The spinal cord in rheumatoid arthritis with clinical myelopathy: a computed myelographic study

JOHN M STEVENS,*† BRIAN E KENDALL,* H ALAN CROCKARD*‡

From the National Hospitals for Nervous Diseases,* St Mary's Hospital,† and the Middlesex Hospital,‡ London, UK

SUMMARY Thirty one patients with suspected myelopathy due to rheumatoid arthritis were examined by plain radiography and 27 had computed myelography. Clinical features and radiological findings were compared. Deformity of the spinal cord could occur in the absence of combined anterior and posterior compression and correlated closely with clinical features only when considered in combination with skeletal and adjacent soft tissue abnormalities. The best surgical results were achieved by transoral odontoidectomy.

The myelopathy associated with long-standing rheumatoid arthritis is acknowledged to be due to spinal cord compression in the great majority of cases. The mechanisms by which the cord may be compressed have been described extensively.¹⁻⁴ However, it is recognised that the degree of atlanto-axial subluxation shown on plain radiographs is not related closely to the presence of myelopathy.⁴⁻⁶ Even when the minimum sagittal diameter of the spinal canal has been taken into account there remain a considerable number of cases where compression cannot be inferred from plain radiographs alone.^{7,8} Indeed, Stevens *et al* found seven of 24 patients with myelopathy in whom the minimum sagittal diameter of the canal exceeded 16 millimetres.⁸ Redlund-Johnell and Peterson have recently shown that vertical atlanto-axial subluxation has the highest incidence of myelopathy,⁹ but this accounts for only a minority of cases. Alternative mechanisms have been suggested, based in the main on observations from isolated case reports: non-bony granulomatous masses,^{12,10,11} vertebral artery compression or thrombosis,^{12,13} and ischaemic myelomalacia from small vessel disease.^{4,8}

Adequate clinical investigation can be difficult in these patients. Myelography has been performed infrequently in most published series, the consensus opinion being that it should be reserved only for those cases where subluxation alone was inadequate to explain the myelopathy, and to localise the site of

cord compression where this was in doubt.^{2,4,8,14,15,16} Computed tomography (CT) has been advocated recently because it has detected more erosions, accurately defined the pathological anatomy in severely deformed patients, and shown soft tissue as well as bony abnormalities.^{1,17-19} However, a better definition of soft tissue and neural elements is provided if CT is performed after the introduction of intrathecal water-soluble contrast medium, that is computed myelography and cisternography.^{20,21}

At the National Hospital for Nervous Diseases (Maida Vale), we have been performing regularly both conventional and computed myelography on such patients and have been impressed by the greater understanding these studies have provided of the mechanical factors affecting both cord structure and function. In addition to localising accurately the site of maximum cord involvement, the importance of anterior deformation of the spinal canal regardless of the width of the subarachnoid space dorsal to the cord has been revealed. This data has enabled us to follow a surgical approach for these patients which has thus far yielded better results than other commonly reported surgical methods.

The purpose of this study has been to examine the correlations between clinical features, abnormalities on plain radiographs, and the pathological anatomy revealed by computed myelography, and to use these to establish a more acceptable concept of cord compression than can be gleaned from existing literature.

Materials and methods

Our material consisted of 31 patients with rheumatoid

Address for reprint requests: Dr JM Stevens, The National Hospital for Nervous Diseases, Maida Vale, London W9 1TL, UK.

