CERVICAL CORD COMPRESSION DUE TO EXTRADURAL GRANULATION TISSUE IN RHEUMATOID ARTHRITIS

A REVIEW OF FIVE CASES

HIROSHI KUDO, KUNIO IWANO, HISAYOSHI YOSHIZAWA

From Sagamihara National Hospital, Kanagawa

Five patients with long-standing, severe rheumatoid arthritis who developed cervical myelopathy at the subaxial levels are presented. In each patient the myelopathy occurred in the absence of major subluxation. At laminectomy the cause of the cord compression was found to be a band-like mass of ligamentous and granulation tissue in the posterior half of the extradural space, extending to the underlying portion of the dura and forming a constricting ring. In three patients this constricting ring was released by longitudinal division of the dura followed by application of a fascial patch graft. This release seems to be essential for effective decompression of the cord and good neurological recovery was achieved in these three patients. Of the other two patients, the result was fair in one and poor in the other, this latter result being due to severe intra-operative bleeding with a massive wound haematoma.

Cord damage caused by atlanto-axial subluxation in patients with rheumatoid arthritis has been well documented (Conlon, Isdale and Rose 1966; Cregan 1966; Crelin, Maccabe and Hamilton 1970; Stevens et al. 1971; Smith, Benn and Sharp 1972; Nakano et al. 1978). Reports of myelopathy at lower spinal levels are, however, relatively scarce, and most describe only cases in which the myelopathy occurred as a consequence of major subluxation.

In this report we describe five cases in which subaxial cervical myelopathy occurred with only minor subluxation. In each the cause of the cord damage was found to be compression by extradural granulation tissue, probably of rheumatoid origin.

CLINICAL MATERIAL

The clinical details of the five patients are summarised in Table I. Their age at the onset of neural symptoms ranged from 56 to 67 years, with an average of 59 years. All five patients fulfilled the diagnostic criteria of classical rheumatoid arthritis, as defined by the American Rheumatism Association and all were seropositive. The duration of the rheumatoid disease varied from 10 to 35 years, with an average of 22 years. All five patients had severe contractures and deformities in most of the peripheral joints, and three of them had typical opera-glass deformities of their hands.

Neurological findings. All five patients developed numbness and weakness in the upper limbs first, and these symptoms gradually progressed to the lower limbs. None gave a history of any relevant injury or other incident to account for these symptoms. At the time of admission all were unable to stand or walk and were in fact completely bedridden.

In all five patients examination of sensibility revealed areas of definite hypo-aesthesia and hypo-algesia in all four extremities as well as in the trunk. Muscle paralysis was more severe in the upper than in the lower limbs. There was a moderate to marked increase of the deep tendon reflexes in the upper limbs and the knee and ankle jerks also were increased. In no patient could patellar clonus be elicited but in four there was ankle clonus. In all five patients the plantar responses were extensor. Two patients (Cases 3 and 4) had complete urinary incontinence, and one (Case 2) had partial incontinence.

Radiographic findings (see Figs 1, 5, 6 and 10). Lateral radiographs of the cervical vertebrae showed minor subluxation (2 mm or less) at one of the subaxial levels in every patient. At the level of the subluxation severe erosive changes were seen in the facet joints in most of the patients. Radiographs taken in flexion and extension revealed instability of the cervical spine in only one patient (Case 2), in whom an anterior slip of 4 mm was demonstrated in flexion. At the atlanto-axial level no patient showed instability.

Myelography (see Figs 2, 7 and 11). In all patients a myelographic block was seen: four patients showed a partial block (Figs 10 to 12) and one patient a complete block. This myelographic block occurred at exactly the same level as the minor subluxation and the severe
Table 1. Clinical details of the five patients

<table>
<thead>
<tr>
<th>Case</th>
<th>Sex</th>
<th>Age (years)</th>
<th>Duration of RA (years)</th>
<th>Duration of neural symptoms (months)</th>
<th>Level and degree of subluxation</th>
<th>Myelo- graphic block</th>
<th>Neurological status</th>
<th>Operation</th>
<th>Neuro- logical recovery</th>
<th>Functional status at final follow-up</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>F</td>
<td>56</td>
<td>18</td>
<td>12</td>
<td>C3-C4</td>
<td>C3-C4 Partial</td>
<td>Spastic tetraplegia</td>
<td>Laminectomy of C3-C5 Facet fusion</td>
<td>Fair</td>
<td>Able to sit up Unable to stand and walk at 4 years</td>
</tr>
<tr>
<td>2</td>
<td>F</td>
<td>58</td>
<td>35</td>
<td>2</td>
<td>C4-C5</td>
<td>C4-C5 Partial</td>
<td>Spastic tetraplegia Urinary incontinence</td>
<td>Laminectomy of C3-C6 Splitting of dura Facet fusion</td>
<td>Good</td>
<td>Able to walk short distances at 1 year 6 months Dead at 2 years</td>
</tr>
<tr>
<td>3</td>
<td>F</td>
<td>57</td>
<td>24</td>
<td>5</td>
<td>C4-C5</td>
<td>C4-C5 Partial</td>
<td>Spastic tetraplegia Urinary incontinence</td>
<td>Laminectomy of C3-C6 Splitting of dura Facet fusion</td>
<td>Good</td>
<td>Able to walk short distances at 1 year 3 months</td>
</tr>
<tr>
<td>4</td>
<td>F</td>
<td>67</td>
<td>21</td>
<td>12</td>
<td>C4-C5</td>
<td>C3/C4 Complete</td>
<td>Spastic tetraplegia Urinary incontinence</td>
<td>Laminectomy of C3-C6</td>
<td>Poor</td>
<td>Completely bedridden at 1 year Urinary incontinence</td>
</tr>
<tr>
<td>5</td>
<td>M</td>
<td>60</td>
<td>10</td>
<td>3</td>
<td>C6/C7</td>
<td>C6/C7 Partial</td>
<td>Spastic tetraplegia</td>
<td>Laminectomy of C3-C7 Splitting of dura</td>
<td>Good</td>
<td>Able to stand Unable to walk Wheelchair-bound at 1 year</td>
</tr>
</tbody>
</table>

