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Case Report

MRI and pathological findings of rheumatoid meningitis

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Abstract

Rheumatoid meningitis (RM) is one of the severest complications of rheumatoid arthritis (RA) and the mortality rate is relatively high. RM diagnosis is sometimes very difficult. We present the case of an 80-year-old woman who was diagnosed with microscopic findings of RM from the biopsy specimens from a brain lesion. MRI revealed meningeal enhancement in the brain, and the pathological findings were meningeal lymphocytic infiltration, vasculitis, and rheumatoid nodules. RM is a treatable disease and this is an important RM case that was diagnosed on the basis of biopsy findings.

Key words: rheumatoid arthritis; meningitis; MRI; treatment; pathology

Introduction

Neurological manifestations of rheumatoid arthritis (RA) are rare (1); however, there are some reports on central nervous system involvement in RA. Rheumatoid meningitis (RM) is one of the severest complications of RA and the mortality rate is relatively high (2). An ante mortem diagnosis of RM is often difficult; some case reports describe its diagnosis at autopsy. Here, we report an important case of RM along with the magnetic resonance imaging (MRI) findings; RM was diagnosed on the basis of biopsy findings and treated with steroid therapy.

Case report

An 80-year-old woman with a 20-year history of RA was admitted to our hospital for repeated attacks of intermittent weakness and numbness in the right extremities for 2 months. The condition was classified as stage 4 and class 2 according to Steinbrocker Criteria for RA (3). On admission, her neck, hands, and knees were operated for RA. The low-dose steroid, anti-inflammatory drugs salazosulfapyridine, bucillamine, and etanercept had been already prescribed for RA when she was admitted. Laboratory data, including the levels of the rheumatoid factor and other autoimmune antibodies, showed no significant abnormalities. The cerebrospinal fluid (CSF) test revealed a slightly increased white blood cell count (18 cells/mm³; 93% mononuclear cells), protein level (55 mg/dl), IgG index (1.38), and interleukin (IL)-6 level (4.6 pg/ml). The brain MRI revealed

hyperintense lesions in the cortex of the left parietal lobe on T2-weighted (T2W) and fast fluid-attenuated inversion-recovery (FLAIR) MRI images (Fig.1-A) and meningeal thickening with abnormal enhancement on the surface of the left parietal lobe (Fig.1-B). Examination of the brain biopsy specimens from the left parietal lesion revealed inflammatory cell infiltration in the dura, suppurative granuloma formation with macrophages and polynuclear giant cells on the surface of the leptomeninges, and extension of the inflammation along the vessels in the cortex (Fig.2-A, B). No infection or tumor was observed in the other immunostainings and cultures of the biopsy specimens, and microscopic findings were consistent with RM. The patient received pulsed therapy with methylprednisolone at a dosage of 1000 mg/d for 3 days, followed by oral prednisolone at a dose of 60 mg/day. The symptoms improved gradually, and the attacks disappeared. CSF abnormalities also improved. The hyperintense lesions were not observed on the T2W and FLAIR MRI images (Fig.1-C), and meningeal enhancement decreased (Fig.1-D). The dose of prednisolone was tapered gradually, and other immunosuppressive treatments were not administered. The patient did not show any recurrence of symptoms or MRI findings for 1 year.

Discussion

Thus far, there are 17 reports on RM diagnosed by biopsy or autopsy listed in Table 1 (1, 2, 4–17).

Half of them had severe long standing RA. RM sometimes cannot be distinguished from other

diseases, especially tumors, without a pathological procedure because there are no specific RM markers in the blood or CSF. In one case, a marked increase of CSF IL-6 level was reported (2); our patient also showed a slight increase in CSF IL-6. CSF analysis also shows nonspecific findings with pleocytosis and an elevated protein level. For RM diagnosis, MRI and biopsy are critical. Meningeal enhancement in the brain MRI scan is the key feature of RM (1, 2, 4, 7–9, 11, 16). Pathological findings of RM are characterized by meningeal lymphocytic infiltration, vasculitis, and rheumatoid nodules (2, 11). Although meningeal infiltration is often noted, the presence of all the 3 typical findings in a single patient is rare (11). In our case, all the findings were observed; this was quite helpful in diagnosing RM. RM treatment has not yet been established; however, steroid therapy is considered the first choice according to previous reports (4, 8, 16). Monotherapy with steroid is insufficient in some cases (1, 2, 9), and recent reports have mentioned the efficiency of immunosuppressive agents such as cyclophosphamide (CY) (1, 5, 7, 9, 11, 12), azathioprine (AZA) (6), cyclosporine (CyA) (9), and methotrexate (MTX) (16) against RM. In our case, etanercept, a relatively new anti-RA agent, was used for RA treatment prior to RM occurrence. Etanercept, however, could not prevent the development of RM in this case. In our case, monotherapy with steroid was effective. RM is a treatable disease, and its accurate diagnosis is critical for treatment. Pathological findings offer useful information for diagnosis.

