Wasting of the small hand muscles in upper and mid-cervical cord lesions

J.A. MATHEWS

From The Department of Rheumatology, St Thomas’ Hospital, London, UK

Received 2 March 1998 and in revised form 3 July 1998

Summary

Four patients are described with destructive rheumatoid arthritis of the cervical spine and neurogenic wasting of forearm and hand muscles. The pathological connection is not immediately obvious, but a relationship between these two observations is described here with clinical, radiological, electrophysiological and necropsy findings. Compression of the anterior spinal artery at upper and mid-cervical levels is demonstrated to be the likely cause of changes lower in the spinal cord. These are shown to be due to the resulting ischaemia of the anterior part of the lower cervical spinal cord, with degeneration of the neurones innervating the forearm and hand muscles. These findings favour external compression of the anterior spinal artery leading to ischaemia in a watershed area as the likeliest explanation for this otherwise inappropriate and bizarre phenomenon.

Introduction

Wasting of the muscles innervated by the C6–T1 nerve roots associated with upper cervical compression was first described by Oppenheim in 1913. Wasting of the forearm muscles was also noted with tumours of the spinal cord extending into the posterior fossa by Elsberg and Strauss (1929) and Garcin et al. (1933). Symonds and Meadows (1937) described similar findings with lesions in the neighbourhood of the foramen magnum. They provided no explanation for the phenomenon but speculated that ‘it may be due to interruption of a descending arterial supply’. The study reported here is an attempt to explain the mechanism of the association particularly with lesions in the mid-cervical spine.

Rheumatoid disease affects synovial tissue throughout the body and leads to instability of joints. This is of special importance in the cervical spine due to the close relationship with the spinal cord. Davis and Markley, for example, reported a patient in 1951 with severe rheumatoid disease in whom destructive changes in the upper cervical spine had led to upward and backward displacement of the odontoid process and features of medullary compression. Storey reported a rheumatoid patient in 1958 who died suddenly and was found to have vertical subluxation, the foramen magnum having descended threading itself over the odontoid process.

Radiological evidence of lesser cervical joint instability is shown to be relatively common and results from ligamentous damage. In a study of 44 patients, Sharp, Purser and Lawrence showed upper cervical disc narrowing, erosion of vertebral end plates and destructive changes in apophyseal joints. In a survey of 333 cases, Conlon, Isdale and Rose showed antero-posterior atlanto-axial in 25% and serial sub-axial subluxations in 23%. Similar findings were reported by Mathews and Meikle and Wilkinson. In general, the incidence of neurological sequelae is low.

Vertical subluxation, i.e. descent of the skull base over the odontoid process of axis however requires more than ligamentous laxity. Actual joint and bone destruction is necessary, and it therefore represents more severe disease. Minor amounts were described in the studies above, but the particular danger of this lesion with respect to the spinal cord was first emphasized by Ball and Sharp in 1971. Webb, Hickman and Brew described a fatal case in which...
death resulted from massive thrombosis of the distorted vertebral arteries at the level of combined antero-posterior and vertical subluxation. Three further patients were described by Swinson et al. in 1972, emphasizing the frequent discrepancy between radiological changes and neurological features. Sub-axial subluxations were reported by Creglin, MacCabe and Hamilton showing C4–5 to be the commonest level for severe changes. The surgical management in severe cases is described by Meijers et al.

The prevalence of neurological features of cervical subluxations has been assessed by Stevens et al., who found evidence of cervical myelopathy in 24/36 patients with atlanto-axial subluxation. Marks and Sharp described such features in 31 patients, and Hopkins described four patients with tetraplegia which appeared to be due to compression of the spinal cord, although curiously not necessarily related to subluxation. Suspicion was directed to the vascular supply of the spinal cord by the report of a case with both atlanto-axial and C4–5 subluxation. The detailed pathology of C4–5 subluxation was examined by Hughes who showed anterior infarction of the spinal cord resulting from compression of the anterior spinal artery. The cases reported here are a clinical, radiological, pathological and electromyographic (EMG) extension of this work.

