

Thrombosis of the retinal artery in a patient with Churg-Strauss syndrome

Sirs,

A 48-year-old man was admitted to our hospital with a 2-year history of asthmatic crises. The patient presented an expired general condition, difficulty in walking and maintaining an upright position, pale skin, and several petechia mainly on the inferior extremities. The lymphonodes (at the right inframandibular and left inguinal levels) were swollen and hard; the muscular apparatus appeared hypotonic and hypotrophic; basal bilateral bluntness was detected in the lungs; at the abdomen deep and superficial palpation in the epigastric periumbilical area was painful. Moreover, the patient reported vision loss in the right eye of at least 3 days duration. Blood tests showed 54000 leukocytes (67% eosinophils and 565000 thrombocytes); increased globulin alpha-2; and negative CPK, LDH and ANCA. At cytomorphological examination the medulla appeared infiltrated, equal to 63% by eosinophilic granuloblasts, prominently granulocytes presenting basophilic granulation. The electromyography indicated a serious extended neurochordal situation with damaged segments of the inferior extremity; Doppler echocardiography showed a paradox septal motion and slight pericardial effusion. Cytochemical assay of the liquor showed a slight increase of proteins and derangement of the haemato-liquoral barrier; myeloencephalic RM showed an in-

crease of the perihemispheric liquor space and a deranged signal with hypodensity in all vertebral metamers. Examination of the *fundus oculi* showed complete occlusion of the central retinal artery on the right side with papilloedema and oedema of the posterior pole, a cherry-red spot, and on the left a trophic papillary with marked thinning of the arterial vessels and venous congestion. Capillaroscopic examination showed remarkable meandering of the segments, however, thin and subtle and flow velocity increased with sludge phenomenon. The anisgmoidoscopy revealed the presence of aphthosis-like lesions and blots of hyperaemia which biopsy was suggestive for a granulocytic infiltrate like for the cutaneous biopsy. Thus, in this patient 5 of the 6 classification criteria for Churg-Strauss syndrome established in 1990 by the American College of Rheumatology (asthma, hypereosinophilia, the mono- or polyneuropathia, migrating pulmonary infiltrates, paranasal sinusitis and extravascular eosinophilia) were fulfilled (at least 4/6 criteria must be met for the diagnosis) (1). The subject starts with a prodromic stage characterized by asthmatic attacks and cutaneous allergies lasting 2-3 years, followed by a second period of hypereosinophilia associated with gastroenteric discomfort and migrating pulmonary infiltrates, and the third stage with generalized vasculitis and polyneuropathy. In our case the non-specific phase of allergic manifestation lasted a long time compared to the second and third stages, which were quite serious and evolved rapidly. About 20-25 days passed from the appearance of oedema in the face to the loss of vision in the right eye. Thrombosis of the retinal artery caused the total and persistent loss of the vision on the right eye. Ocular involvement in Churg-Strauss syndrome is very rare (2,3). Ophthalmologic lesions can be due to ischaemia of the optical nerve or retinal circulation, or both (4-7). In our patient the vascular insult was concentrated in the retina. The loss of vision is usually persistent and rarely temporary. As a consequence a diagnosis can be made early and timely pharmacological treatment instituted. The subject was treated with cortisone (80 mg/day), cyclophosphamide (200 mg/day), and thrombocytic antiaggregants.

The early diagnosis and treatment with high dose corticosteroids and cyclophosphamides prevented irreversible damage of the other eye by vascular, necrotizing and thrombotic lesions (8). In addition, this led to a regression of the polyneuropathia, which is conditioned by the mobility of the inferior extremities (9). The patient achieved a satisfactory physical condition with the capacity to walk independently, even if he did not

recover the vision on his right eye.

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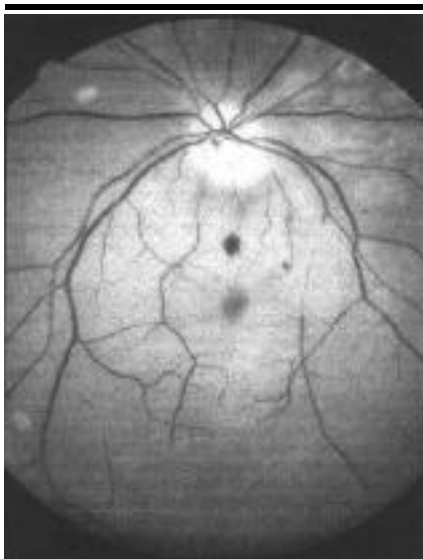


Fig. 1. Ophthalmoscopic picture of occlusion of the central retinal artery, papilloedema, and oedema of the posterior pole in a patient with Churg Strauss syndrome.