

Case report

Pachymeningitis and optic neuritis in rheumatoid arthritis: MRI findings

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Abstract. Rheumatoid arthritis is a systemic disease in which cerebral and eye involvement is neither common nor fully understood. Although it is rarely the cause of pachymeningitis and optic neuritis, rheumatoid arthritis should always be kept in mind in these two conditions. We present a 52-year-old male with an 8 month history of rheumatoid arthritis who was referred to the neurology department with headache and decreasing vision and was diagnosed as having rheumatoid pachymeningitis and optic neuritis on the basis of MRI findings.

Rheumatoid arthritis (RA) is a systemic disease with a mortality rate. Although the commonest cause of death in RA is cardiovascular involvement, central nervous system involvement causes death in up to 18.6% of cases [1]. Cerebral involvement in RA is neither common nor fully understood. Dural nodules, hyperviscosity syndrome and cerebral vasculitis are reported manifestations but reports are few and most do not involve controlled studies [1–3]. The rheumatoid process affects the eye in very few cases [4–6]. Here we present the MRI appearances of dural and optic nerve involvement in a patient with rheumatoid arthritis.

Case report

A 52-year-old male with an 8 month history of RA was referred to hospital with headache, fatigue, right-sided weakness and swelling of the left eye. He also complained of decreased vision in both eyes, especially the left. On physical examination, there were rheumatoid nodules on both elbows. Ophthalmological examination showed swelling of the left optic nerve and the patient had diplopia on lateral gaze. A slight right hemiparesis and ataxia were evident on neurological examination. The patient also had mild dysarthria. He had been treated with prednisolone, methotrexate and chloroquine for 2 months after his initial diagnosis and had then stopped the medication.

The patient's rheumatoid factor level and erythrocyte sedimentation rate (64 mm h⁻¹, normal 0–20 mm h⁻¹) were elevated. Haemoglobin was 14.5 g dl⁻¹ (normal 16.0 ± 2.0 g dl⁻¹) and haematocrit

was 43% (normal 39–40%). Leucocyte count was 11 500 cells μl⁻¹ (normal 4500–11 000 cells μl⁻¹). The antinuclear antibody test was negative. The patient's cerebrospinal fluid glucose level was 57 mg dl⁻¹ (normal 40–70 mg dl⁻¹), total protein was 50 mg dl⁻¹ (normal 20–50 mg dl⁻¹), and chloride level was 119 mEq l⁻¹ (normal 116–122 mEq l⁻¹) and all were within normal limits.

Brain and orbital MRI was performed on a 1.0 T system, using a head coil. T₁ weighted spin echo (SE) axial and sagittal images (TR/TE: 500/15 ms), T₂ weighted turbo spin echo (TSE) axial images (TR/TE: 5000/105 ms) and T₁ weighted SE sagittal and coronal images (TR/TE: 500/15) after intravenous gadolinium administration and fat suppressed T₁ weighted sagittal images were obtained. The subarachnoid space in the upper frontal and interhemispheric region showed low signal intensity on axial T₁ weighted, and high signal intensity on axial T₂ weighted images relative to brain tissue (Figures 1a and b). Diffuse meningeal enhancement in the upper frontoparietal convexity, more prominent on the left and in the interhemispheric region, was seen after gadolinium injection (Figure 1c). The left optic nerve was thickened and diffusely enhanced when compared with the right side (Figures 2a and b). It was concluded that the lesions within the subarachnoid space represented nodules with dural involvement of RA.