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Fig 1 Classification of deformities of external form of the spinal cord at the cranio-vertebral junction, also applicable to subaxial levels. Measurements are compared to classification by visual assessment.

arthritis referred to the National Hospitals for Nervous Diseases (Maida Vale) and St Mary's Hospital (Paddington) with suspected compressive cervical myelopathy. There were 19 females and 12 males, and the age range was 22 to 77 years. All were investigated by plain radiographs, 29 had water-soluble myelograms performed by lumbar puncture, of which 27 were followed by CT on a GE 8800 or Somatom II scanner. Slice thicknesses of 1.5 mm–2 mm were performed sequentially across the foramen magnum and 5 mm thick slices were made with 2 mm overlap between slices to examine the cervical spine from C.2 to T.1. Sagittal and coronal reformatted images were prepared from this data. All patients were scanned supine with the head supported in a neutral position, and in six, the foramen magnum region was scanned in both flexion and extension. Measurements were taken of the mid-sagittal diameters of cord, bony canal, theca and width of the subarachnoid space anterior and posterior to the cord. In addition to measurements, a visual classification of the variations in cross-sectional shape of the cord was devised: loss of normal anterior convexity was designated *mild*, anterior concavity *moderate* and obvious antero-posterior flattening *severe* deformity (figs 1, 2, 3 and 4).

Measurements were made from plain radiographs according to methods described by Inger Redlund-Johnell, including minimum mid-sagittal diameter of the bony canal.¹

Clinical features were arranged to conform in general with those discussed by other authors, and they will be defined in the *Results* section. Each was compared with the radiological variables defined above. Operation was performed on 24 patients. Six had posterior occipito-vertebral or atlanto-axial fusions, eight had subaxial fusions and 15 had transoral odontoidectomy combined with posterior fusion. The technical aspects of the latter are described elsewhere.²² Some patients having subaxial fusions had cranio-vertebral operations as well. Follow up periods varied from three months to four years, and not all operations were performed by the same group of surgeons.

Fig 2(a) Axial CT with sagittal reformat. Note atlanto-axial subluxation, and mild congruous cord deformity on the axial image which is only just perceptible on the sagittal image. (b) Coronal reformat, showing lateral atlanto-axial subluxation which the axial image in (a) shows to be associated with more distortion of the right side of the cord. This patient had left arm paraesthesia.

Statistical significance was calculated using Fisher's Exact Probability Test.

Results

The results are presented as follows: (1) observations on the spinal cord from computed myelography, (2) skeletal and soft tissue factors (3) correlations with clinical features, and (4) results of the various types of operative procedure.

1 The spinal cord

On CT the cross-sectional shape of the cord was either normal or deformed. Two kinds of deformity occurred: those "mirroring" deformations of the theca, which were called congruous or "com-

Fig 3 Axial CT. Moderate congruous cord deformity.

Fig 4 Axial CT with sagittal reformat. Severe congruous cord deformity. Note the small ossific nodule lying in the atlanto-dental interval, and this subluxation was immobile on plain radiographs.

pressive", and those unassociated with thecal deformation at the same level, which were regarded as incongruous or "atrophic".

Incongruous deformity of the spinal cord In nine patients the cord looked atrophic throughout most of

the spinal canal. Seven had minimal or no compression at the craniovertebral junction and eight had moderate or severe compression usually at multiple subaxial cervical levels. Therefore atrophy is related to subaxial cord compression and not to compression at the craniovertebral junction, in this series ($p = 0.008$).

Congruous deformity of the spinal cord This was seen at the cranio-vertebral junction in 21, at subaxial levels as well in five and alone in five. Sagittal measurements of the cord tended to overlap between the classification based on visual assessment as shown in fig 1, and it was felt that for clinical correlations cord deformity was best assessed visually. The anterior surface of the cord was the only surface deformed at the cranio-vertebral junction in 17 (71%), and the cord was often shown not to be in contact with the deforming agent (mean separation 1.2 mm, range 0 to 4 mm). CT in flexion and extension (fig 5) showed an average percentage reduction in sagittal diameter of the theca of 31% (range 0–67%) on flexion, and of the cord of 15% (range 0–50%). The cord came into contact with the theca anteriorly when its sagittal diameter became narrowed, but in two cases there was up to 4 mm of subarachnoid space behind the cord. However, when the cord was deformed on flexion it never reverted to normal on extension in this group of six patients. It is concluded therefore that mechanical cord deformity becomes fixed, and can occur without the necessity for a posterior compressive element.

