Rheumatoid Arthritis with Neurological Involvement Manifested as Hemispheric Masses

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Dear Sir,

Central nervous system (CNS) involvement in patients with rheumatoid arthritis (RA) is rare. The pathological findings include rheumatoid nodule, pachy- or leptomeningitis, and vasculitis of necrotizing or plexiform type [1–3]. We report a patient with the unusual presentation of multiple masses in the cerebral hemispheres which were found to be associated with lymphocytic infiltration of some cerebral vessel walls. The masses regressed after steroid treatment.

Case Report

A 58-year-old right-handed male farmer had a 3-year history of RA manifested as intermittent painful swelling of the ankles and interphalangeal joints of both feet (fig. 1a), and an elevated rheumatoid factor titer of 197 IU/ml (normal value <30 IU/ml). He took only analgesics for symptom relief. He did not take any disease-modifying antirheumatic drugs or steroids. One month prior to admission, he started to experience painful swelling in his right wrist, which was followed by slurred speech and progressive weakness in his right limbs. At admission, he developed a low-grade fever and polyarthritis in both of his knees, ankles, shoulders, and right wrist. Neurological examination revealed that he had anoma, visual agnosia, mild right central facial palsy, right hemiparesis (Medical Research Council, MRC, grade 4/5), and right hemihypesthesia of all sensory modalities. Laboratory findings were unremarkable except for an elevated level of rheumatoid factor (238 IU/ml). A brain MRI scan disclosed 3 mass lesions in the bilateral temporal and the left periventricular regions. The lesions were hypointense in T1-weighted imaging, hyperintense in T2-weighted imaging (fig. 1b, c), and were not enhanced by gadolinium. The patient underwent a stereotactic biopsy of his periventricular lesion. The pathology revealed perivascular lymphocytic infiltration and reactive gliosis (fig. 1d, e), which were compatible with vasculitis. We initiated pulse therapy of methylprednisolone (1 g/day); however, the patient developed steroid psychosis 2 days later. We therefore discontinued the pulse therapy and administered oral prednisolone 60 mg/day instead. After 1 month of therapy, follow-up MRI disclosed regression of the 3 brain lesions (fig. 1f), and his symptoms improved with residual minimal right hemiparesis (MRC 4+/5). The patient’s clinical features remained stable during the subsequent 3 years of follow-up.

Discussion

Direct CNS involvement in patients with RA is rare. The well-known condition of rheumatoid vasculitis typically involves the skin and peripheral nerves, rarely does it affect the CNS. In addition, although nodular formation is a frequent and characteristic extra-articular feature of RA, nodular formation within the CNS is uncommon. Bathon et al. [1] reported on 19 patients with seropositive long-standing RA who developed inflammatory CNS lesions. In these patients, rheumatoid nodules were the most common pathological finding (68%), followed by lepto- or pachymeningeal inflammation (63%) and vasculitis (37%). These 3 pathological changes were present with similar frequencies in symptomatic patients, but CNS nodules alone were often found in asymptomatic patients. CNS disease occurs in a significant number of patients without active synovitis and extracranial vasculitis and nodules. CNS vasculitis in patients with RA may manifest as necrotizing vasculitis [4] or arterial plexiform change with amyloidosis [5]. These findings, however, were not observed in our patient.

Recently, Tajima et al. [6] reported a case of RA with multiple CNS lesions that resolved after steroid treatment. Regard-
In conclusion, we report a rare case of RA with multiple lesions in the brain involving both the gray and white matters of both hemispheres. The lesions were considered as vasogenic edema caused by a mild form of vasculitis that has not been reported previously.

References
1 Bathon JM, Moreland LW, DiBartolomeo AG: Inflammatory central nervous system involvement in rheumatoid arthritis. Semin Arthritis Rheum 1989;18:258–266.