Case reports

Patient 1

This female patient, aged 57 at death, was referred to hospital 7 years previously complaining of pains in the shoulders, toes and sides of the feet. She developed sero-positive, nodular, and mutilating rheumatoid disease with vasculitis and was treated with corticosteroids and azathioprine. After 3 years, her neck became slightly painful, and 3 months later still the neck was noticed to ‘clunk’ on flexion. X-rays showed anterior subluxation of C4 on C5. A collar was supplied. Despite this, she noticed weakness in all four limbs during the next year and by the following year hyper-reflexia, clonus and bilateral extensor plantar responses were recorded. She was admitted as an emergency due to falling, reporting paraesthesiae in the arms and painful spasms in the legs. Severe rheumatoid deformities were noted in all limbs. Power was reduced in both arms and legs with gross spasticity, increased tendon reflexes and bilateral extensor plantar responses. There was slightly reduced sensation to all modalities up to the level of C4. EMG showed spontaneous activity with evidence of denervation in left deltoid, biceps, extensor digitorum communis and right deltoid, extensor digitorum communis and 1st dorsal interosseous. However, motor conduction in the relevant peripheral nerves was normal. X-rays of the cervical spine (Figure 1) showed gross forward subluxation of C4 on C5 to about half the total antero-posterior depth of the vertebral body, with some diminution and irregularity of the inter-vertebral disc below. A diagnosis of cervical cord compression was made and reduction of the subluxation by skull traction was attempted. Tongs were inserted under local anaesthetic and 3 lb (1.5 kgms) traction was applied. The patient died of cardio-respiratory failure and a chest infection.

At autopsy, the cervical spine was fixed with the cord in situ. A post-mortem vertebral arteriogram showed these vessels to be patent. The spinal canal was opened by a laminectomy and the cord removed. The spine was sawn in the sagittal plane. Necrotic changes were found in the disc between C4 and C5 (Figure 2) and reduction in height of the disc between C5 and C6. The anterior subluxation of C4 on C5 was demonstrated, together with compression of the anterior aspect of the spinal cord against the upper posterior aspect of C5. The anterior aspect (Figure 3) of the cord at this level was seen to be blanched. A section of the cord (Figure 4) showed triangular distortion by this pressure, and compression of the anterior spinal artery, which was empty of blood.

Figure 1. Patient 1. A lateral X-ray of the cervical spine shows gross forward subluxation of C4 on C5.
Table 1  Clinical, radiological, EMG and post-mortem findings

<table>
<thead>
<tr>
<th>Patient</th>
<th>Age at death/Sex</th>
<th>Years of disease</th>
<th>Motor signs (arm weakness)</th>
<th>Sensory symptoms</th>
<th>Plantar responses</th>
<th>X-rays</th>
<th>EMG denervation</th>
<th>Treatment</th>
<th>Post-mortem findings</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>1</td>
<td>57/F</td>
<td>7</td>
<td>+</td>
<td>Paraesthesiae arms</td>
<td>↑↑</td>
<td>Severe vertical and ant.-post</td>
<td>C₄–C₆</td>
<td>Right C₄, C₇, C₈, T₁</td>
<td>Foramen magnum decompression, mid-cervical fusion, collar</td>
</tr>
<tr>
<td>2</td>
<td>59/F</td>
<td>13</td>
<td>+</td>
<td>Numbness hands</td>
<td>↑↑</td>
<td>Severe vertical</td>
<td>C₄–C₆</td>
<td>Right T₁</td>
<td>Collar</td>
</tr>
<tr>
<td>3</td>
<td>54/M</td>
<td>20</td>
<td>+</td>
<td>Numbness hand and finger tips</td>
<td>↑↑</td>
<td>Gross vertical</td>
<td>C₄–C₆</td>
<td>Left C₄, C₇</td>
<td>Collar, skull traction</td>
</tr>
<tr>
<td>4</td>
<td>54/F</td>
<td>4</td>
<td>+</td>
<td>Paraesthesiae hands</td>
<td>↑↑</td>
<td>Gross ant.-post with myelographic block C₁</td>
<td>No</td>
<td>Right C₆, C₇, T₁</td>
<td>Skull traction C₁–C₂ fusion</td>
</tr>
</tbody>
</table>

RA of the spine and wasting hand muscles.
Figure 2. Patient 1. Necrotic changes are shown in the intervertebral disc between C4 and C5 and diminution in the size of the disc between C5 and C6. The upper posterior border of C5 is shown protruding into the intervertebral canal.