Normal cord shape Only two patients showed no cord abnormality at any level, and six showed none at the cranio-vertebral junction. In these the mean separation between the anterior surface of the cord and theca was 1.0 mm, indicating that the close approximation of cord and theca does not necessarily produce congruous deformity of the cord.

2 Skeletal and soft tissue abnormalities

Severity of congruous cord deformity and measurement of atlanto-dental separation and sagittal canal diameter are compared in fig 6. Trends are visible in this graphical representation but the actual figures as arranged do not reach statistical significance, presumably because the numbers are too small in each group.

Pannus was responsible for most of the distortion of the theca associated with congruous cord deformity in nine of the 21 relevant cases (43%) (fig 7). Rarer types of craniovertebral subluxation were also seen: anterior atlanto-occipital, four cases; and two had severe cord deformity; vertical atlanto-axial, three cases, with only mild or moderate deformity of the cord in each (fig 8); and posterior atlanto-axial, two cases.

Fig 5(a) and (b) Flexion and extension CT, the sagittal reformats are not quite in the same sagittal plane. The cord is compressed in flexion, not in extension. (c) Axial images of the same examination in flexion showing severe congruous cord deformity.

The width of the subarachnoid space anterior and posterior to the cord is indicated in fig 9. The posterior subarachnoid space was compressed from behind in seven cases, owing to anterior atlanto-occipital subluxation in two, pannus in two, and atlanto-axial rotation plus pannus in three. The cord was compressed against a normal posterior theca in a further

Fig 6 Graphical presentation of the relationship between severity of cord deformity at the cranio-vertebral junction and atlanto-dental separation (open diamonds) and minimum sagittal diameter of the spinal canal at this level (solid diamonds). (measurements are in mm).

Fig 7(a) and (b) Axial CT images from a case in which pannus was the dominant agent determining the pattern of congruous cord deformity. Note the importance of posterior pannus also.

Fig 8 Sagittal reformat (a) and axial images (b) showing mild vertical atlanto-axial subluxation and mild congruous deformity of the infraolivary segment of the medulla. This patient had downbeat nystagmus and swallowing difficulties.

three cases. Combined anterior and posterior compression was therefore shown in only ten of the 21 relevant cases (48%).

Lateral atlanto-axial subluxation was seen in 15 cases, and in nearly half (7) it was associated with lateralised cord deformity (fig 2).

Subaxial cord deformity occurred with a fixed angular kyphosis and vertebral collapse in three, single level anterior subluxation in four and multiple levels of subluxation ("staircase") in four.

Mobility of the atlanto-axial subluxation and severity of cord deformity are compared in fig 2. A significant relationship between increasing mobility

Fig 9 Graphical representation of the relationship between cord deformity at the cranio-vertebral junction and width of the subarachnoid space anterior (open diamonds) and posterior (closed diamonds) to the cord. (measurements are in mm).

and greater cord deformity is shown: eleven of 14 cases with at least moderate cord deformity has more mobile subluxation, compared to three of 13 with mild or no cord deformity ($p = 0.006$). Flexion produced little or no movement at the atlanto-occipital joint in 26 (84%), anterior atlanto-axial subluxation in 14 (45%) and posterior atlanto-axial subluxation in two (6%).

Several other skeletal and soft tissue features were considered: lengths of clivus, basal angle and anterior angulation of the neuraxis across the cranio-vertebral junction. No relationship with cord deformity was revealed.

3 Clinical features

The only clinical features which correlated with severity of cord deformity on CT were a combination of motor long tract signs and a sensory level, and they correlated more closely with mobility of the atlanto-axial subluxation than with its magnitude on plain radiographs. The duration of clinical myelopathy was shortest in patients with severe cord deformity at the cranio-vertebral junction, and longest in those with only subaxial involvement.