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Figure Legends

Fig. 1

Axial magnetic resonance images (A–D). FLAIR image (A) and gadolinium (Gd)-enhanced T1W image (B) obtained on admission showing a left parietal lesion. After treatment, marked improvement in the lesion was observed on FLAIR images (C) and Gd-enhanced T1W images (D).

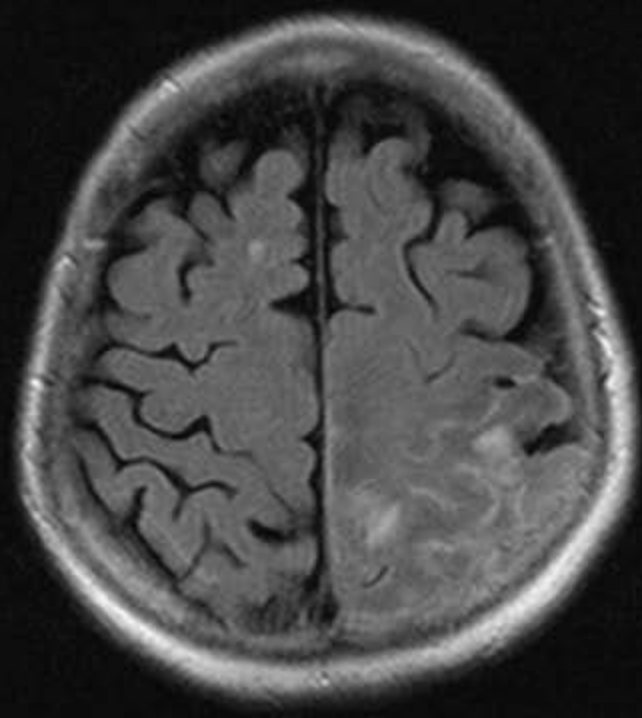
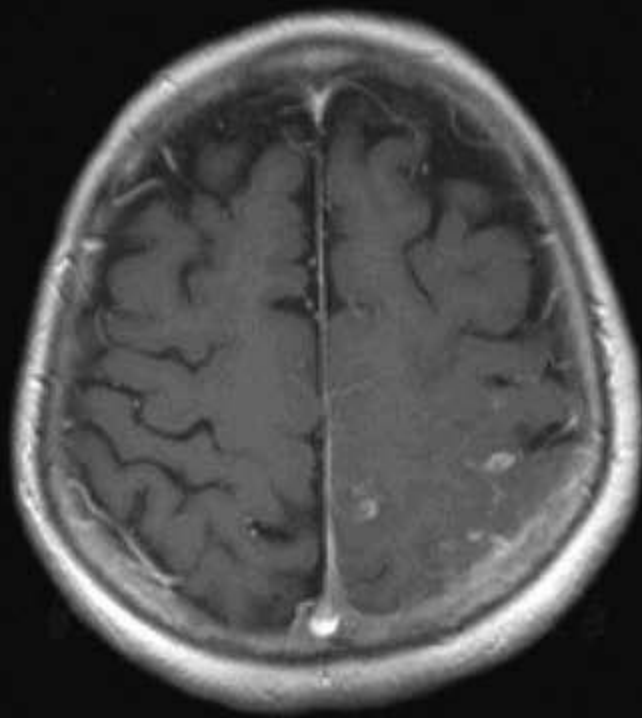
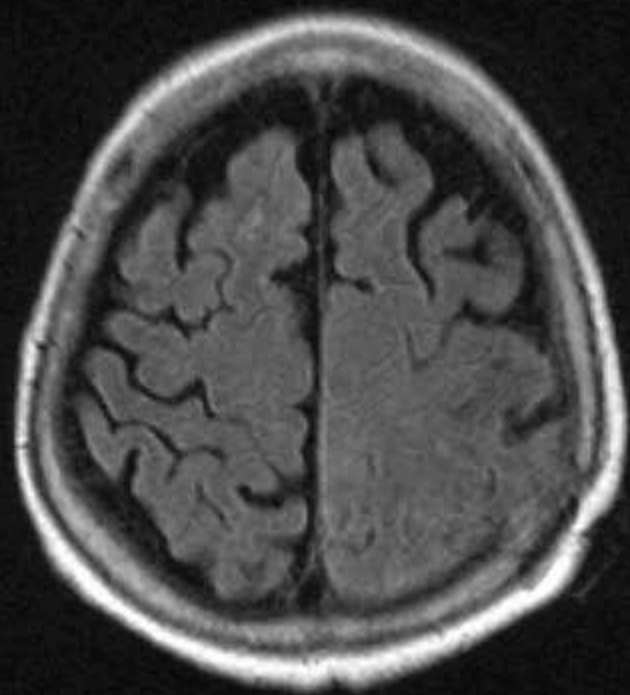
Fig. 2

