Management of cervical spinal cord injury in ankylosing spondylitis: the intervertebral disc as a cause of cord compression

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Twenty-one patients with universal syndesmophytopsis due to ankylosing spondylitis were identified in a consecutive series of 1578 patients with acute spine and spinal cord injuries. They were predominantly male, older than spinal cord-injured patients in general, and most were injured by falls. Approximately one-half were managed by halo-vest immobilization alone with good clinical and radiological outcomes. The remainder required surgery either for recurrent dislocation or for spinal cord compression associated with neurological deterioration. Extrudal hematoma, a recognized cause of spinal cord compression in ankylosing spondylitis patients with spinal fractures, was encountered in two patients. Herniated intervertebral disc as a cause of spinal cord compression in ankylosing spondylitis does not appear to have been previously reported and was recognized three times in the present series, once in association with extradural hematoma. The pathology of ankylosing spondylitis is such that the nucleus pulposus tends to be spared, allowing disc herniation to occur in the heavily ossified spine. In virtually all patients, satisfactory correction of the flexion deformity could be safely accomplished following spinal fracture.

It is concluded that fracture/dislocations of the cervical spine should be managed initially by halo-vest immobilization, without prior traction and with careful incremental correction of flexion deformity. Decompression is performed as required for extradural hematoma or intervertebral disc herniation, and internal fixation is carried out for recurrent dislocation.

Key Words: ankylosing spondylitis • spinal cord injury • cervical spine • spinal extradural hematoma • intervertebral disc herniation • halo vest

ANKYLOSING spondylitis increases the risk of cervical spine and spinal cord injury as a result of relatively minor trauma, frequently due to falls. In these cases, neurological deterioration after injury is not uncommon. Spinal cord compression due to recurrent dislocation or to extradural compressive lesions occurs relatively frequently and may require surgical intervention. Universal syndesmophytopsis ("bamboo" or "railroad track" spine) causes the spine to resemble a long bone and makes immobilization more difficult.

Our experience with 21 consecutive cases of cervical injury complicating long-standing ankylosing spondylitis and the management principles that have evolved during that experience serve as the basis for the following report.

Clinical Material and Methods

The present series is derived from 1578 patients consecutively admitted to the Acute Spinal Cord Injury Unit at Sunnybrook Health Science Centre between 1974 and 1990. Of these, 1028 injuries involved the cervical spine. Twenty-one patients with pre-existing ankylosing spondylitis were identified from the records based on historical and radiological evidence. All of the patients in this series had known ankylosing spondylitis of many years' duration, with radiographic evidence of universal syndesmophytopsis. Patient follow-up data were obtained from hospital and clinic records and by telephone from patients or their physicians. The radiological follow-up study was performed by re-examination of films.

Results

Patient Population

The 21 patients with pre-existing severe ankylosing spondylitis constituted 2% of our consecutive series of 1028 patients with cervical spine injuries; 16 (3.5%) of
them were among the 462 cases of cervical spinal cord injuries. There were 19 men and two women, ranging in age from 35 to 77 years (mean 61 years). In 14 patients, the duration of ankylosing spondylitis prior to spinal cord injury could be accurately established and ranged from 7 to 40 years (mean 24.4 years).

Falls were the cause of injury in 15 patients, and motor-vehicle accidents in five. Two of the motor-vehicle accidents were car-pedestrian mishaps in which the patient was struck by a slow-moving vehicle, perhaps caused by the patient's severely kyphotic posture which prevented his seeing the approaching vehicle. As determined from patient histories or from eyewitness accounts, the mechanism of injury appeared to be flexion, with or without axial compression, in nine patients and extension in four. In the remaining eight patients, no reliable account of the injury could be obtained.

Severity of Spinal Cord Injury

Sixteen (76%) of the 21 patients had spinal cord injuries as well as spine injuries, while five had spine injuries only. Despite careful management, four patients (19%) showed some degree of neurological deterioration. Four patients (19%) sustained initially complete neurological deficits, but seven (33%) had complete neurological deficits on final follow-up examination. In other words, three additional patients deteriorated from incomplete to complete injuries.

Radiological Findings

All 21 patients sustained fractures through a cervical disc space or vertebral body plus posterior elements, with some degree of displacement in most instances. In 14 patients, displacement was at least 25% of the anteroposterior body diameter. An additional component of body compression was seen in four patients. In six patients, distraction was identified radiographically at some time during early clinical management, and two of these deteriorated clinically, one to a complete deficit.