Motor long tract signs were counted present when there was spastic weakness worse in the legs, clonus, bilateral Babinski reflexes and gait disturbance. About half our patients (15) had these features, and about half of these (7) also had a sensory level somewhere between the sixth cervical and fifth thoracic segment. These signs, in combination, were taken as evidence of definite cord involvement.

Twelve patients complained of progressive weakness in the hands, legs or both but no signs could be elicited against a background of joint disease. Seventeen patients complained of paraesthesia usually in the arms, and six showed a positive Lhermitte's phenomenon. Bilateral glove-and-stocking anaesthesia

Table 1 Clinical features compared to findings on computed myelography in 27 cases

Clinical feature	No	Cord deformity at or near the cranio-vertebral junction			Subaxial cord deformity	Site of deformity of spinal theca	
		Severe or moderate	Mild	Nil		Mainly anterior	Antero-posterior
Cord—Motor	8	4	3	2 (subaxial only)	2	4	4
—Sensory-motor	7	7	0	0	2	0	7
—Nil	12	3	4	5	4	11	1
Subjective weakness	12	3	4	5	5	11	1
Paraesthesiae	17	7	5	5	5	10	7
(No paraesthesiae)	10	4	2	4	3	6	4
Lhermitte's sign	6	2	1	2	2	3	3
Glove-and-stocking anaesthesia	4	3	0	1 (subaxial only)	1	2	2

was found in four, hemianaesthesia in one, and patchy sensory loss in eight others.

Neck pain was a dominant feature in 19 patients, with occipital radiation in five. In eight, pain preceded the myelopathy by less than one year (five cases) or up to 17 years (three cases). Pain and myelopathy appeared together in seven, and myelopathy preceded pain by six to eleven months in three.

Features indicating hind-brain involvement were present in four patients, with swallowing difficulties in two and nystagmus in two (fig 8).

How these features were related to findings on computed myelography is shown in table 1. There was a significant correlation between definite cord involvement and severe or moderate cord deformity: seven of seven with involvement compared to three of 12 without ($p = 0.002$). A close relationship between these sensorimotor cord signs and combined anterior and posterior narrowing of the subarachnoid space was also revealed: seven of seven with signs compared to one of 12 without ($p = 0.002$). No patient with definite cord involvement had a normal cord on CT. The three patients with a normal cord on CT had subjective weakness, and one also had arm paraesthesia and a positive Lhermitte's phenomenon. Definite cord signs were lacking in three of the patients who had moderate or severe cord deformity (25%), but were present in all such cases when cord

compression was not just anterior alone. The presence of motor long tract signs, or any of the other clinical features considered in isolation, did not show a significant relationship with these CT findings.

Unilateral clinical features predominated in eleven cases. In over half (six) congruous deformity was concentrated on one side of the cord. Ipsilateral compression was seen with monobrachial paresis, paraesthesia and proprioceptive loss, and contralateral compression with hemiparesis, hemianaesthesia and in one, monobrachial paraesthesia (fig 2).

Further relationships with CT findings are shown in table 2. Short duration of clinical symptoms other than neck pain, was related to moderate or severe cord deformity at the cranio-vertebral junction (nine of eleven with short, compared to five of 16 with long or moderately long; $p = 0.013$). Conversely long duration of symptoms correlated with the presence of subaxial sites of compression only (0 of 11 with short compared to eight of 16 with long, $p = 0.012$).