Biopsy was performed on the dura of the left parietal convexity and the left optic nerve sheath. Histological examination showed hyalinization, a few lymphocytes, and plasma cell infiltration of the dura and optic nerve sheath. Decompression surgery to the left optic nerve was applied. In the light of these findings, the diagnosis of rheumatoid

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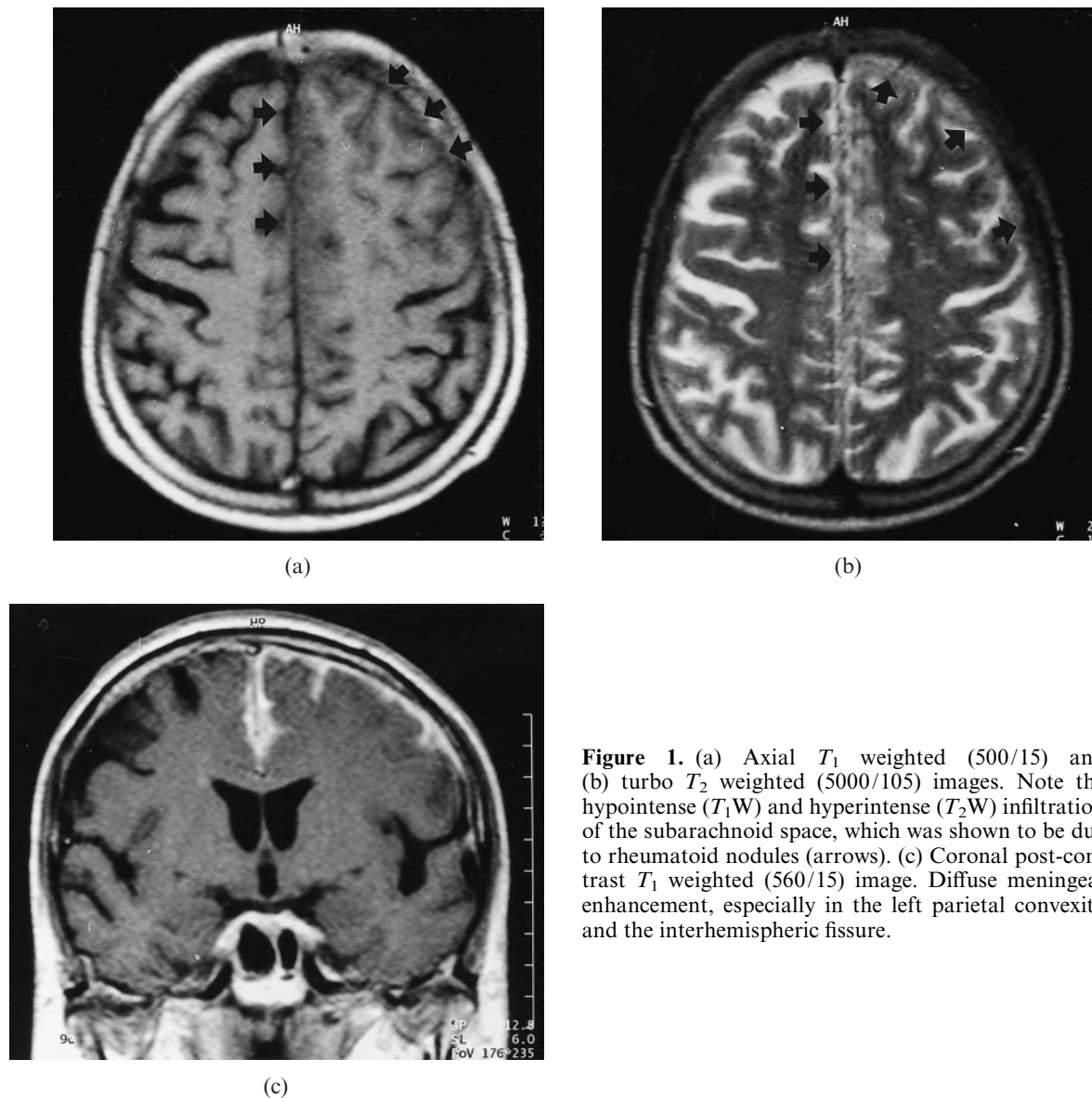


Figure 1. (a) Axial T_1 weighted (500/15) and (b) turbo T_2 weighted (5000/105) images. Note the hypointense (T_1W) and hyperintense (T_2W) infiltration of the subarachnoid space, which was shown to be due to rheumatoid nodules (arrows). (c) Coronal post-contrast T_1 weighted (560/15) image. Diffuse meningeal enhancement, especially in the left parietal convexity and the interhemispheric fissure.

pachymeningitis was established and a new steroid regimen was prescribed for the patient.

