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PACHYMENINGITIS AS AN ATYPICAL INITIAL MANIFESTATION OF RHEUMATOID ARTHRITIS: A CASE REPORT.

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BACKGROUND

Rheumatoid arthritis (RA) can rarely present with neurological symptoms in the absence of typical joint manifestations. We report a case with an unusual clinical onset characterized by pachymeningitis, in which RA emerged as a possible underlying etiology based on a comprehensive diagnostic workup.

CASE DESCRIPTION

A 73-year-old man with a history of hypertension and pulmonary fibrosis treated with nintedanib, presented with an acute episode of disorientation and confusion lasting approximately ten days, without fever or other systemic symptoms, which resolved completely without treatment. Over the previous year, he had experienced mild verbal recall difficulties and occasional instability, without gait or sphincter disturbances. Neurological examination at the time of evaluation was unremarkable. During the acute episode, cognitive testing using MoCA revealed mild language and delayed recall deficits (score 23/30). Brain MRI showed multiple aspecific gliotic lesions in the bilateral supratentorial white matter and diffuse pachymeningeal thickening. A follow-up contrast-enhanced MRI three months later confirmed persistent diffuse pachymeningeal thickening with marked enhancement. CSF analysis showed elevated protein, a few oligoclonal bands with a mirror pattern, and normal levels of total tau, p-tau, and beta-amyloid 1-42. Laboratory findings revealed positive lupus anticoagulant, anti-cyclic citrullinated peptide (anti-CCP) antibodies, and elevated IgG4. Tumor markers showed a weakly positive CA19-9, but no clinical or imaging evidence of malignancy. Tests for anti-onconeural antibodies, anti-NMDAr, anti-VGKC (in serum and CSF), viral markers, ACE levels, CSF cytology, EEG, and salivary gland and fat biopsies were all negative. Neuropsychological testing at four months was normal. Given the clinical picture, imaging findings, and autoimmune markers, a working diagnosis of rheumatoid pachymeningitis was proposed. The patient was treated with oral steroids followed by Rituximab, resulting in clinical stabilization and radiological improvement. A six-month follow-up MRI showed clear improvement with a marked reduction in pachymeningeal thickening.

OCTOBER 2024 (ACUTE PHASE)

- **Acute episode of disorientation and confusion for 10 days**
- **MoCA in acute**: mild language and delayed recall deficits (score 23/30)
- **MRI**: multiple aspecific gliotic lesions in the bilateral supratentorial white matter and **diffuse pachymeningeal thickening**

DIAGNOSTIC PATHWAY

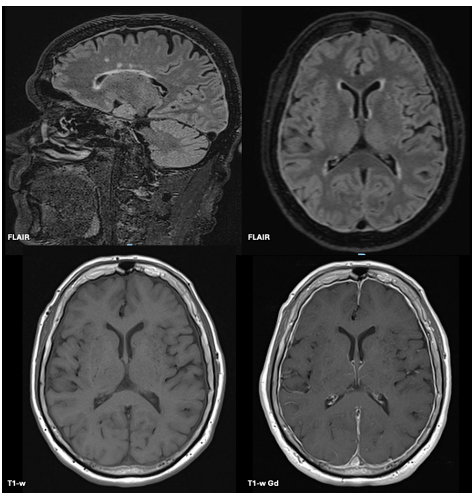
- **MRI 3 months later**: diffuse pachymeningeal thickening with marked enhancement
- CSF: **elevated protein, a few oligoclonal bands with a mirror pattern (normal levels of** tau, p-tau, and beta-amyloid 1-42)
- Serum: **positive LAC and anti-CCP; elevated IgG4.**
- Others test negative

DIAGNOSIS & TREATMENT

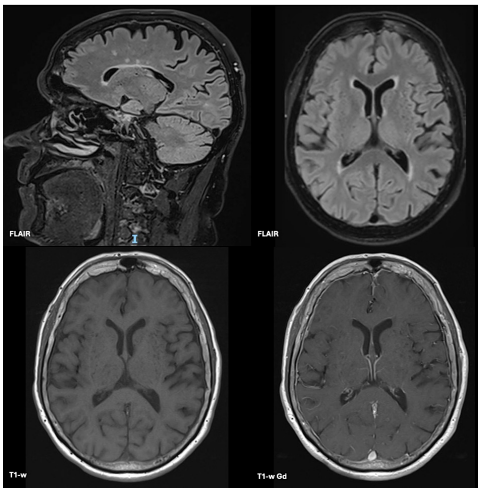
- **RHEUMATOID PACHYMENINGITIS**
- Oral **steroids followed by Rituximab iv**

FOLLOW-UP

- Neuropsychological tests 4 months later: negative
- **MRI 6 months later:** improvement with a marked reduction in pachymeningeal thickening.



October 2024



April 2025

CONCLUSION

THIS CASE PRESENTS **A RARE AND ATYPICAL FORM OF RHEUMATOID ARTHRITIS WITH ISOLATED NEUROLOGICAL SYMPTOMS, RAISING THE POSSIBILITY OF RHEUMATOID PACHYMENINGITIS.**
IT HIGHLIGHTS THE NEED FOR ONGOING EVALUATION AND A BROAD DIFFERENTIAL IN PATIENTS WITH UNEXPLAINED PACHYMENINGITIS AND IMMUNE ACTIVATION.

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