Correlations with plain radiograph findings are shown in table 3. There was a significant relationship between atlanto-dental separation of more than 10 mm and definite cord signs (five of 14 with less than 10 mm, compared with six of eight with more than 10 mm, $p = 0.048$). Unpredictability therefore occurred most frequently with atlanto-dental separations of between 5 and 10 mm. Paradoxically neck

Table 2 Clinical features compared with findings on radiographs of the cervical spine in 31 cases

Radiological feature		No	Cord signs	Paraesthesia	Pain	Lhermitte's sign	Duration of clinical features (months)		
							1-6	6-12	12+
*Maximum ADI (mm)	≤4	14	5	9	12	2	3	5	6
	5-9	9	4	7	5	2	5	2	2
	≥10	8	6	1	2	2	5	2	1
†Mobility at C1-2	Marked	9	7	5	7	2	5	3	1
	Mild-moderate	5	4	5	4	2	3	2	0
	Fixed	17	3/4	7	8	2	5	4	8

*ADI is the atlanto-dental separation.

†"Mobility" refers to mobility at the atlanto-axial joint as defined in Figure 10.

Table 3 Duration of clinical features compared to findings on computed myelography in 27 cases

Duration of clinical features	No	Level	Cord deformity		Compression of the subarachnoid space	
			Severe or moderate	Mild or nil	Anterior alone	Posterior
Long (> 12 months)	9	C1	3	6	6	3
		Subaxial	5		2	3
Moderate (6–12 months)	7	C1	2	5	2	5
		Subaxial	3		2	1
Short (< 6 months)	11	C1	9	2	3	8
		Subaxial	0

Fig 10 Bar graph to show the relationship between cord deformity at the cranio-vertebral junction and mobility of the atlanto-axial subluxation assessed from flexion and extension radiographs of the cervical spine. Fixed-movement less than 2 mm; moderate, 2–9 mm; marked, more than 10 mm.

pain was more often a feature when atlanto-axial subluxation was relatively less (12 of 14 with less than 4 mm, compared to seven of 17 with more than 5 mm, $p = 0.014$). There were no correlations between these radiological features and paraesthesia, Lhermitte's phenomenon, duration of symptoms other than neck pain or sensory changes other than a sensory level.

Mobility of the atlanto-axial subluxation was significantly related to the presence of definite cord signs (11 of 14 with mild or marked mobility compared to four of 17 with no mobility, $p = 0.003$). No correlations between mobility and other clinical features were found.

4 Surgical results

Only ten patients had no operation at the time of writing. Three refused, two were considered too poor a surgical risk for medical reasons, in one myelopathy was doubtful and myelography normal, and four are still being considered for operation. Of these, four were not followed up at our hospitals, one died, and those awaiting operations have not deteriorated as far as we know at this time.

Sixteen patients had a transoral odontoidectomy.

In 14 it was combined with posterior cranio-vertebral fusion, and in one with an anterior fusion; one operation had to be abandoned because the vertebral artery was encountered in front of the dens (fig 11). All patients successfully operated upon by this approach improved, in both neck pain and myelopathic features. Two required subaxial fusion as well. Two have deteriorated within two years but the rest have had sustained improvement for up to four years. There was not a significant relationship between improvement and any of the radiological variables considered above.

Post-operative radiographs of the cervical spine showed that seven of the posterior fusions in this group failed within two years. Moreover in nine odontoidectomy and interlaminar wiring caused vertical atlanto-axial subluxation of between 5 and 10 mm. However, it was conspicuous that in all but two patients there was no recurrence of neck pain or any of the other clinical features despite this failure of immobilisation.

Four patients were treated by occipito-atlanto-axial interlaminar wiring alone. The dislocation was reduced in only two. One died of continuing cord compression within a year, one remained unchanged, one improved for about four months and then rapidly deteriorated, and one only showed a good sustained improvement.