Discussion

The causes of pachymeningitis are many and include RA, sarcoidosis, tuberculosis, Wegener's granulomatosis, syphilis and other infections [4]. Clinically apparent rheumatoid pachymeningitis is an unusual complication of RA, but there have been reports of central nervous system involvement diagnosed on biopsy [2].

Contrast enhancement of the meninges on MRI is a non-specific finding seen in autoimmune, bacterial, fungal or viral inflammatory processes. The diagnosis of rheumatoid pachymeningitis requires the correlation of pathological, laboratory and clinical data, and radiological findings. Most of these manifestations result from rheumatoid

nodules and may be due to hyperviscosity syndrome. Some neurological complications in RA appear to be related to vasculitis [3].

MRI is very effective at demonstrating meningeal pathology. Cranial pachymeningitis is shown as marked thickening of the meninges (Figure 1). In the case presented, meningeal thickening with enhancement was seen on MRI (Figure 1c). Allison and Marano [7] showed marked enhancement of the tentorium on CT in a patient with RA. Shuntani et al demonstrated marked unilateral enhancement of the dura and falx cerebri on MRI, suggestive of inflammation, in a 72-year-old man in whom the exact cause of pachymeningitis was unknown, although RA or other collagen disease could not be excluded [4]. Higgins et al attributed an increase in gallium uptake at 24 h in the tentorium cerebelli and posterior aspect of the superior sagittal sinus to the chronic inflammatory

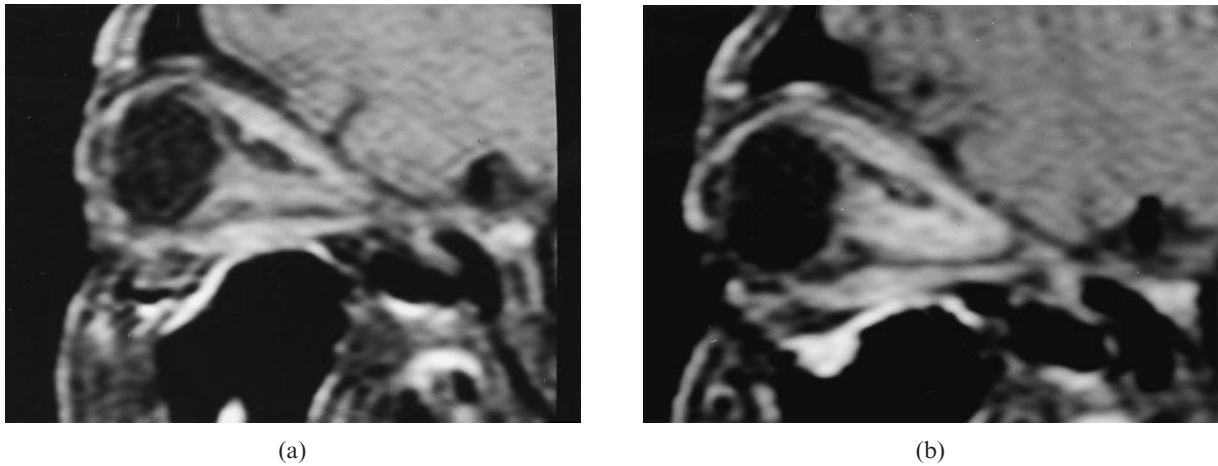


Figure 2. Sagittal fat-suppressed T_1 weighted images (680/15) of (a) the right and (b) the left optic nerve after gadolinium injection. Notice left optic nerve thickening and enhancement when compared with the right optic nerve.

