LOWER CERVICAL RHEUMATOID SUBLUXATION WITH TETRAPLEGIA

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It has been said that the cervical spine is involved in approximately 40 per cent of patients suffering from rheumatoid arthritis (Garrod 1890, Sharp 1957), and various authors have described the resulting changes (Kornblum, Clayton and Nash 1952, Sharp 1957, Sharp, Purser and Lawrence 1958, Martel and Page 1960, Ball 1961). Sharp and his colleagues in particular described the radiological features and emphasised the frequency of vertebral subluxation of more than mild degree.

Atlanto-axial subluxation has been well recognised especially in ankylosing spondylitis, and the possibility of cord damage, occasionally severe, has been pointed out (Kornblum et al. 1952, Margulies, Katz and Rosenberg 1955, Werne 1956, Sharp and Purser 1957, Hauge 1958, Martel and Page 1960, Ball 1961). However, there have been surprisingly few accounts of the incidence and natural history of vertebral subluxation below the axis, and the few authors who have written on the subject have suggested that associated cord damage is not a problem (Kornblum et al. 1952, Hauge 1958, Sharp et al. 1958, Martel and Page 1960).

Seemingly only two cases have been reported of severe cord damage associated with lower cervical rheumatoid subluxation (Hauge 1958, de Blécourt and Veenstra 1960). In this article four further patients are described who had cord damage with nearly complete tetraplegia. They were treated at one orthopaedic hospital in the last three years. I know of two patients from other hospitals; so the condition is probably not so rare as a study of the literature might suggest.

CASE REPORTS

Case 1—A man then aged thirty-seven began to suffer from rheumatoid arthritis in 1945. A year later he was admitted to hospital with widespread joint involvement, including the cervical spine. He was given treatment but was not given steroids or a support for the neck. He was next seen in early 1964 when he was able to walk with one stick but with pain. On admission in August 1964 he had extensive joint involvement, especially of the left hip and right knee. There was some stiffness of the neck but no abnormal neurological findings. Arthroplasty was done on the right knee in August and cup arthroplasty on the left hip in October. In November the patient complained of numbness in the right hand on the ulnar side, and on examination intrinsic weakness with early claw-hand deformity was found. Radiographs of the cervical spine showed a forward shift of the fourth on the fifth cervical vertebra (Figs. 1 and 2). On the supposition that this might have caused traction on the nerve roots lower down, it was decided to try to reduce the displacement by caliper skull traction in some extension. Radiographically the displacement became less and the patient seemed to lose some of his numbness (Fig. 3). A Minerva plaster was applied incorporating the skull calipers. The following day definite weakness of both hands developed. The plaster was removed and skull traction recommenced; but the neurological signs increased in spite of increasing the amount of traction and extending the neck further, until four days later almost complete tetraplegia at the fifth cervical root level had developed. Some altered sensation was retained in the legs and very weak movements in the fingers of both hands (grade 2). Control of bladder and bowel was lost. At this time the skull caliper traction was replaced by halo fixation (Perry and Nickel 1959, Thompson 1962) preparatory to surgery.

Operation—At operation (December 1964) the dura was taut and not pulsating. It showed an area of constriction where it seemed to have been compressed against one of the neural arches or possibly the infolded ligamentum flavum (Robinson 1957). Gentle probing laterally and anteriorly did not reveal extruded disc material or any other compressive agent. The dura was not opened, but at the end of the operation visible pulsation had returned. Because it was thought that the laminectomy had increased the instability of the cervical spine anterior fusion of the bodies of the fourth and fifth cervical vertebrae with a dowel (Wiltberger 1957, Smith and Robinson 1958) was done.
Progress—After the laminectomy slight improvement in muscle power occurred within two to three days. Recovery continued until, three months after operation, sensation was virtually normal, as was control of bladder and bowel. The patient had some power (grade 3 or more) in all muscle groups, was able to sit out of bed and to feed himself. Within the next month he was able to stand and begin walking. Further rehabilitation was hampered by the general extent of the rheumatoid arthritis.

