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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 venous outflow occlusion, unregulated overflow into the penis and autonomic dysregulation (1). 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 auto-immune thrombocytopenia. 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 nephrotic syndrome related to an extramembranous glomerulonephritis. Treatment with azathioprin (100mg/dj) and prednisolone 1mg/kg was associated with his previous treatment (hydroxychroloquine, 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 treatment (intracorporal injection of 5mg of Ephedrin, renewed) was ineffective and the patient therefore underwent surgery at 2 pm. These surgical procedures were also ineffective and detumescence was finally obtained by bilateral cavernous spongiosal shunting. Ten hours after, penile erection 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 (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 92 µmol/l. Proteinuria varied between 1 and 1.25 g/24 hours, and the INR was 2.7.

This is the first case report of priapism related to an antiphospholipid syndrome in a SLE patient. There are many possible causes of priapism (1). The most common are medications. Other common causes are pelvic tumors (including prostate adenoma), spinal cord damages, pelvic arterial-venous shunt, hematological disorders for example polyglobulia, thrombocythemia or venous thromboembolism. Some causes are less common : toxic causes (marijuana, cocaine, alcohol), amyloid, intravenous hyperlipidic nutrition and Fabry disease. The occurrence of priapism in systemic diseases is exceptional, but has nevertheless been reported during Behçet disease (2), Henoch-Schönlein purpura (3), Crohn's disease and ulcerative colitis (4) and Kawasaki disease (5). The only case reported in a SLE patient was described during a nephrotic syndrome associated with SLE in a 29 year-old patient (6). During nephrotic syndrome, the risk of developing venous thrombosis is elevated and inversely related to the antithrombin III level (antithrombin III is excessively cleared by the kidney) (7). In our case, the nephrotic syndrome was moderate when the priapism started (proteinuria 2.64 g/24 hours and albuminemia 25 g/l), and there was an evident relationship between the withdrawal of antivitamin K treatment 2 days before, the normalisation of INR and the occurrence of a priapism related to a thrombosis of cavernous spongiosal. APS usually requires an INR above 3 to avoid recurrences (8) and some authors (9) proposed to target INR between 2.5 and 3 in APS patients without other risk factors for thrombosis.

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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