Clinical vignette

Retinal vasculitis in primary Sjögren’s syndrome

A 49-year-old man presented to our hospital with dry mouth for 1 year, chilblain rash, mottled appearance of the skin of his hands and legs and a 4 week history of bilateral blurred vision. Funduscopic evaluation revealed intra- and pre-retinal haemorrhages, retinal exudates and vascular sheathing (Fig. 1A). Fundus fluorescein angiography showed non-perfusion areas and extravascular leakage of the dye with blocking defect (Fig. 1B). Both anti-SSA and anti-SSB antibodies were positive, but ANA, anti-dsDNA antibodies and ANCA were negative. The unstimulated salivary flow rate was low and salivary scintigraphy demonstrated decreased uptake in his parotid glands. No signs of large vessel vasculitis or SLE were observed. Skin involvement in his legs worsened to form multiple ulcers (Fig. 1C). Skin biopsy revealed predominantly lymphocytic infiltration surrounding small-sized blood vessels, consistent with lymphocytic small vessel vasculitis (Fig. 1D). The patient was diagnosed with retinal and cutaneous vasculitis associated with primary SS [1]. He was treated with pan-retinal photoocoagulation for retinal haemorrhages. Oral prednisolone 30 mg/day was also initiated. Thereafter his retinal and cutaneous symptoms gradually improved. Retinal vasculitis in SS is rare and timely diagnosis and treatment may aid in avoiding blindness [2].

Fig. 1 Findings of the patient’s images

(A) Funduscopy showed retinal haemorrhages (black arrows), retinal exudates (black arrowhead) and vascular sheathing (black asterisks). (B) Fundus fluorescein angiography showed blocking defect (white arrow), extravascular leakage of the dye (white arrowhead) and non-perfusion areas (white asterisk). (C) Multiple skin ulcers on the patient’s legs. (D) Skin biopsy revealed predominantly lymphocytic infiltration surrounding small-sized blood vessels (haematoxylin and eosin stain; original magnification ×100).

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