pyelonephritis, secondary to mucormycosis, and worsened before any surgical intervention could be planned. To the best of our knowledge, these are the fourth and fifth cases of isolated renal mucormycosis in pediatric patients with bilateral involvement.

Both the patients were non-diabetic, non-neutropenic, and not on any immunosuppressive medications; prolonged hospitalization with broad-spectrum antibiotic usage and hemodialysis could have placed them at risk for nosocomially acquired mucormycosis. However, community-acquired infections have been described. While relatively uncommon in the literature from developed countries, isolated renal involvement has been described by Indian authors. It is unknown as to what factors, inherited or environmental, place apparently immunocompetent individuals at risk of renal mucormycosis, in the absence of dissemination.

In a child with a history suggestive of urinary tract infection, features such as flank pain, gross hematuria or pyuria, acute kidney injury (anuria), and enlarged kidneys on ultrasonography, a high index of suspicion is required to suspect angioinvasive fungus. Further workup is warranted in such children with renal ultrasound to demonstrate hypoechoic shadows or hypodensities and Doppler to show altered blood flow in renal vessels. CT abdomen reveals enlarged kidneys with poor enhancement, hypodensities (parenchymal abscess), emphysematous changes, and infarcts (features of angioinvasion). Urine culture for fungus and kidney biopsy revealing broad-based (ribbon like) aseptate fungal hyphae branching at right angles clinches the diagnosis. Early aggressive management with amphotericin (or antifungal with activity against molds) is desirable and nephrectomy or partial excision to remove the infected and necrotic tissue is often required.

To summarize, isolated renal mucormycosis is a rare fungal infection with a fatal outcome unless detected early and treated aggressively with antifungal and surgery, if required.

**DISCUSSION**

Mucormycosis is a rare fungal infection with high morbidity and mortality seen most often in immunocompromised hosts, especially in hematologic malignancies, transplant recipients, and diabetics. Other predisposing factors include IV drug abuse, deferoxamine therapy, burns, trauma, and malnutrition. Neutrophil or macrophage dysfunction, especially when associated with acidosis and hyperglycemia, predisposes to this infection. Isolated involvement of the kidneys by Mucorales is a rare entity in children. In a review of pediatric mucormycosis involving 187 patients, Roilides \textit{et al.} described the most common patterns of organ involvement — rhinocerebral (18%), cutaneous (27%), pulmonary (16%), and gastrointestinal (21%).\(^\text{[1]}\) In 2004, Jianhong \textit{et al.} reported three cases with isolated renal involvement, of whom two were immunocompetent children, both of whom improved with surgery and antifungal therapy.\(^\text{[2]}\)
Table 1: Cases of pediatric isolated renal mucormycosis published in literature

<table>
<thead>
<tr>
<th>Author/year</th>
<th>Age/sex</th>
<th>Predisposing factors</th>
<th>Unilateral/Bilateral</th>
<th>Management</th>
<th>Outcome</th>
</tr>
</thead>
<tbody>
<tr>
<td>Chugh et al., 1993[4]</td>
<td>17/male</td>
<td>None</td>
<td>Bilateral</td>
<td>Amphotericin B</td>
<td>Died</td>
</tr>
<tr>
<td>Gupta et al., 1999[5]</td>
<td>17/male</td>
<td>None</td>
<td>Bilateral</td>
<td>Amphotericin B</td>
<td>Died</td>
</tr>
<tr>
<td>Jianhong et al., 2004[2]</td>
<td>3 months/female</td>
<td>None</td>
<td>Unilateral</td>
<td>Amphotericin, partial excision</td>
<td>Survived</td>
</tr>
<tr>
<td>Jianhong et al., 2004[2]</td>
<td>12/male</td>
<td>None</td>
<td>Unilateral</td>
<td>Amphotericin B, drainage of renal abscess</td>
<td>Survived</td>
</tr>
<tr>
<td>Jianhong et al., 2004[2]</td>
<td>14/male</td>
<td>None</td>
<td>Unilateral</td>
<td>Amphotericin B, nephrectomy</td>
<td>Survived</td>
</tr>
<tr>
<td>Sharma et al., 2006[6]</td>
<td>14/male</td>
<td>Aplastic anemia/ATG</td>
<td>Unilateral</td>
<td>Amphotericin B, nephrectomy</td>
<td>Died</td>
</tr>
<tr>
<td>Dhua et al., 2012[8]</td>
<td>7/male</td>
<td>Post-pyeloplasty</td>
<td>Unilateral</td>
<td>Amphotericin B, nephrectomy</td>
<td>Survived</td>
</tr>
<tr>
<td>Sotbi et al., 2013[9]</td>
<td>4/female</td>
<td>None</td>
<td>Unilateral</td>
<td>Amphotericin B, nephrectomy</td>
<td>Survived</td>
</tr>
<tr>
<td>Our cases</td>
<td>12/male</td>
<td>None</td>
<td>Bilateral</td>
<td>Amphotericin B</td>
<td>Died</td>
</tr>
<tr>
<td>Our cases</td>
<td>10/male</td>
<td>None</td>
<td>Bilateral</td>
<td>Fluconazole</td>
<td>Died</td>
</tr>
</tbody>
</table>

ATG: Anti-thymocyte globulin

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Conflicts of interest

There are no conflicts of interest.

References