AUTOIMMUNE CONNECTIVE TISSUE DISEASES

CRYOGLOBULINEMIC VASCULITIS WITH RENAL INVOLVEMENT AND SJöGREN'S SYNDROME.

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Background: A 67-year-old female presented a painful and pruritic dermatosis on lower limbs that appeared after prolonged standing, for 3 years, after the initiation of anticoagulant regimen with acenocoumarol. The anticoagulant scheme had been rotated to warfarin and then to apixaban without improvement of the disease. She had a history of portal cavernomatosis due to protein C deficiency, portal hypertension, deep vein thrombosis and recurrent thromboflebitis.

Observation: Physical examination found round violaceus macules with discrete margins, 3 cm in diameter in lower limbs. Diascopy was negative.
Serum creatinine was 0.7mg/dL, urine sediment contained 15-20 erythrocytes per high-power field, and 24 hs proteinuria was 1.3 grams. ANA was positive (1/160), anti-SSA antibody was >150 and rheumatoid factor was 876 U/m. dsDNA, Sm and ANCA antibodies where negative. C3 was 61 mg/dL, C4 1 mg/dL, and CH50 <10 mg/dL. Cryoglobulins tested positive on three different occasions. Serum protein electrophoresis was normal. Anti-HBV, HCV and HIV antibodies were negative. PCR testing for HCV was negative.
Left leg skin specimen exhibited leukocytoclastic vasculitis of small vessels. DIF revealed granular deposits of IgG, C3 and fibrinogen.
Renal biopsy evidenced type I membranoproliferative glomerulonephritis with 30% of sclerosis and 20% of wafers.
The patient referred arthralgia and xerostomia, so a minor salivary gland biopsy was performed that was compatible with Sjögren's syndrome.
We conclude this case as cryoglobulinemic vasculitis with renal involvement associated with Sjögren's syndrome.
Meprednisone 40 mg and azathioprine 150 mg was initiated. Furosemide, irbesartan and enalapril were indicated by renal involvement. She presented complete remission of dermatologic lesions. Renal function remained normal but proteinuria persisted, reason for which she remained follow-up by nephrology.

Key message: We highlight the value of a complete examination in any patient presenting with cutaneous vasculitis, always trying to discard systemic affection and trying to diagnose

associated diseases.