Biopsy specimens of the left parietal lobe stained with hematoxylin-eosin (A and B). Low-power (A) and high-power (B) photomicrographs indicate inflammatory cell infiltration in the dura (arrowhead) and suppurative granuloma with macrophages and polynuclear giant cells (arrows) on the surface of the leptomeninges.

Table 1

References	Age(y) gender	Symptoms	CSF findings (WBC [/ mm^3] / MRI Protein [mg/dl] / glucose [mg/dl])	enhance	Treatment	Outcome	Biopsy or autopsy	Pathological findings (cell infiltration / rheumatoid nodule / vasculitis)
(10)	63 M	loss of consciousness, seizure	N.D.	N.D.	untreated	death	autopsy	+ / + / -
(14)	58 F	loss of consciousness, confusion, seizure	3 / 80 / 58	N.D.	steroid	death	autopsy	+ / + / +
(15)	63 M	confusion, dementia, blindness	0 / 133 / 57	N.D.	untreated	death	autopsy	+ / - / +
(17)	69 M	seizure	many / 50 / 260	N.D.	steroid	death	autopsy	+ / + / -
(13)	70 F	weakness	220 / 421 / <5	N.D.	steroid	improvement	biopsy	+ / - / -
(12)	63 F	diplopia, left dysesthesia	232 / 260 / 6	N.D.	aspirin + CY	death	autopsy	+ / + / -
(6)	52 M	loss of consciousness, seizure, right paresthesias, aphasia	24 / 75 / 49	N.D.	steroid + AZA	improvement	biopsy	+ / - / +
(5)	37 M	headache, visual disturbance	70 / 100 / 53	N.D.	steroid + CY	improvement	biopsy	+ / - / -
(1)	52 M	diplopia	8 / 53 / ND	+	steroid + CY	improvement	biopsy	+ / - / -
(2)	71 F	fever, arthralgia,, headache, nausea	377 / 74 / 9	+	steroid	improvement and relapse	autopsy	+ / - / +
(8)	78 F	headache, left paresthesias, left hemiparesis	23 / 73 / 60	+	steroid	improvement	biopsy	+ / + / +
(9)	68 F	left paresthesias, left hemiparesis	9 / 71 / 62	+	steroid + CyA + CY	improvement	biopsy	+ / - / -
(11)	58 F	left paresthesias	reactive process	+	steroid + CY	improvement	biopsy	+ / - / -
(7)	58 F	headache, numbness, dysarthria, weakness, seizure	N.D.	+	steroid + CY	improvement	biopsy	+ / + / -
(4)	77 M	headache, dysphasia, involuntary movement, confusion	88 / 67 / 48	+	steroid	improvement	biopsy	+ / - / -
(16)	67 M	hearing loss	N.D.	+	steroid + MTX	improvement	biopsy	+ / - / -
(16)	76 F	confusion, seizure	N.D.	+	steroid	improvement	biopsy	+ / + / -
This case	80 F	weakness, numbness	18 / 55 / 64	+	steroid	improvement	biopsy	+ / + / +

N.D.: not described, CY: cyclophosphamide, AZA: azathioprine, CyA: cyclosporine, MTX: methotrexate

A**B****C****D**