A man in his twenties was admitted to hospital with headache, dizziness, vomiting, lethargy and weight loss. Cerebral MRI showed diffuse, confluent changes in white matter, consistent with cerebral vasculitis. Conventional angiography with selective injection in both carotids and the right vertebral artery showed normal arteries with no calibre changes or occlusions. Cerebrospinal fluid tests showed an elevated number of mononuclear cells, by far the majority of which were mature lymphocytes, with 107 leukocytes per cubic millimetre (≤ 5 leukocytes per mm³) and elevated total protein 0.94 g/l (0.10–0.40 g/l). An eye examination showed moderate papilloedema, normal vision and no visual field defects.

While in hospital the patient became acutely ill with reduced consciousness. A further cerebral MRI showed several infarctions. He was treated with cyclophosphamide and steroids and gradually improved.

Several weeks later the patient again became acutely ill, and new cerebral infarctions were detected. He now had visual disturbances and there were visual field defects in both eyes. An eye examination revealed increasing papilloedema with fresh bleeding around the papilla in both eyes. The picture shows papilloedema and bleeding in the right eye. Unfortunately we have no pictures of the peripheral retina. The patient again improved following treatment with steroids and cyclophosphamide. One month later the papilloedema and retinal bleeding had disappeared and there was only a small visual field defect.

Diagnosis of central nervous system vasculitis is difficult. The diagnostic tools are cerebral MRI, cerebrospinal fluid analyses and conventional angiography. The latter, however, has low sensitivity and specificity for small-vessel vasculitis. In some cases a brain biopsy may be required. Symptoms such as blurred vision, visual field defects and double vision are relatively frequent clinical manifestations and can help in making the diagnosis (1).

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