Priapism related to an antiphospholipid syndrome in a patient with systemic lupus erythematosus

Sirs,

Priapism is persistent penile erection not associated with sexual stimuli. Its pathophysiology remains unclear but is partially related to cavernous occlusion, unregulated autoregulation into the penis and autonomic dysregulation (1). An antiphospholipid syndrome (APS) is defined as the association of a thrombotic event and the presence of an antiphospholipid antibody. Priapism has never been reported as a clinical manifestation of APS. The case we reported occurred after the withdrawal of antivitamin K treatment from a patient with a history of APS secondary to systemic lupus erythematosus (SLE).

A 39 year-old man was admitted on the first of February 2004 for priapism. Since 1991, he had suffered SLE, diagnosed in the presence of auto-immune hemolytic anemia associated with anti-cardiolipin and antiphospholipid antibodies. In 1996, he developed a right kidney infarction related to a lupus anticoagulant and antivitamin K treatment was started. In 2003, the patient presented with a nephritic syndrome related to an extra- membranous glomerulonephropathy. Treatment with azathioprine (100mg/dj) and prednisolone 1mg/kg was associated with its previous treatment (hydroxychloroquine, candesartan-cilexetil, acenocoumarol). The treatment stabilized proteinuria at between 2 and 3g/day, albuminemia at around 22 g/L, creatininemia at between 75 and 95 µmol/L. On the first of February 2004 at noon, the patient presented with priapism in the Urology department. INR was 1. The medical history included intracorporeal injection of Spongilium. The patient was discharged on day 8 treated with anti-coagulant and prolonged antivitamin K therapy (INR > 3). One month later, recovery was complete, detumescence persisted and fibrosis of corpora cavernosa was developed, INR was 2.54. In September 2005, lupus was still quiet, hemoglobinemia was 12.7 g/dl, platelet count 283000/ml and creatininemia was 1.4. In 2006, we reoccured. Arterial blood was extracted manually but detumescence was not achieved. The patient was discharged on day 8 with a persistent painless semi-penile erection treated with cyproterone acetate and prolonged antivitamin K therapy. As INR was 2.5, the patient was discharged on day 8 with a persistent painless semi-penile erection treated with cyproterone acetate and prolonged antivitamin K therapy.

In conclusion, our observation adds a new clinical manifestation to APS, and suggests that cases of unexplained priapism should be tested for APS.

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References