Case 2—In 1934 a woman then aged thirty-six developed generalised rheumatoid arthritis. In 1958, when she was sixty, steroid treatment was started and continued thereafter. Two years later she complained of paraesthesiae in both hands, thought to be from carpal tunnel syndromes. Operation on the left side afforded no relief. In 1962 she was admitted to hospital because she had become bedridden. After arthrodesis of the right knee and extensive rehabilitation she was discharged from hospital just able to walk. Two years later she complained of increased paraesthesiae and weakness in the hands. After a further month she was almost fully tetraplegic. There was complete loss of motor power below the fifth cervical level; sensory loss below this level was incomplete and patchy in distribution. There was sparing of sacral sensation. Normal control of bladder and bowel was retained. Radiographs of the cervical spine showed rheumatoid changes, with a forward slip of the fourth on the fifth cervical vertebra of about a quarter of the diameter of the body (Fig. 4). The patient was put on halter traction immediately and this was changed to skull caliper traction the next day. No attempt was made to extend the neck, which was kept in the neutral position. Check radiographs showed the vertebral displacement to be reduced. However, the patient’s neurological condition deteriorated; during the next five days she lost bladder and bowel control. Myelography showed an almost complete block at the level of the fifth and sixth cervical vertebrae (Fig. 5). The patient was placed in the halo apparatus before operation. There was immediate improvement in the neurological findings in that sensation returned in the fingers and feet, and some active power of movement of the fingers of both hands and of the left foot was regained.

Operation—At laminectomy (February 1965) the dura was tight under the laminae and there was much adhesion formation between the ligamentum flavum and the dura. When the dura was exposed it was pulseless and very constricted in both planes at about the fifth and sixth cervical level, and here the dissection was carried laterally; on the left side a large piece of sequestrated disc material was
found and removed. Because the dura and cord still remained constricted the dura was divided longitudinally and separated from the pia mater. It was then seen that the dura itself was thickened at the area of the constriction and appeared to be infiltrated with rheumatoid-like granulation tissue (Hauge 1961).

At a subsequent operation two weeks later the bodies of the fourth and fifth cervical vertebrae were fused.

Progress—After operation the patient gradually regained power of movement in the upper limbs and to a less extent in the lower limbs. Sensory recovery was slight but she regained control of bladder and bowel. Five months after operation she was able to feed herself. Because she was almost certain to be chairbound from the rheumatoid arthritis, the recovery in the upper limbs and in bladder and bowel function was rewarding.

Case 3—A woman then aged forty-six developed generalised rheumatoid arthritis in 1947. Various forms of conservative treatment were used and in 1956 steroid therapy was started and continued thereafter. In 1960 she complained of paraesthesiae in both hands. This was not cured by carpal tunnel decompression. In 1964 she was admitted with acute pain in the left knee, thought to be a flare-up of the rheumatoid process; she was then sixty-three. Radiographs of the cervical spine showed extensive changes, with slipping of the third on the fourth, and fifth on the sixth cervical vertebrae (Figs. 6 and 7). Some six months after admission the patient suddenly became shocked and semi-comatose. A diagnosis of an "Addisonian"-like crisis, associated with extensive steroid therapy, was made. Her condition responded to supportive measures, including increased steroid administration. However, the next day she was virtually tetraplegic, with loss of sensation from the fourth cervical level downwards and motor loss, apart from a flicker of movement in the toes, below the fifth cervical segment. Skull traction with calipers was started but there was no neurological improvement; her general condition forbade exploration of the cervical spine and she died one month later without any recovery of the tetraplegia.

Necropsy—The cervical spine showed gross rheumatoid involvement (Fig. 8); there was destruction of the cervical three-four disc space with a ridge pressing backwards at this level consisting of soft
Case 3. Figures 6 and 7—Active flexion and extension lateral radiographs showing unstable subluxations between the third and fourth and fifth and sixth cervical vertebrae. Note the severe destructive changes. Figure 8—Sagittal section of the cervical spine and cord. Note the disruption of the disc space between the third and fourth cervical vertebrae, narrowing of the cord at this level by soft-tissue projections anteriorly and posteriorly, and the fracture of the body of the fifth cervical vertebra with subdural haematoma and further cord narrowing.

Case 4—A woman then aged sixty-eight was admitted to hospital in 1962 having had rheumatoid arthritis for some thirty years with involvement of virtually all peripheral joints. For the last three and a half years she had been confined to bed and during this time had been on steroid therapy. On admission she said she had had increasing weakness of the arms and legs and persistent severe paraesthesiae in the hands for three months. Examination showed advanced generalised rheumatoid arthritis with particularly severe deformity of the elbows and hands. She had a moon face and a thoracic kyphosis, apparently of recent onset, suggestive of steroid overdosage. Radiographs confirmed that she was osteoporotic; those of the cervical spine showed subluxation of the second cervical vertebra forwards on the third, and the third backwards on the fourth. The cervical vertebral bodies showed erosions, particularly the second, third and fifth. The fourth cervical vertebra was particularly osteoporotic (Fig. 9).

