

Bilateral dacryoadenitis as a presenting symptom of an extra-ocular disease

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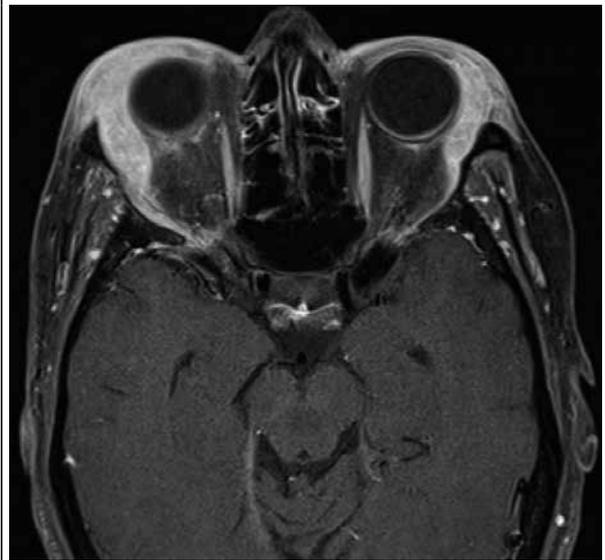
CASE REPORT

A 49-year-old woman, with no medical history, presented with bilateral swollen eyelids, progressive conjunctivitis and proptosis for the last month (*figure 1*). She was referred by the ophthalmologist because an underlying disease was suspected. Magnetic resonance imaging of the orbitae showed bilateral enlargement of the lacrimal glands (*figure 2*). Treatment by the ophthalmologist with antihistamines, topical corticosteroids and antibiotics had no effect. She had no other complaints. Physical examination showed evident swelling of both eyes without nasal or oral inflammation. Laboratory investigations showed: C-reactive protein 109 mg/l (0-10), haemoglobin 6.6 mmol/l (7.5-10.0), with a normal cellular volume, leucocytes $10.0 \times 10^9/l$ (4.0-10.0), and a normal serum creatinine; IgG4 levels were within normal limits. Urinalysis showed erythrocytes (3+), leucocytes (2+) and protein (1+).

Figure 1. Patient with bilateral swollen eyelids



Figure 2. Magnetic resonance imaging after gadolinium administration showing bilateral swelling of the lacrimal glands



WHAT IS YOUR DIAGNOSIS?

See page 285 for the answer to this photo quiz.

Antineutrophil cytoplasmic antibody (ANCA) associated vasculitis with renal involvement was suspected. Autoimmune investigation showed positive ANCA immunofluorescence for myeloperoxidase of 47 kU/l (<10). Microscopic examination of the urine showed >40 % dysmorphic red cells without casts. A chest X-ray, computed tomography (CT) of the thorax and sonography of the kidneys, showed no abnormalities.

Renal biopsy revealed a segmental necrotising crescentic glomerulonephritis, without immune deposits (pauci-immune) on immunofluorescence. Because granulomatosis was absent, the diagnosis of 'microscopic polyangiitis' (MPA) was made, although granulomatosis with polyangiitis (GPA) could not be entirely excluded.^{1,2}

MPA is an ANCA-associated autoimmune vasculitis affecting the medium and small arteries and veins. Typically the upper airways and kidneys are involved. Ophthalmological involvement occurs in approximately 5 to 30% of cases and may be, as in our patient, the

presenting symptom. Conjunctivitis is present in 30% of the patients with ocular complications; severe episcleritis and uveitis are rare.³ We did not find any literature relating dacryoadenitis to MPA.

The patient was treated with oral cyclophosphamide in a dose of 2 mg/kg/day and oral prednisone 60 mg once daily. Her complaints subsided quickly and she was discharged in a good condition.

REFERENCES

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