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NECROTIZING SCLERITIS AS ONSET PRESENTATRIONM IN ANCA-ASSOCIATED VASCULITIS PATIENT

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BACKGROUND

Granulomatosis with polyangiitis (GPA) is a rare autoimmune vasculitis characterized by granulomatous inflammation and necrotizing vasculitis, combining vascular wall inflammation and peri- and extravascular granulomatosis. One of the uncommon manifestations of GPA is necrotizing scleritis. This is a severe painful inflammatory condition characterized by edema and cellular infiltration of the sclera and episclera. We will report a case of a patient with vasculitis who developed severe necrotizing scleritis.

CASE REPORT

A 61-year-old man presented with a lesion in the palpebral region of the left eye, which had evolved over 2 months. subsequently, he presented pain, hyperemia and abundant secretion in the same eye. initially, topical treatments were performed with ciprofloxacin eye drops and corticosteroids, and systemic antibiotic treatments with antibiotics cephalexin,trimethoprim/sulfamethoxazole. No improvement after treatment. Serology for HIV, viral hepatitis and syphilis showed negative results. Diagnostic investigation continued with autoantibodies and markers for systemic rheumatologic diseases: rheumatoid factor (RF), antinuclear antibody (ANA), peripheral antineutrophil cytoplasmic antibodies (P-ANCA) and C3, C4, with negative results. However, the antineutrophil cytoplasmic antibody (C-ANCA) test was reactive. The positive marker (C-ANCA) associated with the clinical manifestations and the anatomopathological examination, allowed to confirm the diagnostic hypothesis of necrotizing scleritis by vasculitis C- ANCA. Was initiated immunosuppressive therapy with azathioprine with partial improvement of the condition however after 3 presented increase of transaminases, with need of suspension of the treatment. an orbital resonance imaging was performed showing an expansive lesion suggestive of orbital inflammatory pseudotumor. due to the set of manifestations, it was programmed to start pulse therapy with monthly cyclophosphamide for 6 months.

CONCLUSION

Antineutrophil cytoplasmic autoantibody disease has an unknown etiology and diagnosis is based on recognition of the clinical picture and detection of serum antineutrophil cytoplasmic agents (ANCAs). We report the case of a patient with this disease who presented with ophthalmic lesions untreated with antibacterial and corticosteroid eye drops, and immunosuppressants. This case is relevant because it is a severe form of a disease that presents a complex clinical reasoning.

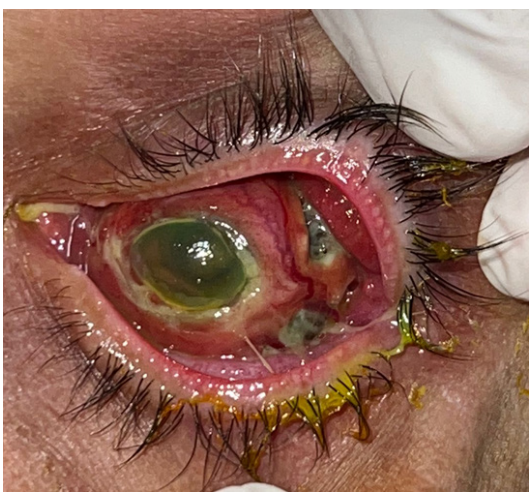

Figure: Diffusely hyperemic conjunctiva with fibrin depositio