Discussion

Distortion of the external form of the cord usually constitutes the only radiological evidence of myelopathy. Abnormalities within the substance of the cord can sometimes be shown by CT,^{23,24} ultrasound²⁵ and magnetic resonance imaging (MRI)²⁶; but the conditions required for consistent results are too restricted for them to contribute in the present context, with the probable exception of MRI which has yet to be directed specifically at the problems addressed here. Although conventional myelography can show alterations in transverse and sagittal diameter of the cord, the cross-sectional images provided by CT give a more sensitive indication of distortion of external form than mea-

Fig 11 Axial CT images from a patient with atlanto-axial rotation and severe cord deformity largely due to pannus. In section 16 it is possible to deduce that the vertebral artery passing between the lateral masses of C1 and C2 has been rotated into the midline, and now lies in the transoral operative field just below the anterior tubercle of the arch of C1. This arch is excised during operation; in this case the vertebral artery was injured and the operation had to be abandoned.

surements, for measurements contain unavoidable and considerable inaccuracies both on CT and conventional myelography.^{27,28} Plain CT usually does not show the cord well enough to detect the small changes which our results have suggested may be relevant. Therefore, at the time of writing, intrathecally enhanced CT is the most sensitive radiological test for the detection of myelopathy.

To date, this aspect of the radiology of the spinal cord has received little attention. Some consideration, however, had been given to it in cervical spondylotic myelopathy,²⁸ which has much in common with changes in the cord seen with rheumatoid arthritis, and recently in rheumatoid arthritis itself.²⁹ Data from necropsy material has indicated only a very general and inconsistent relationship between the severity of external cord deformity, clinical features and histo-

logical degeneration, and usually it is the surprising lack of close correlation between them that is emphasised.^{30,31} In spondylotic myelopathy for example, severe cord deformity may be associated with no degeneration in cord substance, and in rheumatoid arthritis Hughes has stated that often little degeneration is seen in even markedly deformed cords, and this has been so despite the presence of clinical myelopathy.³⁰ It has been suspected from necropsy material that cord deformity found in association with osteophytes was often nothing more than a post-mortem fixation artefact,³⁰ presumably because in these cases antero-posterior compression could not be inferred from the configuration of the spinal canal. Computed myelography has demonstrated emphatically, however, that these cord deformities exist during life: they are, as far as we can tell, fixed alterations

in external form and are not dependent upon the continuous application of deforming forces.

Several distinct but closely related processes are operating to produce external cord deformity, neurophysiological dysfunction, and internal histological degeneration.

Mechanism of cord deformity

We have taken care to distinguish two types of cord deformity: congruous or compressive and non-congruous or atrophic. Congruous deformities were by far the most common. Graphical representation of the relationship to increasing atlanto-dental separation and decreasing sagittal diameter of the spinal canal shows the expected tendencies, but the figures in the sample were not large enough to be significant. The disparity between severity of cord deformity and these skeletal features usually could not be explained by pannus as some have suggested,⁸ and vertical atlanto-axial subluxation was not associated with severe cord deformity as it has been implied.⁹

In our material mobility of the atlanto-axial subluxation was significantly related to the severity of congruous cord deformity. Flexion and extension CT studies in six cases showed a consistent reduction in canal size on flexion which was twice as large as the changes seen in the cord. It has been shown on computed myelography that the cord moves anteriorly at the foramen magnum on head flexion by between 1 and 2.5 mm, a movement which has been attributed to anterolateral traction from the upper digitations of the dentate ligament.²¹ The cord is therefore pulled against a potentially subluxing dens to a degree which varies somewhat from patient to patient. In this series anterior angulation of the neuraxis across the cranio-vertebral junction was not related in any way to anterior cord deformity. However, it may be that longitudinal tension in the cord is relevant, and this is known to be influenced by the posture of the entire spinal column,³² as is the relative position of the cord within the spinal canal. Combined anterior and posterior compression accounted for only about one third of cases in this material. Therefore it is suggested that the variable mechanical forces which tend to move and hold the cord anteriorly, and are brought to bear on head flexion, establish the conditions for deformation of the cord at the cranio-vertebral junction irrespective of the amount of free space present posteriorly.

