Keeping with emerging clinical evidence for effectiveness, the decision was made to proceed with anti-TNF therapy, in view of persistent active disease (DAS-28 of 8.3), a decision that was in keeping with the British Society for Rheumatology (BSR) guidelines [1]. However, in view of previous malignancy, and in keeping with the British Society for Rheumatology (BSR) guidelines [1], anti-TNF therapy was chosen in preference to anti-TNF therapy because of previous malignancy, and in keeping with the British Society for Rheumatology (BSR) guidelines [1].

Despite the high prevalence of cervical spine involvement in RA, neurological deficits are uncommon. Cervical spine involvement in RA may occur in up to 80% of cases [3, 4]. Risk factors include extensive joint involvement, high seropositivity, male gender, vasculitis and prolonged use of corticosteroids [4-6]. The most common deformities affecting the cervical spine are atlantoaxial subluxation, vertical subluxation leading to superior migration of the odontoid peg ('cranial settling') and subaxial subluxation [3]. Compression of the spinal cord, brainstem or cranial neuropathies can result from subluxation of the spine or direct pressure by a synovial pannus.

Despite the high prevalence of cervical spine involvement in RA, neurological deficits are reported in only 7–34% of patients [4, 7]. Cranial nerve palsies involving CNs IX, X and XII have been reported only rarely in patients with RA. Most of the cases have been secondary to nerve compression at the base of the skull by vertical [8, 9] or horizontal [10] subluxation at the atlantoaxial joint. Cranial neuropathy related to compression by a rheumatoid pannus has only been reported once before and involved the CN XII bilaterally [8]. Our patient had unilateral CNs IX and XII cranial neuropathy due to compression by the inflammatory pannus. To our knowledge, this is the first case reported where cranial neuropathy resolved with medical and safety of this approach after rituximab [2], in accordance with the patient’s wishes, and just before her developing neurological symptoms.

At the time of presentation, examination revealed reduced sensation of the left posterior oropharynx [indicating glossopharyngeal nerve, cranial nerve (CN) IX, dys-function] and tongue deviation to the left indicating a left hypoglossal nerve (CN XII) palsy. There was normal symmetrical movement of the uvula (i.e. normal vagus nerve, CN X, function), no dysphonia and a good cough impulse. There were no long tract signs. Active synovitis involved several joints (DAS-28 of 8.45).

Investigations showed thrombocytosis, normal biochemistry and an elevated CRP of 67 mg/l. Cervical spine X-ray revealed a marked erosion of the odontoid process and anterior subluxation of 6 mm on forward flexion. MRI showed a large inflammatory pannus measuring ~17 mm at the atlantoaxial joint, extending to the left into the region of the hypoglossal canal (Fig. 1). There was associated atlantoaxial subluxation, but no myelopathic signal change within the spinal cord. A recent mammogram and isotope bone scan had both been negative for cancer recurrence.

Treatment with i.v. methylprednisolone 1 g daily for 3 days improved her symptoms and signs. Therefore, it was felt that neurosurgical intervention was not indicated. Etanercept 50 mg s.c. once a week was commenced soon after. On clinic review after 8 weeks, the function of the glossopharyngeal nerve had returned to normal, and her swallowing and speaking difficulties had resolved. She had minimal residual tongue deviation to the left. There was a significant reduction in the DAS-28 to 4.3. This improvement was maintained at 6 months and no infections were observed. A repeat MRI scan confirmed moderate reduction in the size of pannus.

Cervical spine involvement in RA may occur in up to 80% of cases [3, 4]. Risk factors include extensive joint involvement, high seropositivity, male gender, vasculitis and prolonged use of corticosteroids [4-6]. The most common deformities affecting the cervical spine are atlantoaxial subluxation, vertical subluxation leading to superior migration of the odontoid peg ('cranial settling') and subaxial subluxation [3]. Compression of the spinal cord, brainstem or cranial neuropathies can result from subluxation of the spine or direct pressure by a synovial pannus.

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References
management, which involved induction of disease remission with high-dose i.v. corticosteroids and maintenance of disease control with anti-TNF therapy. In conclusion, lower cranial nerve palsies in rheumatoid patients should raise suspicion of cervical spine involvement. In our case, aggressive medical management has obviated the need for neurosurgery and this approach might be worth trying in similar cases before subjecting patients to surgical intervention. Furthermore, it is important to consider anti-TNF therapy early on in patients with aggressive RA and successfully treated malignancy.

Rheumatology key message

- Cranial neuropathy due to rheumatoid pannus can respond to aggressive medical management.

Disclosure statement: The authors have declared no conflicts of interest.

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References


Comment on: Longitudinal examination with shoulder ultrasound in patients with polymyalgia rheumatica

Sir, We read with interest the recent work of Macchioni et al. [1] on the usefulness of ultrasonography (US) and power Doppler US (PDUS) in PMR to detect at diagnosis the