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SHORT COMMUNICATION

Magnetic resonance imaging of rheumatoid meningitis: a case report and literature review

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Abstract Rheumatoid meningitis is a rare and serious complication of rheumatoid arthritis (RA) with high mortality rate. Clinical importance of the disease is high because diagnosis is difficult, and the disease is treatable if diagnosed successfully. We present the clinical and cranial magnetic resonance imaging findings of 62-year-old female patient with RA who has been followed up for 4 years.

Keyword Rheumatoid arthritis · Rheumatoid meningitis · Magnetic resonance imaging

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Introduction

Although neurological manifestations of rheumatoid arthritis (RA) are rare, central nervous system (CNS) involvement has been reported [1]. CNS involvement can occur as parenchymal and meningeal vasculitis, rheumatoid nodules, pachymeningitis or leptomeningitis. Rheumatoid meningitis (RM) is one of the most serious complications of RA with a high mortality rate [2, 3]. In this report, we present the clinical and cranial magnetic resonance (MR) imaging findings of a 62-year-old RA patient who has been followed up for 4 years and diagnosed as RM by demonstrating findings of meningitis on cranial MR scan and excluding other etiologic factors. In our case, clinical recovery is provided after using high-dose corticosteroids (CS) and methotrexate (MTX), and we emphasize the importance of cranial MR imaging findings on the diagnosis and the treatment for RM.

Case report

A 62-year-old woman with a 4-year history of RA who was admitted to our hospital after experiencing headache over 1 month and newly developed paresis and paresthesis on her left arm. RA diagnosis made by criteria of American College of Rheumatology [4]. According to her medical history, the patient suffered twice from deep vein thrombosis on her right lower extremity. Physical examination showed arthritis with synovial hypertrophy on her hand, foot and wrist joints, compatible with RA. There were no abnormal findings in neurologic examination and mental status. The patient was using CS and MTX for treatment for RA and warfarin for deep venous thrombosis. Laboratory data showed increased rheumatoid factor as high as 351 IU/mL

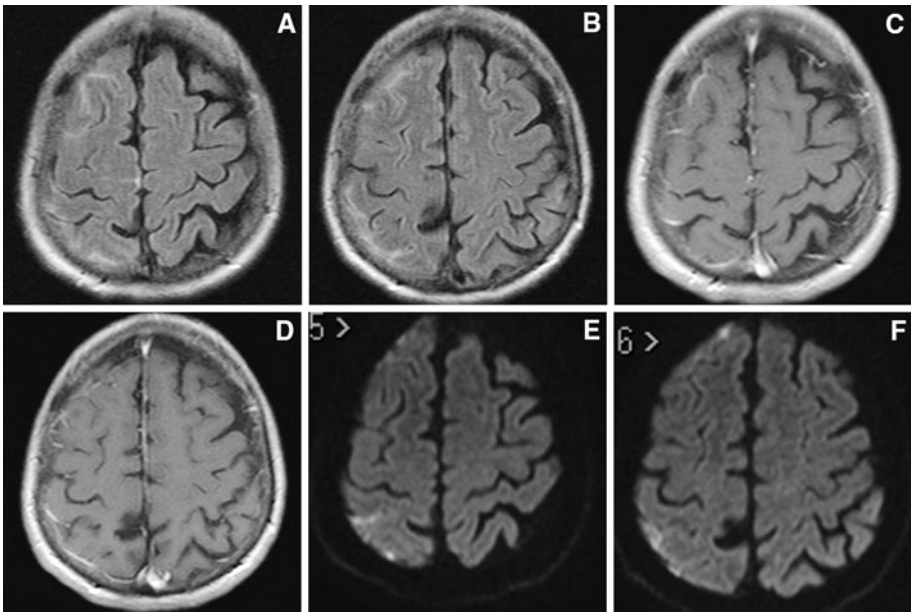

Fig. 1 Axial fluid-attenuated inversion recovery (FLAIR) MR images showing hyperintensity in the *right* frontoparietal subarachnoid space (**a**, **b**) and gadolinium-enhanced T1-weighted MR images showing meningeal thickening and pathologic contrast enhancement (**c**, **d**). Diffusion-weighted MR images showing restriction of diffusion at the same localization (**e**, **f**)