Histopathology showed no occlusion of spinal vessels at other levels, thus in all other sections the anterior and posterior spinal arteries were patent. In the central parts of the spinal cord from C4 to C8, there was proliferation of capillaries, and the spinal veins were moderately enlarged and patent. The spinal cord showed a large irregular area of partial infarction at C5, C6 and C7. In addition, the bony specimen was decalcified, embedded, and sections cut to demonstrate the state of the intervertebral foramina, the nerve roots, and the apophyseal joints (Figure 5). These did not reveal any encroachment upon or fibrosis within the intervertebral foramina. The pathology of this case has been reported by Hughes.20

Patient 2
This female patient, aged 59 at death, had been referred to our unit 2 years previously with an 11-year history of seropositive, erosive, nodular rheumatoid arthritis which had been treated with corticosteroids and ACTH. She gave a history of several months of increasing pain in the cervical region, helped slightly by a collar. Later, however, the pain became very severe. She then began to have numbness and weakness of the hands and subsequently the legs followed by flexor spasms of the left arm and both legs. Neurological
examination demonstrated that the arms were weak, with severe wasting of the intrinsic muscle of the hands, and the legs were spastic. Tendon reflexes were all brisk with absent abdominal reflexes and bilateral extensor plantar responses. Sensation to pin prick was diminished bilaterally from C5–T5.

X-rays showed vertical subluxation of a generous size foramen magnum over the odontoid process and unstable forward subluxation of C4 on C5 and to a lesser extent of C5 on C6. Myelography (Figure 6) showed a partial block at the C4–5 level. EMG revealed fibrillation in the small muscles of median and ulnar innervation in both hands, with no evidence of impaired peripheral motor conduction. As there was some improvement in a restrictive collar, surgery was not offered, but after a further 2 years she died.

At post-mortem, the odontoid process could be detected protruding 1.5 cm above the foramen magnum. The medulla showed some atrophy of the pyramids and leptomeningeal thickening anteriorly. In the cervical region severe compression of the spinal cord with flattening was demonstrated at the level of C4–5, with blanching similar to that seen

**Figure 4.** Patient 1. The anterior spinal artery is compressed and the spinal cord at the level of C5 is distorted into a triangular shape. The arterial wall appears normal.

**Figure 5.** Patient 1. Sections through the apophyseal (a) joint did not reveal any foraminal encroachment or perineural (b) fibrotic changes.
Figure 6. Patient 2. Myelography in antero-posterior and lateral views shows partial block to the flow of myodil at the level of the C4–5 subluxation. The canal is deep at the level of the atlas and axis.

Figure 7. Patient 3. A lateral X-ray of the upper cervical spine shows the descent of the skull and atlas over the axis permitted by the resorption of the lateral masses of the atlas.

in patient 1. Section of the cord showed bilateral symmetrical long tract degeneration principally in the lateral corticospinal tracts. Stains showed depletion of neuronal cell bodies in the anterior horns of C4, C5, C6, C7 and C8. No other lesions lower in the spinal cord were detected.

