A renal biopsy yields sight as well as insight

A 34-year-old woman presented with periorbital oedema and progressive visual loss. Periorbital oedema was first observed 2 weeks before admission, particularly in the mornings. Visual loss started 5 days prior to admission. The patient was born in Thailand. She had a renal disease of unknown origin in 1974 and was treated for malaria in 1975. She has an 11-year-old healthy son and has lived in Germany since 1996. The patient was normal, except for pronounced periorbital oedema and bilateral conjunctivitis. Minor cutaneous changes included maculopapular eruptions with fine scaling over the bridge of the nose and some residual hyperpigmentation. Her erythrocyte sedimentation rate (ESR) was elevated at 77 mm at 1 h; however, the C-reactive protein (CRP) was normal. Autoantibodies directed against nuclear components (ANAs) were positive at a titre of 1:1280 with a fine speckled pattern. A precise nuclear antigen could not be determined. Complement C3c (0.36 g/l), C4 (0.015 g/l) and C1q (72 mg/l) were all reduced. Rheumatoid factor was negative. The microscopic urinalysis was normal, except for single hyaline casts and erythrocytes. The protein excretion was 1.2 g per 24 h; the creatinine clearance was mildly reduced (54 ml/min) for her size.

Neurologically, the patient was also normal. EEG and cranial MRT showed no pathological changes. She had a reduced visual acuity of 0.1 bilaterally. Ophthalmological examination revealed pronounced oedema of the retina and serous retinal detachment involving the fovea (Figure 1A). Early and late phase fluorescein angiograms show numerous leak sites, multiple punctate hyperfluorescences corresponding to the area of retinal alteration as well as filling of retinal detachment spaces (Figure 1B).

Question

What is your diagnosis?
Answer to the quiz on the preceding page

Because of the acute visual loss accompanied by periorbital oedema and protein excretion, the patient underwent a renal biopsy. Conventional histology showed no pathological glomerular alterations (Figure 2A). However, fluorescein isothiocyanate (FITC) immunofluorescence staining for IgG revealed granular IgG deposits predominantly in the mesangium (Figure 2B). These findings are consistent with lupus glomerulonephritis class I according to the 2002 WHO/ISN/RPS classification. Based on these findings, the diagnosis of systemic lupus erythematosus (SLE) was made. The patient was treated with 250 mg of prednisolone for 3 days. The eyes were also treated locally with prednisolone. The visual acuity improved markedly by 3 weeks. Six months after treatment, prednisolone was reduced to a maintenance level. Her visual acuity was 0.8 bilaterally and the protein excretion decreased to 0.3 g per 24 h. Our therapy was confined to steroids since the patient’s vision improved and her renal involvement was mild. Should her clinical course change, we would probably review our treatment and commence cyclophosphamide therapy.

SLE is a chronic inflammatory disorder of unknown aetiology that may affect all organ systems. The serological hallmark of SLE is the production of high titre ANAs directed against a variety of nuclear components. Much of the tissue damage, especially in blood vessels and kidney, is caused by the deposition of autoimmune complexes in the capillaries of the affected organs. Ocular involvement is common (prevalence up to 30%) and extremely varied [1–3]. Anterior segment and chamber findings include keratoconjunctivitis, scleritis, keratitis and iridocyclitis. Posterior segment and chamber abnormalities range from relatively common, asymptomatic microvascular changes, including cotton-wool spots and intraretinal haemorrhages, to rare, vision-threatening retinal arterial or venous occlusions. Optic neuritis and papilloedema have also been described. However, only occasional reports of SLE patients with central serous chorioretinopathy have been published [1]. The underlying mechanisms are believed to include choroidal vascular perfusion and permeability changes, which result in the alteration of the choroidal interstitial fluid composition followed by its extension. Any disease process, be it inflammatory, infectious, vascular, degenerative or genetically determined, which can cause such choroidal changes may therefore induce serous retinal detachments. SLE, but also sarcoidosis, Wegener’s granulomatosis, Behçet’s disease, and Vogt–Koyanagi–Harada syndrome may be associated with chorioretinopathy and serous retinal detachment [2,4]. In our patient, we had no evidence for cytomegalovirus infection, syphilis, HIV, ischaemic vasculopathy, thrombotic thrombocytopenic purpura, disseminated intravascular coagulation or eclampsia that may also cause the syndrome. The diagnosis of SLE was supported largely by the histopathological findings on renal biopsy (Figure 2A). The biopsy made other renal-associated causes of the ophthalmological symptoms, such as IgA nephropathy or type II membranoproliferative glomerulonephritis, unlikely.

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References


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Fig. 2. (A) Light microscopy section showing normal renal architecture (PAS; ×20). (B) FITC immunofluorescence staining for IgG of a glomerulus revealing granular IgG deposits predominantly in the mesangium.