A particularly private pain

Case

A 55-year-old man presented to his physician because of arthralgia involving the elbows, shoulders, knees, and ankles of 2 years’ duration. Occasionally, he noted frank arthritis of these joints with swelling and erythema. He denied skin manifestations or sun light intolerance. However, his physician had noted several weeks earlier that an anaemia was present. On physical examination he had a blood pressure of 160/80 mmHg. His eyes, mucous membranes, skin and joints were not remarkable. Full range of motion was preserved. His cardiovascular examination disclosed a systolic ejection murmur along the left sternal border. No oedema was present. However, the patient had evidence of glomerulonephritis on the basis of urinalysis and protein excretion. A procedure was performed, diagnosis established, and treatment with prednisone and sulfasalazine was begun by the patient’s physician.

One month later, the patient presented to the urology service with sudden scrotal pain and swelling. Elevation of the scrotal contents did not relieve the pain. The left testis was enlarged and painful. An emergency operation was performed. Postoperatively, the patient was referred to the nephrology service for further evaluation.

Laboratory and pathology

A renal biopsy was performed elsewhere but examined by our pathology department. This biopsy showed a severe proliferative glomerulonephritis with extracapillary proliferation and tubulointerstitial changes (Figure 1a). Immunofluorescence showed immune complex disease (Figure 1b). The patient’s physician had diagnosed systemic lupus erythematosus. At operation 1 month later, the non-viable appearing, swollen testis was removed. Histological section showed that the artery to the testis was acutely and completely thrombosed (Figure 2a). Another vessel was observed showing an older, organized thrombus (Figure 2b).

We first saw the patient after his unilateral orchiectomy. The haemoglobin was 5.5 mmol/l, the haematocrit was 26 vol%. Platelets were slightly

Fig. 1. (a) Light photomicrograph showing severe proliferative glomerulonephritis with extracapillary proliferation and interstitial nephritis. (b) Immunofluorescence showing granular deposits along the basement membrane.
reduced at 200 000 µ³ while the white blood cell count was normal. The Coombs test was negative, the total protein was 59 g/l, albumin 30 g/l, and the cholesterol 7.3 mmol/l. Urinary sediment revealed erythrocytes, leukocytes, red cell and granulated casts. The protein excretion was 3.5 g/day. The C3 concentration was reduced to half normal values while the C4 concentration was also reduced at 0.06 g/l. Antinuclear antibodies were increased at a titre of 1 : 640. Double-strand DNA antibodies were markedly positive, while antibodies against SS-A (Ro) were also elevated. Anticardiolipin antibodies for IgG were also elevated. Echocardiography disclosed thickened mitral leaflets.

**Question**

What is your diagnosis?
The pathological diagnosis was systemic lupus erythematosus and anticardiolipin antibody syndrome with thrombosis of a testicular artery. Therapy was begun with prednisone, cyclophosphamide, and then phenprocoumone. The common causes of acute scrotal pain are orchiitis, epididymitis, and testicular torsion. Less common are tumours, thromboses, and infarctions [1]. Thromboses of testicular arteries have been described in the literature in patients from various causes, including protein S deficiency [2], embolism from non-bacterial endocarditis in the course of Wegener’s granulomatosis [3], and also in systemic lupus erythematosus [4].

This patient’s renal biopsy was consistent with florid systemic lupus erythematosus. His serological studies verified the presence of anticardiolipin antibody syndrome, a fact that had not been appreciated at his initial evaluation. Patients with anticardiolipin antibody syndrome are at risk for recurrent arterial and venous thrombosis. In this patient, the thrombosis unfortunately involved a testicle resulting in a rare, but by no means unknown, cause for scrotal pain. A severity of systemic lupus erythematosus to this degree warrants agressive treatment with prednisone and cyclophosphamide, in addition to exquisite blood pressure management. The presence of anticardiolipin antibody syndrome in such patients should be addressed with oral anticoagulation, in this case phenprocoumone, sufficient to increase the INR to 2.5–3.0 [5].

References


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