# Rheumatoid pachymeningitis

**H J Tan, A A Raymond, P P Phadke, Z Rozman**


## ABSTRACT

**Symptomatic rheumatoid pachymeningitis is a rare extra-articular manifestation of rheumatoid arthritis. Clinical symptoms are non-specific and diagnosis is frequently made by exclusion. We present a 61-year-old woman with a 9-year history of rheumatoid arthritis presenting with deafness and progressive disability over a two month duration. She was diagnosed as having rheumatoid pachymeningitis based on the cerebral magnetic resonance imaging findings.**


**Keywords: internal auditory canals, magnetic resonance imaging, rheumatoid arthritis, rheumatoid pachymeningitis, sensorineural deafness**

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## INTRODUCTION

Rheumatoid arthritis is a multisystem disease with many extra-articular manifestations. The pathogenesis of rheumatoid arthritis involves an immune process characterised by immunoglobulins, IgM and IgG rheumatoid factors. Part of the localised immune process has been implicated in the development of rheumatoid meningitis. Rheumatoid meningitis is a pachymeningitis that is rarely encountered in rheumatoid arthritis. This is the first case of rheumatoid pachymeningitis presenting with bilateral sensorineural deafness. Brain imaging revealed enhancement of the tissues in both internal auditory canals.

## CASE REPORT

A 61-year-old woman was hospitalised for progressive disability of two months duration. She had a 9-year history of rheumatoid arthritis involving the small joints of her hands, wrists and ankles. She had occasional disease exacerbation for which she was prescribed analgesics and methotrexate. She had initially complained of hearing problems to her family. She slowly became forgetful and confused in the following two months, and she was not able to perform her routine daily activities. There was no headache, fever, nausea or vomiting.

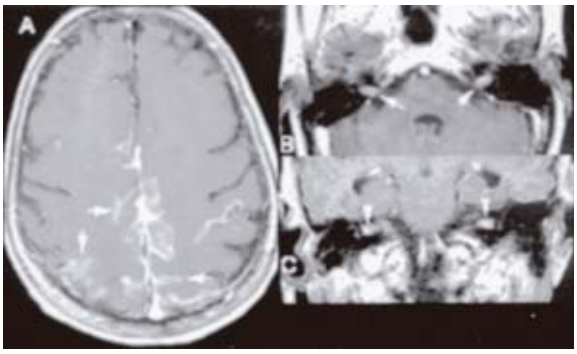


**Fig. 1** Enhanced (a, b) axial and (c) coronal (C) T1-W MR images show enhancement in the convexity sulci (arrows in a) and the interhemispheric fissure, and in both the internal auditory canals (arrows in b & c).

On physical examination, there were bilateral symmetrical deforming polyarthropathies of her hands. The neurological examination revealed bilateral sensorineural deafness. The mini-mental state examination could not be done, as the patient did not understand the instructions. Her muscle bulk, power and tendon reflexes were normal. Plantar responses were flexor. The examination of the heart, lungs and abdomen was normal.

The rheumatoid factor was positive. Antinuclear antibodies and VDRL were negative. The cerebrospinal fluid results were abnormal (Table I). The Gram stain, Indian ink stain, Ziehl-Neelson stain and latex agglutination tests were negative. Mycobacterium and fungal cultures were negative. Brain magnetic resonance (MR) imaging showed prominent periventricular areas of hyperintensity in the parieto-occipital and periaqueductal regions. There was thick meningeal enhancement in the posterior interhemispheric fissure and enhancement of both internal auditory canals (Fig. 1).

## DISCUSSION

Neurological manifestations in rheumatoid arthritis, although not very common, causes death in up to 18.6% of cases[1]. Documented nervous system manifestations of rheumatoid arthritis include spinal cord compression due to subluxation of the cervical vertebrae, cerebral haemorrhage due to

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serum hyperviscosity associated with the elevated titres of circulating rheumatoid factors, vasculitis, rheumatoid meningeal nodules, choroid plexus infiltration, rheumatoid synovial infiltration of the spinal dura and pachymeningitis[2]. Other causes of pachymeningitis include sarcoidosis, tuberculosis, lymphoma, Wegener's granulomatosis and syphilis[1]. Short of doing a brain biopsy, the diagnosis of rheumatoid pachymeningitis is one of exclusion. In this patient, the entire infective screen was negative.

Rheumatoid pachymeningitis is a rare occurrence in rheumatoid arthritis, and previous reported experience with this complication is limited to case reports. Both fibrinoid deposits and rheumatoid nodules cause pachymeningitis. Its clinical course is one of longstanding, erosive and seropositive disease with prominent extra-articular features. There may or may not be clinical evidence of active synovitis when the neurological disease becomes symptomatic. Similarly, this patient who had long-standing rheumatoid disease did not have active arthritis during her neurological presentation. Although patients with pachymeningitis may remain asymptomatic, several case reports have described its clinical manifestations. They include seizures[3], diffuse cerebral dysfunction[4], headaches, meningitis, cortical blindness.