FIG. 9
Case 4—A lateral radiograph of the cervical spine, showing severe subluxation forward of the second on the third, and backwards of the third on the fourth cervical vertebrae. There is much erosion of vertebral bodies and there is osteoporosis.
Neurological examination revealed an incomplete spastic tetraplegia from the third cervical segment. Bladder and bowel control was lost.

Progress—It was thought that she was suffering from cord compression caused by displacement of the cervical vertebrae, but her extremely frail general condition and her already bedridden state contra-indicated operation. Halter traction was applied but not tolerated; a collar did not produce improvement and her neurological state deteriorated. She died six months later.

DISCUSSION

In these four patients long-standing severe rheumatoid arthritis had extensively involved the cervical spines, leading to subluxation. Cord damage occurred late in the progress of the disease, though two patients had developed cervical nerve root symptoms much earlier. Three of the four patients had had prolonged steroid therapy and at least one of these showed obvious osteoporosis. Considering the cause of the cord damage, it is likely that compression was present in all four patients, though the factors causing the compression were varied. Whether the damage to the nerve tissues was caused directly by the compression or whether it was secondary to interference with the blood supply is not certain.

In the first patient it is difficult to avoid the conclusion that cord damage was brought on by attempts to reduce the vertebral subluxation by powerful skull traction and extension of the neck. At operation the dura was found compressed at the back against the neural arches, presumably by tension in the soft tissues because there were no bony or other factors causing compression. Rheumatoid cervical subluxations are likely to occur slowly with associated soft tissue fibrosis. Forcible attempts to alter the position may exert abnormal tension on the cord.

In the second patient it is obvious that the vertebral subluxation was not responsible for the compression of the cord, which was present one segment lower. The compression was caused, first by sequestred disc material, and second by infiltration of the dura with rheumatoid granulation tissue, possibly as in cases described by Hauge (1961).

In the third patient the clinical level of cord damage corresponded to the unstable segment; necropsy confirmed compression at this level, probably by soft tissue rather than bone, and present posteriorly as well as anteriorly. It is likely that the cord lesion at this level was precipitated by handling the patient in her severely ill, semi-comatose state. The fracture in the body of the fifth cervical vertebra probably occurred just before death as a lethal event (Lemmen and Laing 1959).

The last patient showed the most extensive vertebral subluxation in the series, and the clinical features were consistent with an incomplete cord lesion at this level. It is only in this patient that it is likely that the bony displacement narrowed the spinal canal sufficiently to cause cord compression.

It is important to note that cervical traction did not improve any of these patients and in two appeared to be harmful. I have knowledge of a further patient in whom similar deterioration occurred (Jeffreys 1965), and two cases reported by Hauge (1958) are relevant.

The treatment of the cervical spines of rheumatoid patients who have radiological instability is with a protective collar. No forcible attempt should be made to reduce vertebral subluxation and extreme care should be exercised if a general anaesthetic is administered. It is possible that continued support of the neck will result in fusion of the unstable segments. That this can happen is shown in the first patient who obviously had a spontaneous fusion between the sixth and seventh cervical vertebrae (Figs. 1 and 2). The nerve root symptoms may also benefit from a supportive collar or from a period of more rigid fixation in a plaster or halo splint. If severe symptoms persist fusion of the cervical spine may be called for, though operation should not be undertaken lightly.

Evidence of cord damage creates a serious problem requiring urgent attention. Immediate rigid immobilisation of the cervical spine, preferably in a halo splint, is advisable without
attempting to reduce vertebral subluxation, and certainly avoiding extension. If the symptoms of cord damage are slight it is worth waiting to see if recovery will take place. If it does, continued support may be all that is necessary, though spinal fusion is probably indicated if the vertebral subluxation is other than trivial. If the neurological condition does not recover, if it progresses, or if it is severe from the outset, cervical myelography should be done to localise any compressive lesion. Exploration of the cord by generous laminectomy should follow in most cases, allowing all constricting factors to be dealt with. Subsequently the spine will be even more unstable and fusion is needed. This is done more simply by the anterior method and should be carried out before removal of the halo fixation.

These suggestions for treatment, especially the operative procedures, may not be applicable to certain patients whose general condition is extremely poor.

SUMMARY
1. Four patients with tetraplegia from rheumatoid subluxation occurring in the lower cervical spine are described and the common features are noted.
2. Compression appeared to be responsible for the cord damage, although it was not necessarily directly related to vertebral subluxation. The lesions causing the compression were varied.
3. Forcible attempts to reduce vertebral subluxation may be harmful. Two patients were improved by posterior decompression.
4. A suggested programme for the treatment of such patients is outlined.

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REFERENCES