Mechanisms of clinical and histological myelopathy

It is generally agreed that the presence and severity of myelopathy in rheumatoid arthritis is not predictable from plain radiographs.^{14 516} The difficulties are intensified because it is not easy to interpret clinical signs in patients with severe joint disease. Laasonen *et*

*al*²⁹ recently claimed to have demonstrated a relationship between cord deformity on computed myelography and the severity of clinical features as graded by Ranawat *et al*.¹⁵ They emphasised that no other radiological feature showed such a correlation in their material including the presence of pannus. This is an over simplification which can be misleading. In our material, and quite possibly in theirs as well, the only clinical features to correlate with cord deformity were those of definite cord involvement as defined in the last section; there was no correlation with features such as neck pain, paraesthesia, Lhermitte's sign, subjective weakness or any sensory loss other than a sensory level, no matter how they were grouped. Furthermore, severe narrowing of the sagittal diameter of the spinal canal (<15 mm) and severe atlanto-dental separation (>10 mm) do correlate with clinical cord involvement, as has been stated by others.^{8 9 15} Finally, pannus was a major factor in determining the pattern of cord deformity in nearly half our cases, and was certainly of significance in some of the illustrations of Laasonen *et al*,²⁹ although they dismissed this as a factor involved in producing myelopathy. It is significant that unilateral symptoms and signs were often associated with deformity of mainly one side of the spinal cord in our material, indicating that in these cases there was variable selective involvement of motor neurones, or crossed and uncrossed long tracts, and suggesting in turn that the pattern of cord deformity is of clinical importance.

The degeneration seen on histological preparations of the spinal cord in spondylotic myelopathy and rheumatoid arthritis has the appearance of ischaemic change or focal demyelination.^{30 31 33 34} It probably often represents a stage of irretrievable cord damage and tends to be most marked at sites of greatest cord deformity, but may be entirely absent even in the presence of quite severe external changes. Rana *et al*⁷ attributed the inconsistent relationship between skeletal changes and clinical features in rheumatoid patients to the variable response of the cord to trauma, and the concept of repetitive minor trauma has been propounded strongly in the case of spondylotic myelopathy also.³⁵ Abnormalities in somato-sensory evoked responses in patients with intermittent medullary compression due to Chiari malformation have led Larson *et al*³⁶ to suggest that some clinical features were evoked by disrupted synaptic transmission, mediated not by ischaemia, but by sheering strains within cord substance and intermittent geometric distortion of synaptic relationships. In our material, cord deformity was related to the presence of sensori-motor cord signs only when there was combined anterior and posterior

compression of the subarachnoid space, and other clinical features occurred with variable degrees of cord deformity usually associated with anterior distortion of the theca only. Sensorimotor cord signs were also clearly related to mobility of the atlanto-axial subluxation, and less clearly to the magnitude of the atlanto-dental separation itself unless it was gross.

It is predictably very difficult to determine the relative importance of such mutually related variables as atlanto-dental separation, mobility, and available space for the cord. Moreover, even in well controlled animal experiments we know how variable can be the response of the cord to trauma³⁷ and compression.³⁸ Our data indicates that in rheumatoid myelopathy it is intermittently applied anterior deforming forces which are of paramount importance. Their effects depend upon their transmission to cord substance, which are maximised when space for the cord is restricted posteriorly whether by bone or pannus. In these circumstances traumatic disruption of cord substance, ischaemic damage due to reduced capillary perfusion, and overt functional failure are most likely to appear. Indeed, it can be speculated that the development of fixed congruous deformity of the cord is part of an adaptive phenomenon tending to protect cord substance from these effects, and the lack of correlation between clinical features, cord deformity or any other radiological variable then becomes more comprehensible. Diverse functional abnormalities possibly emerge from topographical alterations occurring within cord substance which are physiologically incompletely accommodated, and which need not be reflected in histological degeneration.

Prognosis and results of operation

Nakano⁴ has estimated that 30 or 40% of patients with rheumatoid arthritis develop cervical subluxation, and that only 2.5% will develop myelopathy. He has concluded that the myelopathy usually takes at least six years to develop and that within 14 years about 25% will deteriorate, 25% improve and 50% remain static.