The most frequent level of injury was C6–7, seen in 10 patients, with C4–5 injuries occurring in four and C5–6 in three. Two levels of spine injury were identified radiographically in five patients.

Management

Initial stabilization of cervical injuries was effected by means of a halo vest. In all, 11 patients were managed without surgical intervention.

Ten patients underwent surgical stabilization and/or decompression. Six were initially stabilized and posterior fusion was performed because of difficulty with or concern about maintaining alignment and immobilization in a halo vest. Four patients required decompressive procedures because of clinical neurological deterioration associated with myelographically demonstrated cord compression.

Operative Findings

A herniated intervertebral disc was found to be producing cord compression in three patients (associated with complete myelographic block). In one of these, extradural hematoma was also present and may have contributed to cord compression. Extradural hematoma was identified in one additional patient at autopsy. Soft-tissue disruption was frequently extensive. In several patients there was laceration of the anterior longitudinal ligament and either laceration of or hematoma within the longus colli muscles. Anterior dural laceration was also frequently seen.

Neurological Outcome

Of the 11 patients who were managed conservatively, three initially had complete and eight had incomplete neurological deficits. These proportions remained unchanged at the time of final follow-up examination. Only one (9%) of 11 patients deteriorated neurologically; two died, two remained chronically hospitalized, and seven (64%) are able to walk and/or work.

Three of 10 surgically treated patients were operated on urgently for spinal cord compression resulting from disc herniation or extradural hematoma. One of these patients continued to deteriorate despite apparently adequate decompression, suffered a complete neurological deficit, and ultimately died. The other two did not deteriorate further, but did not experience major recovery and remained chronically hospitalized at the time of final follow-up study. Of the remaining seven patients who were stable neurologically at the time of surgery, two deteriorated to complete lesions and one remained unchanged with a complete lesion. Thus, of the six surgically treated survivors, three remain hospitalized, one is at home but not able to walk or work, and two have become employable.

Mortality and Autopsy Findings

There were six deaths (29%). Two of these were in nonsurgical patients and four were in the surgically treated group. The cause of death in all cases was respiratory. Autopsies were performed on two patients. In one, a spinal extradural hematoma was found, probably accounting for the anterior cord syndrome exhibited clinically by the patient. In the second patient, the spinal cord was completely transected at C4–5, where there was fracture/dislocation.

Functional Outcome on Follow-Up Examination

Of the 15 survivors, six (40%) remained chronically hospitalized as a result of their injury, eight (53%) are capable of walking, and eight (53%) are able to work. The follow-up period for the survivors varies from 4 to 178 months (mean 68 months).

All 16 patients who survived 12 weeks or longer ultimately demonstrated solid bone union at the fracture site, confirmed by tomography and dynamic lateral radiographs in both flexion and extension. Of these 16
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patients, x-ray films adequate to permit measurement of final angulation were available for 11. Nine showed substantial correction of their flexion deformities with lordosis of 6° to 30° (Fig. 1). The remaining two demonstrated kyphosis of 7° and 19°, respectively.

Discussion

Patient Presentation

Severe ankylosing spondylitis was found in 3.5% of the present series, supporting the belief that patients afflicted with this disorder are more likely to sustain cervical spinal cord injury.2,4,21 In this series, patients had been known to have ankylosing spondylitis for a mean period of almost 25 years at the time of injury. From this, it is evident that the spine must be severely affected before the patient is significantly predisposed to injury.2,4,9,11

The mean age of these patients was 61 years, compared with 31.5 ± 13.7 years (± standard deviation) in an unselected series of patients with cervical spine injuries previously reported from the same center.9 Ankylosing spondylitis affects men more commonly than women, particularly when severe, as demonstrated by the striking male preponderance in this and other reported series of ankylosing spondylitis.2,7,9,11,12,16,18

Mechanism of Injury

The tendency of ankylosing spondylitis patients to sustain injury from minor trauma, particularly falls, has been noted in previous reports.2,4,11,16 Weinstein, et al., 21 noted that an inability to see the ground ahead while walking due to extreme flexion deformity of the cervical spine contributed to injury in their patients. This was also apparently the case in the present series.

Flexion appeared to be the most frequent mechanism of injury in our series, although the mechanism still remains uncertain in more than one-third of the cases. Most reported series identify hyperextension as the most common mechanism of injury.2,7,14 Given the flexion deformity of the lower cervical spine, it seems probable that either extension or flexion may produce fracture/dislocation. Since falls are the usual cause of injury,2,4,7,18 it is perhaps not surprising that extension predominates as a mechanism of injury in most cases.

