Clinical vignette

A case of relapsing neurosarcoidosis with brain nodules and hydrocephalus successfully treated by corticosteroid and methotrexate

Sir, A 45-year-old woman presented with a 2-month history of progressive cognitive dysfunction and red plaques on her extremities (Fig. 1A). She was diagnosed as having neurosarcoidosis (NS) by brain biopsy (Fig. 1B) 12 years before presenting and had been prescribed prednisolone 5 mg/day for the past 6 years. Laboratory examinations showed an increased serum angiotensin-converting enzyme level of 29.3 IU/l. Neither infection nor malignancy was detected. Skin biopsy of plaques revealed non-caseating epithelioid cell granuloma with giant cells (Fig. 1C, arrows), like the previous brain pathology. Contrast-enhanced MRI of the brain showed numerous high-intensity foci on the brain surface and hydrocephalus (Fig. 1D, arrows). Queckenstedt’s sign was positive. She was diagnosed as a relapse of NS. She underwent methylprednisolone pulse therapy, followed by prednisolone 50 mg/day and MTX 12 mg/week. After 4 weeks from the initiation of therapy, the high-intensity foci disappeared (Fig. 1E), Queckenstedt’s sign was negative and her cognitive function recovered.

NS develops in 5–13% of sarcoidosis patients [1]. Diagnosis of NS is challenging because of the variety of its clinical presentation and the difficulty of histological confirmation. Although hydrocephalus is a rare manifestation (5–7% of NS patients), it has a poor long-term prognosis [2]. In our case, multiple brain nodules and cognitive dysfunction improved remarkably, while hydrocephalus was unchanged after treatment.

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