MASTERCLASS *Clinical cases from the Royal National Hospital for Rheumatic Diseases*

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Management of rheumatoid neck

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Rheumatoid arthritis of the neck is common but neurological complication occurs only in the minority. Nevertheless, this represents one of the few rheumatological emergencies. Surgery is the only definitive treatment, but diagnosis of this complication is often delayed so that candidates for surgery are frail and disabled, thus jeopardising the outcome. The advent of new forms of imaging, especially magnetic resonance imaging (MRI), when combined with careful clinical examination potentially offers a more efficient way of selecting patients for surgery. This is illustrated in the following case.

Case report

A 55 year old man was admitted to hospital with weakness of the right arm and difficulty in walking, which was interfering with his job as a messenger.

Eighteen years before, he developed sero-positive rheumatoid arthritis which from the beginning affected most of his peripheral joints. The arthritis followed a persistent and slowly progressive course. He had been treated almost continuously throughout with penicillamine 500 mg daily and low dose prednisolone (4-6 mg daily). He remained at work but required extensive joint surgery, which included silastic implants of the metacarpophalangeal joints, bilateral total hip replacements, and bilateral ankle arthroplasties. A year before admission insidiously he developed paraesthesia, first in the right arm and then bilaterally. This was aggravated by neck flexion. Shortly before admission he developed weakness of his hand and difficulty in walking and he complained of 'jumping' of his legs.

In addition to widespread rheumatoid deformities and subcutaneous nodules on both elbows, examination showed weakness of the right hand and loss of sensation to pin prick in the distribution of the C6/7 dermatomes bilaterally. There was hyperreflexia at the knees and ankles with bilateral extensor plantar responses. The plain radiograph (fig 1A) showed subluxation of C6 on C7 with narrowing of the disc space and fusion of the facet joints of C3 to C6 above this level. There was a minor degree of atlantoaxial subluxation. Magnetic resonance imaging was then performed from the posterior

fossa down to T4. T1 weighted (fig 1B) and T2 weighted MAST sagittal images were obtained together with T1 weighted axial images (fig 1C). This confirmed subluxation at the C6/7 level with severe restriction of the thecal sac. In addition there were subaxial subluxations at other levels, particularly obvious on the T2 weighted sagittal views. The axial views showed no evidence of myelomalacia. There was erosion of the odontoid peg but no significant subluxation at this level. The increased signal from the vertebral bodies on T1 weighted images shown in Fig 1B is due to fatty replacement of the marrow, which is typical of fused vertebrae.

Discussion

In the current state of knowledge, the combination of careful clinical examination, a single plain radiograph showing a lateral view of the neck in flexion, and MRI is sufficient to indicate the patient in need of surgery and to provide sufficient information for the neurosurgeon. Magnetic resonance imaging represents a major advance in the imaging of the cervical spine.^{1 2} The pathological anatomy of rheumatoid disease can be defined, including soft tissue inflammation and pannus formation (figs 2A and B), erosions, and subluxations. T1 weighted images show the spinal cord which can be visualised throughout its length. Axial views of the cord at the site of subluxation may indicate irreversible change such as myelomalacia. T2 weighted views demonstrate occlusion of the subarachnoid space by providing a myelogram-like image. Magnetic resonance imaging therefore replaces myelography, which is often an unpleasant experience for the patient, especially when performed by cisternal puncture. Next best is computed tomography enhanced by contrast introduced through a lumbar puncture.^{3 4}

A careful clinical examination, however, is indispensable. Subluxations, most commonly atlantoaxial, can be shown in about 25% of patients with rheumatoid arthritis, but only 5% of these develop myelopathy.⁵ There is a poor correlation between the degree of subluxation and neurological complications and, consequently, surgery is not indicated as a prophylactic measure. Those patients most likely to

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Figure 1 (A) Plain radiograph showing subluxation of C6 on C7 with narrowing of the disc space and fusion of the facet joints of C3 to C6 above this level. (B) T1 weighted MAST sagittal image showing subluxation at the C6/7 level with severe restriction of the thecal sac. (C) T1 weighted axial image of the C6/7 level.

have progressive subluxation, like the case described here, have severe erosive disease of peripheral joints, seropositivity and rheumatoid nodules, and a history of chronic corticosteroid usage.⁶⁻¹⁰ Men tend to be more commonly affected. It is necessary to maintain vigilance, therefore, especially in such cases. Symptoms are often insidious in onset, starting most often with paraesthesia, numbness, and sensations of hot and cold, most commonly affecting the hands.¹¹ Sometimes this happens in a 'glove-and-stocking' distribution and is misdiagnosed as a peripheral neuropathy. Spastic paraparesis often resulting in the complaint that 'the legs won't go' may also be overlooked. This combined with the fact that neurological examination is often difficult in a patient with severe rheumatoid deformities means that there is often a delay in diagnosing myelopathy. This in turn reduces the chances of a good surgical result.

Clinical examination is also important in defining the major site of cord compression: often the plain radiograph shows multiple subluxations ('stepladder effect'). Similarly, MRI may show several occlusions of the subarachnoid space. For technical reasons, however, the T2 weighted image can exaggerate the appearance of cord compression. Therefore, the demonstration of a clear-cut C6/7 nerve root lesion in the case described here was crucial for pinpointing the appropriate level for surgery.

The indications for surgery are severe neurogenic pain and clinical evidence of myelopathy. The procedure entails a combination of decompression and fusion. The constraints of surgery are the common frailty of the patient (if decompression and fusion is done at one sitting, surgery takes about four hours), bone which is often osteoporotic making stabilisation very difficult, and irreversible neurological damage. Halo traction is an effective external means of

Figure 2 (A) T1 weighted image showing soft tissue mass at atlantoaxial level. (B) T2 weighted image showing high water content of inflammatory pannus and occlusion of the subarachnoid space by providing a myelogram like image.

reducing subluxation, but this is generally not required preoperatively. Patients are often a major anaesthetic risk and the combination of reduced opening of the mouth and problems in the neck makes intubation difficult. Fiberoptic intubation helps to overcome this.

Surgery in each case has to be considered individually. For atlantoaxial subluxation, posterior stabilisation with wire or a combination of bone and wire is usually performed together with decompression of the craniocervical junction if necessary. An effective way of accomplishing the latter is transoral removal of the odontoid peg and associated inflammatory tissue as advocated by Crockard *et al.*¹²

The surgical technique for subaxial subluxations is more controversial. The usual finding, as in the case described here, is forward displacement of the upper on the lower vertebral

body. In our experience, techniques of posterior stabilisation using, for example, a Hartshill rectangle, which are effective in other causes of subluxation, are less suitable for rheumatoid patients because of the poor quality of the bone. We favour an anterior approach to decompression and fusion. In the above case, with significant subluxation at one level only, the relatively conservative Cloward's procedure¹³ was carried out using a dowel bone graft. When subluxations extend to more than two levels a more extensive operation is required, in which a trench is cut anteriorly into the bodies of the cervical vertebrae and filled with a bone graft. Postoperatively, the patient is mobilised early and initially provided with a firm collar.

With early surgical intervention and improved anaesthetic and perioperative technique, postoperative mortality is less than 10% and improvement of neurological function and loss of pain are to be expected.^{14 15} This was the case in the patient described here, who was able to return to work. Even successful surgery does not cure the underlying process which, if progressive, will lead to further problems, usually at a different site. Subsequent surgical procedures may then be needed, each requiring more ingenuity and improvisation.

At the present time, however, there is no alternative to surgery. Collars do not prevent progression of subluxation and, once myelopathy has developed, the prognosis in the absence of intervention is very poor.¹¹

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