Severity of Spinal Cord Injury

Our observation that 76% of the patients had spinal cord injuries as well as spine injuries is in keeping with reported experience.2,11,14 The severity of neurological injury complicating spinal fracture appears to be higher in patients with ankylosing spondylitis. Murray and Persellin14 found that 57% of ankylosing spondylitis patients who sustained spinal fractures suffered severe neurological injury compared with 18% of patients with previously normal spines. Similar data are reported by others.2,11 A relatively high incidence of neurological deterioration has also frequently been noted in previous reports.2,7,14,21

Radiological Findings

Fractures of anterior and posterior elements were easily identified on plain x-ray films in most instances, although tomography frequently revealed additional bone injuries.22 Distraction deserves particular comment, because it was observed in six patients in our series, two of whom deteriorated clinically. Distraction is more likely in ankylosing spondylitis patients because calcification of anulus and ligaments causes the cervical spine to behave like a fractured long bone, and also because of a loss of muscle tone related to long-standing ankylosis. Deterioration with distraction has been observed and reported previously.14

Level of Injury

The C6–7 vertebral level was the site of injury in about one-half of the patients in the present series, and has also been the most frequent level in several other reported series.1,12,17 In the literature review carried out by Murray and Persellin,14 21 cases of C5–6 fracture and 16 cases of C6–7 fracture were identified among 83 reported cases.

Management

We now avoid traction prior to placement of the halo vest because it is not required for reduction in ankylosing spondylitis patients, and the risk of distraction with resulting neurological deterioration is thus avoided. We disagree with DeWald and Ray,1 who recommended routine reduction by axial traction. In patients managed by halo-vest immobilization alone, without operative intervention, the incidence of deterioration was low, in agreement with previous reports.8,1,12,14,24,25

Four patients experienced recurrent dislocation while
immobilized in a halo vest, but only one deteriorated clinically. It is possible that a return to halo traction or a longer period of bed rest after application of the halo vest would have obviated a need for surgical stabilization. Sweet and Borges (unpublished data) have suggested an alternative approach to conservative management in the highly unstable patient; they advocated a return to halo-pelvic fixation.2

Experience with anterior decompresive surgery in ankylosing spondylitis patients demonstrates that the incidence of cord compression is high. It is also apparent that, even with the additional stability provided by the halo vest, anterior decompresive surgery should probably always be combined with stabilization, either posteriorly or anteriorly, as better methods of anterior fixation evolve.

**Operative Findings**

_Cord Compression by Spinal Extradural Hematoma._ Two of our 10 surgically treated patients suffered spinal cord compression by epidural hematoma, a recognized complication of spinal fracture in ankylosing spondylitis patients.1,6,0,14 and an occasional occurrence in the absence of fracture.6 The reported incidence of epidural hematoma in ankylosing spondylitis is high, ranging from 20%7 to 50%.1 Spinal epidural hematoma was observed in two (10%) of the 21 patients in the present series; one was identified at the time of anterior decompresive surgery in association with intervertebral disc herniation at the same level, the other was identified at autopsy and was believed to have been clinically significant because the patient had suffered from an antemortem anterior cord syndrome. Interestingly enough, Kiwerski, et al.,12 encountered neither epidural hematoma nor intervertebral disc herniation causing cord compression in their 33 patients, although only two cases were managed surgically. Bohlman1 reported a 50% incidence of epidural hematoma (four of eight cases) in cervical fractures associated with ankylosing spondylitis and noted that the results of decompressive laminectomy tended to be poor. Epidural hematoma was not encountered in any of the remaining 292 patients in that series. Hunter and Dubo11 encountered nine cases of spinal epidural hematoma (17%) in 54 reported cases; however, they did not identify epidural hematoma in any of their own 20 patients, perhaps because a relatively high percentage of their patients were diagnosed after a lengthy delay. Foo and Rossier12 reviewed 38 cases of spinal epidural hematoma from the literature and noted the frequent association of ankylosing spondylitis with posttraumatic spinal epidural hematoma. They also noted that the prognosis for recovery after surgical decompression was better in cases of spinal epidural hematoma not associated with ankylosing spondylitis and spinal fracture.

