Pseudotumours: an atypical presentation of renal sarcoidosis

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Learning Point for Clinicians

Sarcoidosis may present as bilateral kidney pseudotumours. Clinical presentation of this form of renal sarcoidosis is quite unspecific and is usually associated with normal renal function. Its diagnosis relies on imagery [computed tomography (CT) scan] and kidney histology. The literature and the present case suggest the efficiency of corticosteroids.

Case report

A 32-year-old male was referred to our department for bilateral kidney tumours. Past medical history included sarcoidosis with eyes and lung involvement that was diagnosed 10 years ago and successfully treated with oral corticosteroids for 3 months. Afterwards, he remained asymptomatic until recently when he developed a right renal colic. A CT scan revealed the presence of bilateral renal masses (Figure 1A). Infracentimetric retroperitoneal lymph nodes were detected and their association with the kidney tumours led to suspect the diagnosis of lymphomatous disease and the performance of biopsy of the kidney tumours. The histological analysis showed a granulomatous interstitial nephritis with typical multinucleated giant cells and a surrounding lymphocytic infiltrate. The interstitium was largely replaced by both non-caseating granulomas and an extensive interstitial fibrosis. The immunofluorescence analysis was unremarkable.

The patient was then referred to our department with the hypothesis of renal sarcoidosis. On admission, he was complaining of effort dyspnoea. Physical examination was unremarkable. He had no peripheral lymphadenopathies. The 6-min walk test demonstrated oxygen desaturation at 92%.

Laboratory tests showed serum creatinine of 103 µmol/l (modification of diet in renal disease estimated glomerular filtration rate 70 ml/min/1.73 m²). Calcemia was in normal range. The hemogram was normal, and screens for monoclonal gammopathy and auto-antibodies were negative. His urinary sediment was unremarkable, and proteinuria was in normal range (0.12 g/day). Finally, the glomerular filtration rate measured with chromium-51 EDTA was normal (107 ml/min/1.73 m²). The serum angiotensin-converting enzyme level was elevated (94 UI/l, normal range: 5–60 UI/l).

Chest CT scan showed mediastinal and hilar lymphadenopathies with very mild interstitial pneumo-

nia. Pulmonary function tests showed a mild CO diffusion impairment and the echocardiogram was normal. A F(18)-FDG positron emission tomography (PET)/CT was done and showed a bilateral hypermetabolic signal (SUV max = 6) of the renal pseudotumours (Figure 1B). Finally, the diagnosis of sarcoidosis with bilateral pseudotumoral kidney involvement was retained.
The patient was started on oral corticosteroids (prednisone 1 mg/kg/day) with a progressive tapering to prednisone 5 mg/day over 4 months. A control CT done 6 months later showed a complete regression of the renal tumours. After 1 year of follow-up, no relapse of the tumours has been observed under a maintenance regimen of prednisone 5 mg/day.

Discussion

Renal manifestations of sarcoidosis are polymorphous and have variable prevalence (3–23% of patients). Nephrocalcinosis and/or nephrolithiasis secondary to hypercalcemia and hypercalcuria induced by vitamin D hypersensitivity are the most frequent renal presentations. Less frequently, renal involvement may include interstitial nephritis with or without non-caseating granulomas and exceptionally glomerulonephritis. Renal failure is usually the clinical manifestation that leads to suspect the renal localization of the disease. In such cases, a kidney biopsy is often done allowing definite diagnosis. Corticosteroids are the cornerstone of treatment of these parenchymal injuries allowing frequently either complete renal recovery or significant improvement of renal function.

Pseudotumours are a rare presentation of sarcoidosis and only few cases in the literature report renal pseudotumours with clinical presentation, management and outcome of this form still largely unknown. In our patient, the presentation was quite insidious as only transient right renal colics led to perform a CT scan showing the bilateral pseudotumours. Lymphoma with lymphomatous infiltration of both kidneys was first considered because of the associated hilar, mediastinal and intra-abdominal lymphadenopathies. Sarcoidosis was retained based on past medical history, tumour histology showing typical non-caseating granulomas and elevated serum angiotensin-converting enzyme level. In opposition to the classical presentation of renal sarcoidosis, our patient had normal renal function without proteinuria and normal serum calcium level. After reviewing the English literature, only six other cases of sarcoidosis with kidney pseudotumours have been reported.

Based on these cases, the clinical presentation is often polymorphous and unspecific. Pseudotumours are often a fortuitous discovery at CT scan done for disease evaluation. In four of the six previous cases, corticosteroids were used with success allowing tumour size reduction in three cases, suggesting a steroid sensibility of pseudotumours. However, little is known about management of corticosteroids and renal function evolution, as well as long-term outcome in patients with this form of sarcoidosis. In our case, the pseudotumours involved both kidneys at ~30–50% of overall kidney parenchyma. Given the presence of an extensive interstitial fibrosis at tumour biopsy, we thought that without treatment the disease would have spread to the intact renal parenchyma and finally result in renal failure.

As a conclusion, sarcoidosis may present as pseudotumours of kidneys. Our case report and the literature review suggest the efficiency of corticosteroids for treatment of such condition.
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References