Patient 3

This male patient, aged 54 at death, had been referred 12 years previously and already had an 8-year history of rheumatoid arthritis. This was seronegative for rheumatoid factor, and anodular, but erosive. He had insulin-controlled diabetes mellitus, and received systemic corticosteroids for control of the arthritis. For several years his neck had felt stiff and later he began to complain of numbness in the right forearm and hand, and the left fingertips. Subsequently he complained of ‘jumping’ of his legs, especially in bed, and paraesthesiae on coughing from the hips to feet. This was slightly improved by wearing a collar. There was also slight difficulty in initiating micturition. In the cranial nerves there was impaired sensation of the left side of the face and weakness of the pharynx. Power was reduced in both arms proximally, associated with shoulder stiffness, but there was distal weakness as well. No
increase in tone was detected but there was marked hyper-reflexia in the arms. The legs showed mild weakness with increased tone, and there was ankle clonus and hyper-reflexia with bilateral extensor plantar responses. Sensation was reduced in all modalities up to the C5 level. X-rays showed a downward subluxation of C1 on C2 and protrusion of the odontoid process into the foramen magnum due to resorption of the lateral masses of the atlas (Figure 7). There was also fixed forward subluxation of C4 on C5, at which level myelography (Figure 8) showed a partial obstruction. EMG revealed spontaneous motor unit activity indicative of denervation and high-frequency discharges in muscles of the right arm of C6, C7, C8 and T1 root origins. He underwent decompression of the foramen magnum and fusion of C4 to C5. He made a modest improvement, but lived a chair-bound and eventually bedridden existence, and subsequently had a right-sided stroke. He died 6 years later of a haematemesis from a benign gastric ulcer. The clinical aspects of this case were included in a report by Swinson et al.\textsuperscript{13}

At post-mortem the destructive changes of the lateral masses of the axis with vertical descent of the skull and atlas were demonstrated, as well as the nearly complete destruction and disappearance of the C4–5 disc with forward subluxation of C4 on C5. This left a prominent knuckle of bone protruding posteriorly into the spinal canal. The partial occlusion of the spinal canal at this level is shown (Figure 9), and there was severe compression of the cervical cord and the anterior spinal artery. The disc between C6 and C7 was degenerate, and a transverse bar affected the C7–C8 spinal cord segment.

**Patient 4**

This female patient, aged 54 at death, was referred to hospital with a 3-year history of joint pains, later documented as seropositive, nodular, erosive, rheumatoid arthritis. Symptoms included neck pain and stiffness, both of which had been relieved by manipulation. She also complained of headaches, dizziness and subsequently loss of feeling in the hands. She began to fall about and needed a frame to walk. On admission the severe rheumatoid arthritic changes were noted; the neck had a good range of movement but was painful and ‘crunching’. There was a moderate spastic paresis of all four limbs worse on the right, with generalized hyper-reflexia and bilateral
extensor plantar responses. There was a sensory level to pin prick up to C2, and reduced position and vibration sense in the arms. X-rays showed antero-posterior atlanto-axial subluxation with erosion of the odontoid process, and disc space narrowing at C3–4 and C4–5, but no subluxation. She was fitted with a stout collar, mobilized by a physiotherapist, fitted with boots and instructed to avoid further manipulative treatment. There was considerable improvement. One year later she was readmitted because of leg weakness and falling. On one occasion she struck her head and subsequently noticed more marked weakness. The neurological signs were similar to, but more severe than, those noted previously. Electromyography (EMG) of the right arm revealed fibrillation and positive waves in deltoid, triceps, extensor digitorum communis and the small muscles of the hand. No abnormality of peripheral conduction was found. Plain X-rays of the cervical spine (Figure 10) showed forward displacement of the atlas on the axis on flexion, indicating instability of the atlanto-axial joint, and myelography showed a complete block of the extra-dural type at the lower border of C1. The remainder of the cervical spine was unremarkable.

She was transferred to a neuro-surgical unit where skull traction was applied. Two weeks later a posterior fusion was performed between C1 and C2, but two days post-operatively the quadriplegia became total, and she died of bronchopneumonia. Post-mortem findings confirmed the subluxation of C1 on C2 with severe upper anterior cervical cord pressure. The findings in the mid-cervical spine are not available, but there was no subluxation at this or any other level apart from C1–C2.

**Discussion**

The cause, mechanisms and effects by which an upper or mid-cervical problem can lead to wasting of small hand muscles are illustrated by these four cases. The severe destructive rheumatoid synovitis has destroyed ligaments and damaged joints leading to instability. This instability in turn has led to cervical subluxations, causing not only pressure on the spinal cord but compression of its arterial supply. Examples of this are the impression of the odontoid process into the medulla in all four patients, and compression of the cervical cord and anterior spinal artery by the upper posterior border of the 5th cervical vertebra in patients 1, 2 and 3. In patient 4, the only lesion was at C1–C2 and the compression was not directly visualized. This case is included speculatively, as the separation from the root lesions was even greater.