and anti-cyclic citrullinated peptide (120 IU/mL). Serum examination for other autoantibodies was normal. CSF analysis revealed mildly increased white cell count (40 cells/mm^3) and protein levels (40 mg/dL) without any atypical cells. On Doppler ultrasonography which was made because of previous deep venous thrombosis history, chronic thrombotic alterations were detected on lower right extremity. No pathological changes were found on upper extremities. Cranial MR imaging revealed hyperintensity in the right frontoparietal subarachnoid space on fast fluid-attenuated inversion recovery (FLAIR) images and meningeal thickening and pathologic contrast enhancement on T1-weighted images. Diffusion-weighted images showed restriction of diffusion at the same localizations (Fig. 1). Reasons that may lead to leptomeningeal and pachymeningeal enhancement by imaging characteristics are sought. These are infectious (tuberculosis, fungal, pyogenic, cysticercosis, etc), tumoral (lymphoma, leukemia, carcinomatosis and meningioma), inflammatory (Behçet's disease, Sjögren's syndrome, Whipple's disease, Wegener's granulomatosis and temporal arteritis) and other (idiopathic pachymeningitis and intracranial hypotension) causes. But with the patient's clinical history, physical examination, blood count, biochemistry results, CSF analysis findings, lung X-ray, thoracic and abdominal CT, and other diseases except RM were excluded. Definite diagnosis of RM is made by pathologic examination, but we could not perform biopsy because of disapproval of the patient. According to the imaging and laboratory findings and 4 years of RA history, we assumed the diagnosis was RM. Then, we started methylprednisolone 1,000 mg/day for 5 days, continued with oral prednisolone 60 mg/day and added 20 mg/week MTX and 10 mg/week folic acid treatment. We gradually decreased the oral prednisolone dose until reaching maintenance dose of 4 mg/day. Both the clinical response and normalized CSF examination findings supported the RM diagnosis.

Discussion

Rheumatoid arthritis is a systemic inflammatory disease that primarily affects joints, but it may also affect extra-articular structures [5]. RM is a rare but a serious complication of RA with high mortality rate [1, 3]. Neurologic symptoms of RM are headache, signs of cranial nerve involvement, seizures, and alteration of mental status, hemiparesis or paraparesis [6–8]. Diagnosis of RM is difficult since that there are no specific RM markers in the CSF or blood. CSF analysis of RM cases shows non-specific findings such as increased protein levels, pleocytosis and decreased glucose levels. Only two cases of interleukin-6 (IL-6) increase on CSF analysis are reported [2, 8]. In our case, CSF and serum examination was normal except slight increase in white blood cell count and protein level.

Cranial MR imaging and pathological examination has critical role on diagnosis of RM [1, 2, 9–14]. Meningeal thickening and contrast enhancement are important findings of RM on cranial MR imaging. In 2005, Jones et al. reported a case with restriction of diffusion because of proteinous debris accumulation at subarachnoid space adjacent to parenchyma with meningeal infiltration [5]. In our case, we disclosed restriction of diffusion at subarachnoid space adjacent to the right frontal lobe.

Presence of those findings on cranial MR imaging suggests RM diagnosis but other reasons that might cause leptomeningitis and pachymeningitis must be excluded [5].

Definite diagnosis is made by demonstrating of rheumatoid nodules by biopsy. Characteristic pathologic findings of RM are meningeal lymphocytic infiltration caused by infiltration of meningeal structures by mononuclear cells particularly by plasma cells, rheumatoid nodules and vasculitis that induced by lymphoplasmocytic infiltration around small vessels at meninges and parenchyma [2, 5].

