Pachymeningeal Thickening in Rheumatoid Arthritis

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Though infrequent, lepto- [1] and pachymeningeal fibrosis may develop secondary to a diversity of disorders or manifest in the idiopathic form.

Here we present a 56-year-old patient who developed pachymeningeal thickening presumably due to pachymeningitis after a 15-year history of seropositive erosive nodular rheumatoid arthritis (RA).

He presented a progressive left hearing loss, swallowing difficulties and dysphonia in the absence of headache over the last 4 years.

Although fundus oculi were normal and visual fields full, there was marked right proptosis with downward and outward deviation accompanied by facial hyperesthesia with paresis of the masticatory muscles, lack of palate excursion with absence of the gag reflex and vocal chord paralysis, all on the right side. The only relevant finding on the contralateral side was a sensorineural hearing loss and vestibular paresis.

Erythrocyte sedimentation rate was elevated and X-rays of hands and cervical spine were consistent with RA changes. CT scan
showed dural enhancement with hypertrophy mainly in tentorium cerebelli and falx suggestive of thickened meninges and compatible with pachymeningitis. However, although meningeal biopsy showed marked thickening, microscopy disclosed fibrous tissue with no signs of inflammation, vasculitis or caseating granulomas. MRI confirmed meningeal thickening and pinpointed hypointense T1-hyperintense T2-weighted bilateral thalamic foci consistent with ischemia (Fig. 1, 2). No treatment was given and at 3 years’ follow-up the patient’s condition was unchanged.

Central nervous system involvement in RA is indeed rare [2] and only 17 cases of pachymeningitis have so far been reported [2-6]. Neurological evaluation including gallium and technetium scintillography, CT scanning and MRI were only available in 4 of the above [2-5]. Although at necropsy most cases of RA associated with pachymeningitis disclosed rheumatoid nodules throughout the dura [6], such information may be missed when only biopsy material is available.

Recognized etiological factors such as trauma [7], toxins [7], metabolic disorders [8] and infections including tuberculosis [9], syphilis [9] and fungi [10] were carefully searched for but found absent in our patient, so that the meningal involvement was reliably ascribed to RA.
The absence of headache which has characterized most cases of pachymeningitis whatever its etiology and the spontaneous arrest in clinical progress are noteworthy. Though several therapies have been employed to date, steroids seem the only effective medication, but only to alleviate headache since fibrotic reaction proceeds unchecked [1]. In contrast, no treatment was given to our patient, and at 3 years’ follow-up the overall clinical picture remained unchanged.

References


Michel/Scorticati/Pikkelby/Zarra/Gatto Pachymeningeal Thickening in Rheumathoid Arthritis