Nephroquiz for the Beginner  
(Section Editor: G. M. Zeier)  

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Acute renal infarct in a woman with slight asthma

A 54-year-old caucasian female fell ill while visiting Norway. She suffered from asthma and used inhalation corticosteroids, but apart from this she was in good health. After a 3-day history of fever and right flank pain she was admitted to hospital with a suspected upper urinary tract infection. On physical examination there was a remarkable tenderness in both flanks, most prominent on the right side, temperature was 38°C, the blood pressure was 126/80 mmHg, and the pulse was 84. The haemoglobin was 14.6 g/dl, the white-cell count was 16 700/mm³, the platelet count was 230 000/mm³, the erythrocyte sedimentation rate was 80 mm/h and the creatinine level was 72 µmol/l. An HIV test was negative. ECG showed sinus rhythm with no apparent pathology. A chest X-ray was unremarkable. Urine microscopy showed more than 20 leukocytes and 20 erythrocytes per high-power field, but no casts. Repeated urine and blood cultures revealed no growth. Ultrasound investigation showed an enlarged right kidney without signs of hydronephrosis.

Upper urinary tract infection was suspected, but the patient’s condition did not improve on antibiotics. A spiral computer tomography and renal angiography were performed, both showing a right kidney with almost no circulation and with the apparent occlusion of several centrally located branches of the right renal artery (Figures 1 and 2). There was also an underperfused segment of the upper part of the left kidney compatible with bilateral renal involvement. The patient’s diagnosis was re-evaluated and she was treated with subcutaneous low-molecular-weight heparin. Echocardiography was normal however, without evidence of intramural thrombi. All coagulation tests were also within normal limits and ANA, antinuclear antibodies and antineutrophil cytoplasmic antibodies (ANCA) were all negative. The patient continuously had a temperature between 38.0°C and 38.6°C and complained increasingly of pain in the upper right quadrant of the abdomen. A renal biopsy was considered, but not done because the patient had received anticoagulation. Under the suspicion of renal vasculitis she received a 3-day course of methylprednisolone, but without effect on her clinical condition. Eighteen days after her admission to the hospital ultrasound revealed a brim of fluid surrounding the right kidney. At that time she also had developed a striking induration of the skin overlying the right kidney.

Question

What diagnostic and therapeutic procedures would you propose at this point?

Fig. 1. Abdominal CT scan showing enlarged hypodense right kidney compatible with renal infarct. (Incidental finding: previously known liver cysts.)

Fig. 2. Renal angiography of the right kidney showing occlusion of several central branches of the renal arteries.
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A right nephrectomy was performed. Microscopic investigation showed large areas of acute infarction of the kidney and surrounding fatty tissue with infiltration of broad and polymorphic hyphae compatible with mucormycosis (Figure 3). Subsequent fungal cultures showed growth of *Saksenaea vasiformis*. A few days after the operation the patient became critically ill and was treated in an ICU with lipid-based amphotericin B and continuous haemodialysis. After 6 weeks she was well enough to undergo left nephrectomy. This kidney also showed mucormycosis and areas of extensive acute infarction. No other organs than the kidneys seemed involved, and after both kidneys and surrounding necrotic tissue were removed, the patient’s condition slowly improved. One year after the kidneys were removed, she receives intermittent haemodialysis and is awaiting a kidney transplant.

Discussion

Mucormycosis is an opportunistic infection caused by fungi belonging to the order Mucorales. These are ubiquitous fungi of low virulence, but when the disease is invasive the fungi have the capacity to invade vessel walls causing thrombosis and infarction [1]. Infection with these agents is well known and feared in immunocompromised patients [2–4] and can also be seen in patients with diabetes mellitus or in patients treated with corticosteroids [3,5]. In most cases the portals of entry are the nasal sinuses or the lungs. The brain is most often affected in the localized form of the disease, while lungs, heart, liver, spleen, and kidneys are the most often affected organs in the disseminated form [1,3]. In the biopsy, hyphae can be confused with capillaries, but the diagnosis is usually straightforward if one is familiar with the appearance of the fungus. The hyphae are seen in ordinary H&E-stained slides. Cultivation is necessary to identify the particular fungus.

There have been only a few reports of mucormycosis in non-immunocompromised patients and of patients with localized disease in the kidneys [6–8]. The only known predisposing factor in our patient was the use of inhalation steroids. Although this should not lead to a general decrease in host defence, the effect of corticosteroids on the airways could hypothetically reduce the local resistance to micro-organisms. There was however, no evidence of a lung infection.

An enlarged kidney is not a typical feature of renal embolism or renal vasculitic disease, but would speak in favour of neoplastic disease, infections, or storage disorders (amyloid). However, tumours are likely to be focal and often give an asymmetric enlargement of the kidney, and the clinical course in this patient was not suggestive of amyloidosis.

Treatment of localized mucormycosis is combined surgical excision and long-term anti-fungal therapy [3,9]. Amphotericin B at increased dosages is the drug of choice. Lipid-based formulations of amphotericin B are usually better tolerated, but have not been shown to be clearly superior to conventional deoxycholate amphotericin B. The mortality of mucormycosis is high, with a reported survival of only 36% [9], and in a series of 21 patients with isolated renal mucormycosis the mortality was 52% [8]. To our knowledge there are no previous reports in the literature of successful treatment of bilateral renal mucormycosis.

Using an ordinary biopsy needle (gauge 16) we drew 20 biopsies from different parts of the removed right kidney; all biopsies showing hyphae. Although the biopsies were taken from a single massively infiltrated kidney, it supports the idea that renal biopsy may be useful in cases of suspected mucormycosis and emphasizes the importance of performing a biopsy in patients with unclear renal conditions.

Suggested reading


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Fig. 3. Necrotic perirenal fatty tissue with infiltration of broad polymorphic hyphae compatible with mucormycosis (H&E-saffron original ×400).