Case 1. Figure 1—Pre-operative radiograph. Note the bony ankylosis from C1 to C3, and the erosive changes and subluxation between C3 and C4. Figure 2—Myelogram showing a partial block at the interspace between C3 and C4. Note that the site of the block coincides with that of the erosive change in the facet joint. Figure 3—Radiograph taken one year after operation, showing a solid bony fusion between C3 and C4. Figure 4—Histological picture of the newly formed synovial tissue excised from the interspace between the adjoining spinous processes. Note fibrin deposition on the surface and diffuse infiltration of the inflammatory cells in the subsynovial layer. (Haematoxylin and eosin, × 60.)
Case 2. Figure 5 – Pre-operative radiograph. Note the bony ankylosis from C2 to C4, and from C5 to C7. Figure 6 – Radiograph taken in flexion, showing the 4 mm anterior slip of C4 on C5. Figure 7 – Myelogram showing a partial block at the interspace between C4 and C5. Note that the site of the block coincides with that of the erosive changes in the facet joint. Figure 8 – Radiograph taken one year after operation, showing solid bony fusion. Figure 9 – Histological picture of granulation tissue at the extradural space. The invading vascular granulation tissue is seen on the right, and the residue of degenerate ligament on the left. (Haematoxylin and eosin, × 60.)

Case 3. Figure 10 – Pre-operative radiograph taken in flexion showing severe erosive change in the facet joint between C4 and C5. There is bony ankylosis between C1 and C2, and between C3 and C4. Figure 11 – Myelogram showing a partial block at the interspace between C4 and C5. Figure 12 – Radiograph taken one year after operation, showing solid bony fusion.
erosive changes in the facet joint, except in one patient (Case 4) in whom the block occurred one vertebra higher than the subluxation.

TREATMENT AND RESULTS

Pre-operative regimen. One patient (Case 1) was placed on skull traction using Crutchfield tongs for two weeks and another two (Cases 2 and 3) were treated by head-halter traction for two weeks each; none showed neurological improvement.

Operation. All five patients underwent a decompressive laminectomy. On removal of the lamina a transverse band-like mass of soft tissue which had been encircling the posterior half of the dura was found at the level where the myelographic block had been seen. Excision of this mass of tissue, which was adherent to the dura, revealed an annular indentation of the underlying portion of the dura, which could be termed a constriction ring; in no case was pulsation of the dura observed. In three patients a longitudinal mid-dorsal incision was made in the dura with its centre at the constricting ring, and in all three patients pulsation became evident soon after the dura was opened. The dura itself was slightly thickened and was adherent to the arachnoid. A fascial patch graft was applied to the dural opening. In two patients (Cases 1 and 4) the constricting ring was not cut through because in one (Case 1) we did not, at that time, appreciate the importance of this procedure, and in the other (Case 4) excessive blood loss and subsequent shock prevented us from proceeding.

In three patients the facet joints were fused; in the remaining two (Cases 4 and 5) this was not done because of the severe intra-operative bleeding and the poor general condition of the patients.

Histopathological examination (see figs 4 and 9). Histologically the band-like mass of soft tissue, which was present in the epidural space and compressing the cord, consisted mainly of degenerate ligamentous tissue interspersed with many foci of infiltrating vascular granulation tissue. In every specimen taken from all five patients the same basic histological pattern was observed; however, the extent of invasion of ligamentous tissue by the granulation tissue and the degree of infiltration by inflammatory cells varied considerably from patient to patient. The only specimen of the dura (obtained from Case 2) showed unremarkable changes, except for the irregular pattern of the fibrous structure.

