Granulomatosis with polyangiitis patient with retinitis as an initial manifestation

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PURPOSE:

To describe a case of retinitis as the initial manifestation of granulomatosis with polyangiitis (GPA).

METHODS:

Case report.

RESULTS:

A 51-year-old male patient presented to the clinic complaining of hyperemia and visual loss in the right eye for 1 week.

OPHTHALMOLOGICAL EXAMINATION:

VISUAL ACUITY

OD: 20/30p
OS: 20/20

BIOMICROSCOPY

OD: Biomicroscopy demonstrated mild conjunctival hyperemia and anterior chamber reaction

FUNDOSCOPY

OD: Demonstred vascular thinning 1+/4+, serous retinal detachment with yellowish subretinal infiltrates in the superior temporal arcade and temporal periphery, not affecting the fovea (Figures 1 and 2)

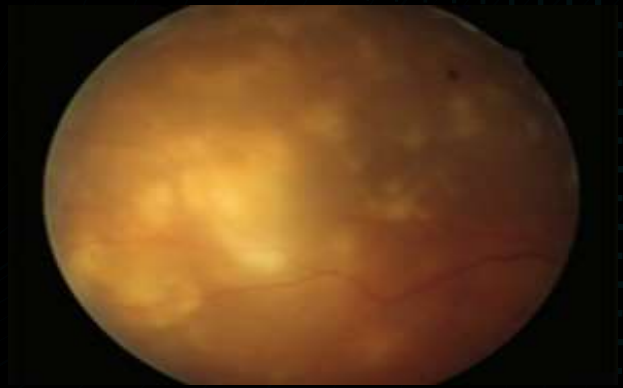

Figure 1: Right eye fundus photograph (superior temporal)

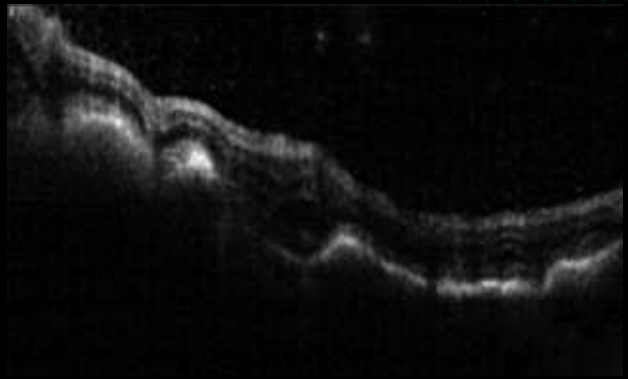

Figure 2: Right eye optical coherence tomography

Considering posterior uveitis as a possible diagnosis, routine examinations were requested.

Examinations for syphilis, HIV, CMV, herpes, tuberculosis, toxoplasmosis, rheumatoid factor, and antineutrophil cytoplasmic antibodies were all negative.

The patient was seen by a rheumatologist and it was confirmed that PR3 antiproteinase antibody was reactive.

SYSTEMIC INVESTIGATION

Showed renal and respiratory changes, with pulmonary lesions on chest tomography.

The diagnostic hypothesis was of GPA with “lung–kidney syndrome.”

TREATMENT

Treatment with oral corticosteroids(40 mg/day) began and after 10 days, the patient developed necrotizing anterior scleritis in the same eye.

Therefore, treatment was modified to cyclophosphamide 200 mg per day, after discussion with the rheumatologist. On the 20th day, the patient showed significant improvement in subretinal infiltrates and scleritis (Figures 3 and 4).

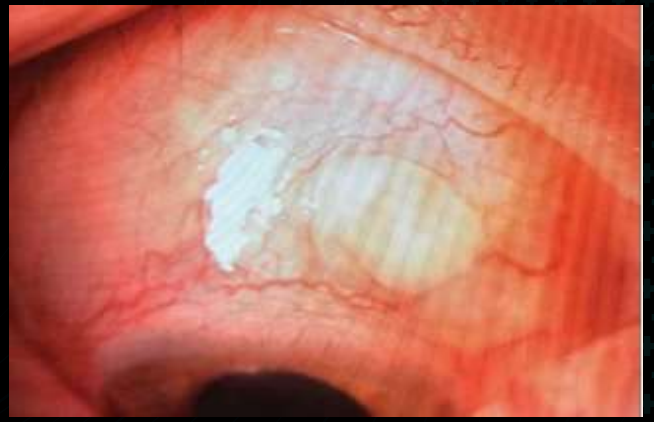

Figure 3: Right eye biomicroscopy

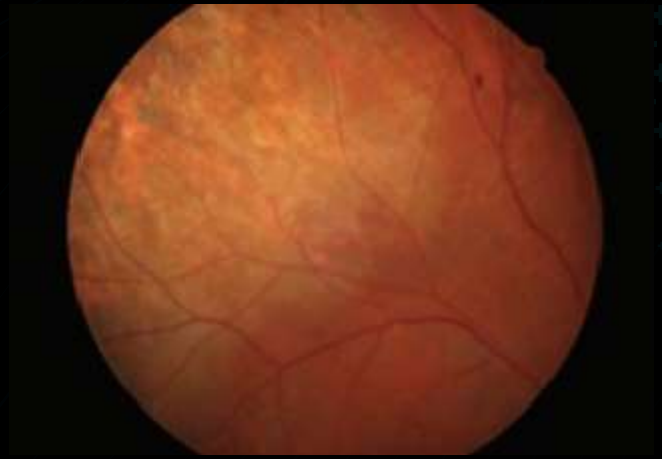

Figure 4: Right eye fundus photograph (superior temporal) - after treatment

DISCUSSION AND CONCLUSION

The current incidence of GPA is approximately 2.4– 11.3 cases per 1 million, predominantly in Caucasians and with no sex predilection.[1] The age at which symptoms appear varies but predominates between 41 and 68 years.[1]
Studies in North America, with 701 patients, reported an association of GPA with ocular involvement of 30%, whereas other reports reached results of up to 50%–58%.[2]

In our case, retinitis presented as the initial manifestation of ocular GPA and later developed scleritis. This differs from the literature since ocular scleritis is more common as an initial manifestation.

Scleritis associated with Granulomatosis with polyangiitis Its association with systemic diseases is approximately 40%–50% of patients, and the most frequently associated morbidity is rheumatoid arthritis. This association was described in 17%–33% of patients. The second most frequently associated disease is GPA.[2]

In cases of advanced disease, recent protocols suggest the induction of disease remission, associating high doses of systemic corticosteroids with cyclophosphamide or rituximab (immunobiological). The cyclophosphamide should be discontinued after remission, due to its toxic effects, and replaced by another drug, such as azathioprine, methotrexate, or immunobiological. If there is no remission, cyclophosphamide should be replaced by immunobiological and vice versa.[3,4]

GPA is a rare disease and misdiagnosis can happen in patients with scleritis. Delay in diagnosis can have consequences for patients' vision, as well as ocular globe preservation, in addition to other systemic disorders, which can be rapidly progressive, leading to long-term disability or mortality.

Cases treated early, with immunosuppressive therapy and well-indicated corticosteroids, evolve with good visual acuity and improvement in the patients' quality of life.

References:

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