changes seen in a case of rheumatoid pachymeningitis [2]. Otsuka et al presented a 53-year-old woman with RA and intermittent headache, ptosis and diplopia. MRI demonstrated hypertrophic dura extending from the falx cerebri to the tentorium cerebelli which was enhanced markedly by Gd-DTPA. The clinical features, pathological and MRI findings were consistent with rheumatoid pachymeningitis. They emphasized that hypertrophic pachymeningitis should be considered as a diagnostic possibility in RA patients with prolonged headache and recommended MRI with gadolinium in such cases [8]. Nishikawa et al reported a 71-year-old woman with rheumatoid pachymeningitis who presented with headache, vertigo and dizziness. She had hypertrophic masses in the meninges of the left cerebellar tentorium, cerebellopontine angle and fourth ventricle on MRI. Her neurological status improved with steroid and azathioprine treatment [9].

Optic neuritis classically refers to the inflammation, infection or demyelination of the optic nerve. Vasculitis is often discussed as a cause of optic neuritis but remains controversial in its significance [5]. RA is a rare cause of this condition. In our case, there was significant thickening and enhancement of the left optic nerve on MRI and this was consistent with perioptic neuritis (Figures 2a and b). Shintani et al presented cranial pachymeningitis cases associated with previous optic nerve involvement [4]. Weinstein et al reported a 42-year-old man with a 15-year history of RA in whom rheumatoid pachymeningitis caused a compressive optic neuropathy [10]. Sklar et al reviewed nine patients with optic neuropathy and vasculitis. MRI of one of these patients, with RA systemic necrotizing vasculitis and visual loss, showed enhancement and enlargement of the intracanalicular and intracranial portions of the

optic nerve and optic chiasm on the clinically affected side [5].

Enhancement represents disruption of the blood–brain barrier within the optic nerve, thus MRI with gadolinium and fat suppression should be performed in cases of optic neuropathy [5]. Some authors do not consider the fat suppression technique to be the primary mode of imaging in patients with optic neuritis, since standard proton density and T_2 weighed spin echo acquisitions are extremely sensitive for areas of demyelination. However, fat suppression on MRI may be useful in cases of isolated optic neuritis without cranial involvement [11]. A higher sensitivity on MRI detection of optic neuritis using inversion recovery sequences with short inversion times has also been reported [6].

Sklar et al postulated that disruption of the blood–brain barrier in RA was vasculitic in nature and might be histopathologically similar to systemic lupus erythematosus and radiation-induced vasculitis [5]. Shintani et al reviewed 12 cases of cranial pachymeningitis associated with optic nerve involvement. Three of these had RA, and although multiple cranial nerve involvement was present in the patients with other conditions, interestingly all three RA patients had no other cranial nerve involvement as in the present case. Involvement of the optic nerve may be caused either by constriction of the nerve through granulomatous thickening in the optic nerve canal or by direct inflammatory cell invasion of the optic nerve [4, 10]. In the reported case, involvement of the optic nerve was thought to be secondary to direct inflammatory cell invasion of the nerve.

Although pachymeningitis and perioptic neuritis are rare neurological complications of RA, MRI is essential in the evaluation of such cases.

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Book review

Essentials of Nuclear Medicine (2nd Edn). Ed. by M V Merrick, pp. 334, 1998 (Springer Verlag, Heidelberg), £55.00
ISBN 3540762051

This single author text gives a broad overview of clinical nuclear medicine in 334 pages and 12 chapters. Coverage of the topic is wide and, therefore, inevitably, not “in depth”, but focuses primarily on techniques and clinical applications.

Imaging, non-imaging and therapeutic nuclear medicine are all covered, and PET is mentioned, briefly, where appropriate. Some readers may be disappointed that more about PET has not been

included; however, I think that the balance achieved in the book accurately reflects the clinical use of nuclear medicine in the UK.

The final chapter entitled *Scientific considerations—selected topics* includes an important reminder about the quality assurance aspects of nuclear medicine.

The text reflects the author’s own firmly held views, some of which would certainly give rise to lively debate! For this reason, I think that the book can best be recommended for clinicians experienced in nuclear medicine rather than new recruits.

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