Clinical picture

Scleritis and retinal vasculitis associated with a type II mixed cryoglobulinemia

The patient, a 64-year-old man, with a past history of an IgM monoclonal gammopathy of undetermined significance (MGUS), presented with a 5-day history of ocular pain and redness in his left eye. The best-corrected visual acuity was conserved at 25/25 in both eyes. Topical instillation of phenylephrine did not blanch leading to the diagnosis of temporal anterior non-necrotizing scleritis (Figure 1a). Fluorescein fundus angiograms revealed a bilateral temporal peripheral retinal vasculitis (white arrows) (Figure 1b). Apart from the ophthalmic symptoms, clinical examination remained normal. Erythrocyte sedimentation rate was 2 mm/h. Interferon-gamma release assay (Quantiferon®-TB Gold-in-Tube, Cellestis) was negative. Screening for antineutrophil cytoplasmic antibodies, rheumatoid factor yielded negative findings. Transient antinuclear antibodies without specificity (controlled negative) and a type II mixed cryoglobulinemia with a monoclonal IgM kappa and polyclonal immunoglobulins were found non-associated with hepatitis C virus. No other complication of this cryoglobulinemia was found with a normal creatinine clearance without proteinuria. Bone marrow biopsy was performed, without plasmocytosis, confirming the MGUS diagnosis.

About one-half of patients with scleritis have an underlying disease, mostly rheumatoid arthritis or vasculitis, such as granulomatosis with polyangiitis. Cryoglobulins, immunoglobulins that reversibly precipitate at temperatures below 37°C, lead to vasculitis affecting small vessels. Skin, glomeruli and peripheral nerves are often involved. However, ophthalmologic presentations, affecting both anterior and/or posterior segment, are rare. A decrease temperature of the cornea from the core temperature has been proposed to explain the cryoprecipitation in the anterior segment associated with a deposition of circulating immune complexes. After a first course of oral non-steroidal anti-inflammatory drugs and topical dexamethasone, his ophthalmic condition worsened with an infero-temporal peripheral ulcerative keratitis leading to the use of high doses of topical dexamethasone and artificial tears with a favorable outcome at follow-up 3 months later.

Figure 1. (a) Scleritis of the left eye after instillation of topical phenylephrine. (b) Retinal temporal vasculitis (white arrow) on the fluorescein fundus angiogram (left eye).
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References