Multiple Hemorrhagic Cerebral Cortical Lesions in Neuro-Behçet’s Disease

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A 27-year-old Japanese man presented with erythema nodosum and consciousness opacity of four days in duration. He had panuveitis and fever. A cerebrospinal fluid (CSF) analysis revealed 100/mm$^3$ pleocytosis (polynuclear cells, 87%) and the elevation of interleukin-6 (5,630 pg/mL). Brain MRI demonstrated multiple hemorrhagic cerebral cortical lesions with gadolinium enhancement (Picture A, DWI; C, T1WI; D, gadolinium T1WI), but normal MR venography (Picture B). Cerebral venous thrombosis was not proven. The patient was positive for human leukocyte antigen-B51. He did not completely fulfil the International Study Group criteria for acute neuro-Behçet’s disease.

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(NBD) (1) because he lacked oral ulceration; however, the acute onset of neurological symptoms and CSF pleocytosis of >6.2/mm$^3$ both matched the criteria. He was treated with prednisolone (1 mg/kg/day; the dose was tapered by 5 mg per month and reduced to 5 mg/day over two years) and infliximab (5 mg/kg bimonthly) for one year, which resulted in remission. Although cortical lesions are rare in NBD (2), it is important to consider NBD when such atypical lesions are encountered.

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References


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