Case Report

Sarcoid granulomatous interstitial nephritis and sarcoid abdominal aortic aneurysms

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Introduction

Sarcoidosis is a systemic granulomatous disorder of unknown etiology, characterized by chronic non-caseating epitheloid granulomatous inflammation with tissue destruction [1,2]. Renal involvement affects ~20% of patients with sarcoidosis [1,2] and can be found in patients with no other localizations of the disease [3]. A common cause of renal dysfunction is hypercalcaemia and hypercalciuria leading to nephrocalcinosis [2]. Granulomatous interstitial nephritis (GIN) is also a cause of renal dysfunction, in which the clinical picture and laboratory evidence of tubular defects point to tubulo-interstitial nephritis [4]. Sarcoidosis is a systemic disease, affecting many organs. However, large vessel involvement such as aortic aneurysms due to sarcoidosis are rare, and only a few papers have reported aortic aneurysms complicating sarcoidosis [5–7]. We report a case of renal sarcoidosis complicated with saccular abdominal aortic aneurysms, confirmed by histology of the surgically resected aortic wall.

Case

A 60-year-old man was referred to our hospital because of renal dysfunction and urinary abnormalities. Although he had no respiratory complaints, an abnormal shadow had been identified on a chest X-ray 10 years earlier in another hospital, but there had been no further work-up. Since that time, he had not suffered from any particular ailment. On admission, he was in good clinical condition. His blood pressure was 116/70 mmHg. Physical examination found no superficial lymph nodes. There was mild hepatosplenomegaly with liver function disturbances. The arterial pulses were found in both lower limbs. Urinalysis showed proteinuria (1+), occult blood (1+) and no significant casts. Proteinuria was 0.44 g/24 h. However, markers of tubulointerstitial damage were very high (β2-microglobulin 123 732 μg/24 h, α1-macroglobulin 34.67 mg/24 h). Serum creatinine level was 127 μmol/l and the creatinine clearance was 39.5 ml/min/1.73 m². Serum electrolytes were: sodium 135 mmol/l, potassium 4.3 mmol/l, chloride 100 mmol/l and bicarbonate 21.5 mmol/l). Serum calcium level was 2.25 mmol/l and phosphorus 1.5 mmol/l. C-reactive protein (CRP) and the erythrocyte sedimentation rate (ESR) indicated some degree of inflammation (CRP 0.8 mg/dl, ESR 50 mm/h). Immunoglobulin and complement levels were normal. Anti-nuclear antibodies were negative. The search for anti-neutrophil cytoplasmic antibodies was negative. The viral titres for B and C hepatitis were negative. The sodium excretion fraction was 2%, suggesting a renal tubulointerstitial disorder. Renal biopsy yielded eight normal glomeruli. Non-caseating tubulointerstitial granulomatous lesions were accompanied by marked tubular atrophy (Figure 1a). High-power microscopy showed a multinucleated giant cell with Schaumann bodies and inflammatory cell infiltration (Figure 1b). The proximal tubule basement membranes were destroyed and tubular epithelial cells detached. Immunofluorescence was negative for IgG, IgA, IgM, fibrinogen, C3, C4 and Clq in the glomeruli and tubules. The serum angiotensin-converting enzyme level was high (34.4 IU/l; normal range 8.3–21.4 IU/l). Tuberculosis skin tests were negative. Serum calcitriol levels were normal (46 pg/ml;
normal range 20–60 pg/ml). There was no evidence of infectious disease, no history of drug use and no uveitis. Cardiac ultrasonography was normal. Non-sustained ventricular tachycardia was found by Holter 24 h electrocardiography. The chest plain radiographs and chest computed tomography were normal. However, there was a significant uptake of 67-Gallium ($^{67}$Ga) in the lungs. Collectively these data were consistent with the diagnosis of systemic sarcoidosis.

The rest of the work-up comprised angiographies that led to the discovery of two abdominal aortic aneurysms. They were saccular and located between the renal artery and the bifurcation of the common iliac artery. Their diameters were 3.5 and 2 cm, respectively. Considering the high risk of rupture of saccular aneurysms, prosthetic replacement surgery was performed before the initiation of steroid therapy. The wall of the resected aneurysm contained granulomas with typical multinucleated giant cells (Figure 2a and b), and the same lesions were found in a neighbouring lymph node (Figure 2c). Two months after recovery, the patient was started on oral prednisolone at a dose of 30 mg/day. At that time, the serum creatinine had risen to 310 $\mu$mol/l. Following 2 months of corticosteroid treatment, serum creatinine had decreased to 97 $\mu$mol/l. The urinary markers of tubulointerstitial disorder had also decreased ($\beta_2$-migroglobulin from 123 732 to 8000 $\mu$g/day), without reaching the normal range.
**Discussion**

Renal involvement complicated with abdominal aortic aneurysm due to sarcoidosis is exceptional. The wall of an aneurysm caused by sarcoidosis is fragile and tends to rupture, as noted in a previous report [7]. In this report, the surgically resected aneurysm contained a granulomatous lesion with multinucleated giant cells in the arterial wall. In our case, we also detected granulomata in the partially resected aneurysm. The aneurysm wall was very friable, suggesting a sarcoid-related inflammation. Considering the rarity of saccular sarcoid aortic aneurysms, it is not surprising that treatment options are not codified, especially regarding the timing of corticosteroid therapy vs vascular surgery. We deemed that surgery should precede steroids for two reasons: (i) considering that saccular aneurysms tend to rupture more readily than the fusiform type; and (ii) because we feared that anti-inflammatory therapy might weaken the aneurysmal wall, and foster rupture.

There are few reports of silent renal failure induced by sarcoidosis [3]. In our case, there were target organ involvement and laboratory abnormalities compatible with the diagnosis: (i) non-sustained ventricular tachycardia; (ii) mild hepatosplenomegaly with liver function disorder; (iii) GIN; (iv) elevated serum angiotensin-converting enzyme level; (v) negative tuberculosis tests; and (vi) high pulmonary $^{67}$Ga uptake. There was no plausible evidence of another cause of disseminated granulomatous disease.

In conclusion, we report on a patient with renal sarcoidosis found by renal biopsy and complicated with abdominal aortic aneurysms that were discovered by chance. Aneurysm surgery was performed and renal dysfunction was improved by corticosteroid therapy. Aortic aneurysms along with renal involvement in sarcoidosis form an exceptional morbid association with a high inherent risk of death by aneurysm rupture if corticosteroid therapy is started before vascular surgery. We therefore suggest that aortic imaging, at least by ultrasonography, should be performed in the case of systemic sarcoidosis.

**Conflict of interest statement.** None declared.

**References**


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