There are 18 cases of antemortem or postmortem diagnosed RM that was reported in literature [4, 5, 7, 10]. Kato et al. suggested increased CSF IL-6 level in patients with RM, and this may come in to use as a therapeutic marker. Additionally, if the IL-6 level is increased, adding of another immunosuppressive agent to CS treatment is necessary. Relying on this fact, in our case, we continued to MTX as an immunosuppressive agent along with CS therapy.

For the treatment for RM, according to previous literature acknowledgements, CS is the first choice [10, 11, 13]. Since monotherapy with steroids does not provide recovery [1, 2, 12], adding of immunosuppressive agents such as cyclophosphamide [1, 5, 10, 12, 14, 15], azathioprine [2], cyclosporine [12] and MTX [13] to therapy is recommended. In our case, MTX with high dose CS therapy likewise provided effective treatment.

Conclusions

Rheumatoid meningitis is a treatable disease with high mortality rate. For treatment, accurate diagnosis has a crucial role. Cranial MR imaging and biopsy findings have primary importance on RM diagnosis. In RA patients, meningeal thickening and contrast enhancement on cranial MR should bring RM to mind. To make accurate diagnosis, as we elucidated, other conditions that may cause meningeal enhancement should be excluded.

References

1. Yucel AE, Kart H, Aydin P et al (2001) Pachymeningitis and optic neuritis in rheumatoid arthritis: successful treatment with cyclophosphamide. Clin Rheumatol 20:136–139
2. Kato T, Hoshi K, Sekijima Y et al (2003) Rheumatoid meningitis: an autopsy report and review of the literature. Clin Rheumatol 22:475–480
3. Bekkelund SI, Pierre-Jerome C, Husby G, Mellgren SI (1995) Quantitative cerebral MR in rheumatoid arthritis. Am J Neuroradiol 16:767–772
4. Arnett FC, Edworthy SM, Bloch DA et al (1988) The American Rheumatism Association 1987 revised criteria for the classification of rheumatoid arthritis. Arthr Rheum 31:315–324
5. Jones SE, Belsley NA, McLoud TC, Mullins ME (2006) Rheumatoid meningitis: radiologic and pathologic correlation. Am J Roentgenol 186:1181–1183
6. Bathon JM, Moreland LW, DiBartolomeo AG (1989) Inflammatory central nervous system involvement in rheumatoid arthritis. Semin Arthr Rheum 18:258–266
7. Cellerini M, Gabbrielli S, Maddali Bongi S, Cammelli D (2001) MRI of cerebral rheumatoid pachymeningitis: report of two cases with follow-up. Neuroradiology 43:147–150
8. Matsushima M, Yaguchi H, Niino M et al (2010) MRI and pathological findings of rheumatoid meningitis. J Clin Neurosci 17:129–132
9. Ahmed M, Luggen M, Herman JH et al (2006) Hypertrophic pachymeningitis in rheumatoid arthritis after adalimumab administration. J Rheumatol 33:2344–2346
10. Chou RC, Henson JW, Tian D et al (2006) Successful treatment of rheumatoid meningitis with cyclophosphamide but not infliximab. Ann Rheum Dis 65:1114–1116
11. Chowdhry V, Kumar N, Lachance DH et al (2005) An unusual presentation of rheumatoid meningitis. J Neuroimaging 15:286–288
12. Claassen J, Dwyer E, Maybaum S, Elkind MS (2006) Rheumatoid leptomeningitis after heart transplantation. Neurology 66:948–949
13. Starosta MA, Brandwein SR (2007) Clinical manifestations and treatment of rheumatoid pachymeningitis. Neurology 68:1079–1080
14. Kim RC (1980) Rheumatoid disease with encephalopathy. Ann Neurol 7:86–91
15. Beck DO, Corbett JJ (1983) Seizures due to central nervous system rheumatoid meningovasculitis. Neurology 33:1058–1061