Postoperative results (see Figs 3, 8 and 12). The length of follow-up varied from one to four years. With the first patient (Case 1), whose constricting ring was not cut, postoperative neurological recovery was very slow; at six months her neurological condition was still unchanged, and only at one year was slight recovery first noted. At two years, examination showed no sensory deficit but there was a feeling of tingling in the distal portions of the four extremities. The tendon reflexes in the upper limbs were still increased and there was still ankle clonus. Bladder and bowel control were normal. Some recovery of motor power and function was also present; she was able to sit up and to feed herself, but was still unable to walk.

In the three patients (Cases 2, 3 and 5) who had the release procedure, very rapid recovery from the paralysis was observed. At one week every patient reported some improvement of motor power and sensibility in the four extremities. At six months examination revealed virtually no sensory deficit in two; the third (Case 3) had an area of hypo-algesia but had normal tactile sensibility below the tenth thoracic level on the left. Recovery of motor power also was evident; all recovered to at least Grade 3 (on the MRC scale) in the four extremities. Although the tendon reflexes were still increased slightly in the four limbs, in none was ankle clonus present. In all, however, the plantar responses were still extensor. The functional status differed considerably; two patients were able to walk short distances but a wheelchair was still needed for long distances; the remaining patient (Case 5), who had been wheelchair-bound for three years before operation, was able to stand but still unable to walk. Urinary incontinence, which two of these three patients had suffered, disappeared after operation.

The remaining patient (Case 4), in whom the operation was complicated by severe intra-operative bleeding, was neurologically worse after the operation; complete anaesthesia and analgesia developed below both elbows and below both nipple lines, with nearly complete motor paralysis of the four extremities and the trunk.

DISCUSSION

A review of the literature shows that in patients with rheumatoid arthritis two different pathological processes may lead to subaxial cervical cord compression: one is severe narrowing of the spinal canal caused by subluxation, the other is intraspinal encroachment by rheumatoid granulation tissue. The former mechanism is now well recognised and well documented (Hauge 1958; Storey 1958; de Blecourt and Veenstra 1960; Whaley and Dick 1968; Lidgren, Ljunggren and Ratcheson 1974; Hughes 1977).

The latter mechanism, on the other hand, seems to be less well recognised. In 1961 Hauge first reported a patient in whom cord compression had occurred at the thoracic level as a result of the intradural encroachment of granulomatous tissue. In 1967 Hopkins reported on two patients in whom the main cause of the cord damage was infiltration of "rheumatoid-like" granulation tissue into the dura; he also described a constricting ring in the dura, and referred to the necessity for releasing this constriction. Since then several authors, including Polyzois and Pearson (1973), Kataoka et al. (1978) and Price (1978), have reported on patients with similar pathology.

In each of our five patients the cause of the cord
compression was a band-like mass of soft tissue in the epidural space, which microscopically consisted of degenerate ligamentous tissue interspersed with foci of infiltrating granulation tissue. The ligament was in fact the ligamentum flavum, and the histopathology indicates that it was being invaded by a pannus of inflammatory granulation tissue. Of interest is the origin or source of this granulation tissue. We found abundant growth of abnormal tissue at the interspaces between the adjoining laminae as well as between the spinous processes; microscopically this was inflammatory synovial tissue. It may be argued that the occurrence of synovial tissue at these sites is rather unusual; however, Bywaters (1982) reported that synovial tissue of bursal origin can occur here. Another possible source of the pannus is the facet joints, where, at operation, marked proliferation of synovial tissue was seen and where, on the radiographs, severe destructive changes were observed.

At operation we found that the abnormal mass in the epidural space had nearly always adhered to the outer surface of the underlying dura. So it seems most likely that the constricting ring in the dura was produced by a combination of external pressure and extension into the dura of the contiguous inflammation. However, in the only specimen which was taken, no significant inflammatory changes were seen histologically in the dura itself. This is in contrast with the reports by Hopkins (1967), Polyzoisdes and Pearson (1973) and Kataoka et al. (1978) who found infiltration of granulation tissue into the dura.

For myelopathy due to intraspinal encroachment of granulation tissue such as was seen in our patients, no method other than a decompressive laminectomy would be effective. It should also be emphasised that laminectomy alone might not achieve sufficient decompression, because the cord is often tightly compressed by the constriction ring in the dura. Whenever this is found and there is no pulsation in the dura it is mandatory to release this constriction. The striking effect of this procedure can be verified by the immediate return of pulsation, and also by very rapid neurological recovery as seen in our three patients (Cases 2, 3 and 5).

The results of operation in the five patients, as far as neurological recovery is concerned, were satisfactory except for one patient: good recovery in three patients (Cases 2, 3 and 5), fair in one (Case 1), and poor in one (Case 4). However, assessed from the standpoint of overall physical function, the results were rather disappointing. This is largely due to the fact that preoperatively all patients had marked physical handicaps as a result of severe, long-standing rheumatoid disease and these remained unchanged or increased postoperatively.

REFERENCES

Bywaters EGL. Rheumatoid and other diseases of the cervical interspinous bursae, and changes in the spinous processes. Ann Rheum Dis 1982; 41: 360–70.