In one prospective survey 40% of atlanto-axial subluxations did not progress in seven years.³⁹ However, it has been estimated by others from different clinical materials that one-third of patients with vertical atlanto-axial subluxation die within ten years,⁹ and that as many as 50% or even 100% of patients with myelopathy will die within a year, although not necessarily of cord compression.²⁶ In our material the patients with the shortest clinical course (less than six months) and most progressive clinical features were those who had mobile atlanto-axial subluxation combined with moderate or

severe cord deformity at the cranio-vertebral junction.

Conservative treatment by collar or traction does not seem to significantly alter the clinical course, and traction may occasionally be harmful.⁶⁷¹¹¹⁶ Operative treatment usually consists of posterior fusion following attempted reduction of the dislocation, sometimes combined with laminectomy; and a period of immobilisation in traction for six to 12 weeks following surgery has been the rule.⁷¹⁵⁴⁰ From the results in various publications, one can estimate an average operative mortality of 20% (range 10 to 70%), and that only about 41% of operations (range 0–60%) result in improvement.²⁵⁶¹⁵¹⁶³⁹ The results of transoral odontoidectomy in this series provide a striking contrast: 86% produced sustained improvement and there were no perioperative deaths. One transoral operation had to be abandoned because rotation of the axis had carried the vertebral artery into the operative field anterior to the dens (fig 11). Symptoms of pain, subjective weakness and gait disturbances were most consistently relieved, sensory disturbances were less responsive. We feel that mobilisation on the third post-operative day was an important factor in achieving the low mortality in this series. Unfortunately, stable posterior fusion was achieved in only about half of these cases, although this is no worse than results reported for posterior fusion alone. It is highly significant that failed fusion resulted in recurrent symptoms in only about 14% of these cases compared with 80 to 90% where the dens had not been removed.²⁵⁶¹⁵¹⁶ Sound bony fusion is notoriously difficult to achieve in rheumatoid arthritis. However, these surgical results indicate that abnormal atlanto-axial movement is less significant once the compressing agents, in these cases the dens and pannus, have been removed.

Therefore a one-stage transoral extradural decompression and attempted posterior fusion with early mobilisation seems to present the best prospects of a satisfactory outcome for patients with atlanto-axial subluxation and myelopathy. Those patients with mobile atlanto-axial subluxation and congruous cord deformity are those most likely to progress to death within a few months if not offered operation. The demonstration of fixed deformity of the external form of the spinal cord at the cranio-vertebral junction we regard as an absolute indication for surgery, for this is firm evidence that atlanto-axial subluxation is affecting the cord. Computed myelography is the most sensitive test to detect this change. It is a safe and accurate test provided contrast is introduced via the lumbar route and not by lateral cervical puncture, where there is a danger of injuring the cord with the spinal needle or

encountering a high cervical block and not obtaining information about the state of the lower cervical canal.

Conclusion

- (1) Intermittent anterior pressure on the spinal cord is the cause of rheumatoid myelopathy, and this does not require combined anterior and posterior compression.
- (2) The presence of clinical features is predictable only when there is combined anterior and posterior compression of the cord, and the only predictable features are combined sensorimotor long tract signs.
- (3) Patients most in need of operation are those showing the most rapid clinical deterioration, and these are patients with mobile atlanto-axial subluxation in combination with cord deformity at the cranio-vertebral junction.
- (4) Attempts at fusing the cervical spine often fail in rheumatoid patients, and immobilisation of the whole spine may be required to eliminate all possible consequences ensuing from anterior pressure on the cord.
- (5) Excision of the actual compressing agents, that is dens and pannus, reduces or eliminates clinical manifestation of cord dysfunction even in the presence of mobile subluxation.
- (6) Computed myelopathy using Iohexol introduced by lumbar puncture is the best way to investigate these patients at the present time.

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