An explanation of the dissociation of level of the subluxation from the level of the root lesions is needed. Symonds and Meadows postulated that it might be due to a descending arterial supply to the motor cells concerned. Subsequent studies demonstrated the vulnerability of the blood supply to the spinal cord. Regular segmental arteries do not exist in the cervical spine and cord, thus a longitudinal blood flow is needed. These studies suggest that the flow is caudal at cervical levels, the anterior spinal arteries being supplied by the two vertebral arteries with variable feeding vessels at C4 or C5, and C1 or C2, exposing between the vulnerable ‘watershed area’.

The results of ischaemia or ‘spinal stroke’ are also explained by the fact that at least the anterior two-thirds of the cord receives its blood supply from the anterior spinal artery. Bolton showed that virtually the whole cord, with the exception of the posterior portion of the posterior columns and the posterior horn, could be deprived by anterior spinal artery occlusion, and this would be expected to lead to the neurological features of these four patients—sensory symptoms in the arms, lower motor neurone lesions in the low cervical cord and spasticity of the legs. An alternative explanation was proposed by
Taylor and Byrnes. Their experiments with hydroscopic balloons inserted into the subarachnoid space of baboons at C2–3 led to lower cervical cord changes thought to be venous infarction and hypoxia. Stark, Kennard and Swash supported this view, but no convincing evidence has been published. Another possible cause of the neurogenic wasting of the hand muscles is pressure in intervertebral foramina or root sleeve fibrosis such as is associated with lesions of the apophyseal joints. To explore this possibility, we examined sections of the intervertebral foramina in patient 1 at the levels of the emerging motor nerve roots responsible for the territory in which denervation was found; i.e. C5, C6, C7, C8, T1 (Figure 5). No evidence of displacement or distortion of the apophyseal joints, fibrosis of the nerve root sleeves nor vascular occlusion was found to explain the weakness and wasting. In patient 1, the lower cervical spine was examined histologically. In patients 2 and 3, there was no radiological evidence of any significant lower cervical spine lesion, and in patient 4 the minor spondylitis at C6–7 would not account for the denervation findings. These studies were carried out before the availability of axial tomography which could have helped further delineate foraminal pathology in vivo.

Wasting of muscles that move inflamed joints is common in most joint disease. Its mechanism is unclear, but is not associated with features of EMG denervation which were found in all these cases. Neurogenic muscle wasting is characterized by fibrillation or positive sharp waves. Fibrillation consists characteristically of small potentials with a duration of 0.5–2 ms and amplitude of 30–150 μV, with a regular but often rallantando repetition rate of 2–10 a second. Positive sharp waves are sometimes of slightly longer duration but in other ways are comparable. Neither is found in the absence of denervation, and they document an important aspect of the muscle wasting and weakness in the forearms or hands of all four of these patients. The mechanism for production of these potentials has been thought to be increased sensitivity of denervated muscle fibre to small amounts of circulating acetylcholine. This phenomenon is not detected in muscle wasting due to joint disease, although it can be found in acute polymyositis, which these patients did not have.

In conclusion, in all four of these patients there was external compression of the anterior spinal artery leading to local and distal ischaemia of the cervical cord. There was severe atlanto-axial subluxation and in all cases, additional C4–5 subluxation, in patients 1, 2, and 3 all potentially leading to this compression. Anterior spinal artery insufficiency has been shown to lead to ischaemia and gliosis in the mid-low cervical cord levels with consequent lower motor neurone weakness, confirmed by EMG examination, as well as reflex and sensory changes. It has not escaped our notice that these clinico-pathological observations might provide an explanation for the suspended neurological features found in cervical cord lesions.

Acknowledgements
I am grateful to Dr R.W. Ross Russell and Professor Lindsay Symon for their help and advice in treating these patients and to Dr J. Trevor Hughes for the pathology examinations. I am also indebted to Dr R.W. Ross Russell also for help in preparing the manuscript.

References
2. Elsberg CA, Strauss I. Tumours of the spinal cord which project into the posterior cranial fossa. Arch Neurol Psychiatry 1929; 21:261–73.
15. Meijers KAE, van Buesekorn KT, Luyendijk W, Duijfjes F.