Our patient presented with confusion and sensorineural deafness. This could be attributed to the thick meningeal lesions, seen as areas of hyperintensity on MR imaging, involving the periaqueductal region, parieto-occipital gray and white matter, and both internal auditory canals. The diffuse meningeal involvement could be a combination of pachymeningitis and vasculitis. The cerebrospinal fluid may show some combination of high protein, mild pleocytosis and hypoglycorrhachia, as seen in our patient. Extremely low glucose content is a well-known characteristic of rheumatoid effusions. The mechanism of hypoglycorrachia in rheumatoid meningitis is due to interference with glucose entry[5]. This differs from the non-rheumatoid meningeal diseases, such as in infections and carcinomatous meningitis where certain inflammatory products interfere with the glucose transport system into the cerebrospinal fluid. Cerebral MR imaging in this patient showed focal hyperintensity in the parieto-occipital region, periaqueductal region and the internal auditory canals, with prominent enhancement. Previous case reports evaluating rheumatoid pachymeningitis with radiological examination include gallium scintigraphy, technetium scintigraphy, enhanced computed tomography and MR imaging[2].

**Table 1. Summary of laboratory results.**

| | | Normal range | |
|---|---|---|---|
| Haemoglobin | 10.5 $\times 10^9$ | (14.0-17.0) | g/dL |
| White cell count | 4.0 $\times 10^9$ | (4.0-10.0) | /L |
| Platelet count | 133 $\times 10^9$ | (150-400) | /L |
| Urea | 4.3 | (2.5-6.4) | mmol/L |
| Sodium | 127 | (135-150) | mmol/L |
| Potassium | 3.5 | (3.5-5.0) | mmol/L |
| Creatinine | 63 | (62-133) | μmol/L |
| Random glucose | 6.4 | (3.0-6.7) | mmol/L |
| Albumin | 28 | (35-50) | g/L |
| Protein | 56 | (67-88) | g/L |
| Total bilirubin | 9 | <23 | μmol/L |
| Alanine transaminase | 23 | (<44) | U/L |
| Alkaline phosphatase | 103 | (32-104) | U/L |
| Erythrocyte sedimentation rate | 10 | (1-20) | mm/hour |
| C-reactive protein | 2.40 | <0.5 | mg/dL |
| Cerebrospinal fluid | | | |
| – protein | 3768 | (150-450) | mg/L |
| – glucose | 0.2 | (2.2 -4.2) | mmol/L |
| – microscopic examination | white blood cells, occasional lymphocytes and neutrophils | 150 | cells/ml |

Methotrexate has long been a recognised treatment of rheumatoid vasculitis. The long-term use of methotrexate in this patient may predispose to accelerated nodulosis. Karam et al[6] reported isolated rheumatoid nodulosis of the meninges in a patient with well-controlled, non-deforming rheumatoid arthritis during methotrexate therapy. Although this patient did not have any rheumatoid nodules, the possibility of methotrexate-induced meningeal rheumatoid nodulosis could not be excluded. Pathologically, the dura has single or multiple rheumatoid nodules, or may show diffuse inflammatory changes consisting of a central area of fibrinoid necrosis surrounded by combination of pallisading epitheloid cells, lymphocytes, plasma cells and multinucleated giant cells. Vasculitis of the leptomeninges and underlying cortex may occasionally be present.

The diagnosis of rheumatoid pachymeningitis is ominous as most patients die within six months of onset of neurological symptoms. The use of corticosteroids and immunosuppressants has not been shown to have any significant effect on this condition[7]. In summary, rheumatoid pachymeningitis should be considered as a diagnostic possibility in

the proper clinical setting, when evidence of abnormal meningeal enhancement is found on enhanced MR imaging.

## REFERENCES


1. Agildere AM, Tutar NU, Yucel E, Coskun M. Case report. Pachymeningitis and optic neuritis in rheumatoid arthritis: MRI findings. Br J Radiol 1999; 72:404-7.
2. William TC, Drew JM, Rizzo M, Ryals TJ, Sato Y, Bell WE. Evaluation of pachymeningitis by contrast-enhanced MR imaging in a patient with rheumatoid disease. AM J Neuroradiol 1990; 11:1247-8.
3. Beck D, Corbett JJ. Seizures due to central nervous system rheumatoid meningovasculitis. Neurology 1983; 33:1058-61.
4. Spurlock RG, Richman AV. Rheumatoid meningitis: a case report and review of the literature. Arch Pathol Lab Med 1983; 107:129-31.
5. Markenson JA, McDougal JS, Tsairis P, Lockshin MD, Christian CL. Rheumatoid meningitis: a localized immune process. Ann Int Med 1979; 90:786-9.
6. Karam NE, Roger L, Hankins LL, Reveille JD. Rheumatoid nodulosis of the meninges. J Rheumatol 1994; 21:1960-3.
7. Rosenbaum RB, Campbell SM, Rosenbaum JT. Clinical Neurology of Rheumatic Diseases. Newton: Butterworth-Heinemann,1996.