_Cord Compression by Intravertebral Disc Herniation._ In ankylosing spondylitis, ossification occurs in the anulus fibrosus of the intervertebral disc and in the apophyseal joints.3,21 Universal syndesmophytosis renders the spine immobile with a loss of elasticity at the disc spaces and apophyseal joints. Because the disease affects the enthes,3 it should be remembered that the nucleus pulposus tends to be spared from the ossifying process.31 Three (14%) of 21 patients in the present series suffered spinal cord compression from herniated intervertebral disc, an entity that does not appear to have been recognized previously. It is important to recognize that intervertebral disc herniation may be an additional cause of non-bony compression of the spinal cord in ankylosing spondylitis. Disc herniation was more frequent than extradural hematoma in our experience and may cause more severe compression of the spinal cord than epidural hematoma, which is distributed more diffusely in the extradural space. Since most previous authors reporting ankylosing spondylitis associated with spinal epidural hematoma have employed laminectomy as a surgical approach, it is possible that some cases of intervertebral disc herniation have gone unrecognized and untreated.

Our overall incidence of non-bony spinal cord compression was thus 19% (four of 21 cases), as one patient had both an epidural hematoma and a herniated intervertebral disc. It is suggested that, since one (25%) of four patients with ankylosing spondylitis and spinal cord injury suffered from non-bony spinal cord compression, myelogram/computed tomography or magnetic resonance imaging followed by decompresive surgery in deteriorating patients may be important.

**Neurological Outcome**

The conservatively managed patients in this series had generally better outcomes at final follow-up examination than did those managed surgically, with seven (64%) of the 11 experiencing good to excellent recovery. Better outcomes in this group do not necessarily indicate that nonsurgical management is superior,14 since the groups are not necessarily comparable.

Surgical decompression in a deteriorating patient appears often to be followed by a poor result, and patients who are neurologically unstable at the time of surgery seem more likely to deteriorate as a result of surgery.1,6 Good to excellent results were achieved in only two (20%) of 10 patients in the surgically treated group. Anterior decompressive surgery must be advised for the patient who is deteriorating neurologically with a demonstrated epidural hematoma or disc herniation, but it is difficult to escape the conclusion that, even with great care and with continuous intraoperative monitoring, the probability of good recovery is poor. It may be preferable for the patient who is deteriorating neurologically because of recurrent dislocation to continue bed rest in the halo vest or possibly halo-pelvic fixation to arrest the progression of deficit, resorting to early surgical stabilization only if this strategy fails. Despite the appeal of early internal fixation, as advised by Detwiler, et al.,4 our surgical results do not justify an aggressive approach to early surgery as routine management.
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Morbidity and Mortality Rates

Reported mortality rates are commonly in the 30% to 35% range for ankylosing spondylitis patients with cervical spine injury and higher for those with cord injuries. The mortality rate associated with spinal fractures occurring in patients with previously normal cervical spines is about half as great, at 17% or 18%. Elderly patients with cervical cord injuries and immobile chest walls can be expected to suffer a high incidence of atelectasis and bronchopneumonia, and vigorous prophylaxis and treatment are indicated. The incidence of pulmonary edema was high in this series although no single cause was evident. A majority of ankylosing spondylitis patients with cervical cord injury require early tracheostomy.

Functional Outcome

Reported series of ankylosing spondylitis patients with cervical cord injury frequently lack specific information on the long-term neurological outcome in terms of activities of daily living. Broom and Raycroft reported that only one (20%) of their five patients achieved independence in activities of daily living; however, their follow-up period was relatively brief. None of the four patients with cervical cord injury reported by Guttmann achieved functional independence. Hunter and Dubo reported major neurological recovery in five (45%) of 11 cases of cervical cord injuries. The status of these patients with respect to employment is not clear; they were definitely ambulatory, but had residual weakness in their hands. Murray and Persellin reported good functional recovery in three (43%) of their seven cervical cord-injured patients and, not surprisingly, were unable to supplement their data on this important aspect of outcome from their extensive review of the literature.

Ohry and Frankel have illustrated the increased difficulty of rehabilitation of cord-injured patients with pre-existing ankylosing spondylitis. Such patients tend to recover less well than patients with comparable injuries but with previously normal spines.

The present series, with six patients (40%) chronically hospitalized, eight capable of walking, and eight able to work, compares well with the best of reported experience.

Radiological Follow-Up Study

Most patients with long-standing ankylosing spondylitis of the cervical spine have relatively severe flexion deformities, and it has been accepted as conventional wisdom that the spine should be immobilized in its usual position of flexion. Cervical osteotomy is carried out therapeutically in patients with intact spines in order to correct severe flexion deformity and interference with the activities of daily living such as limitations in the field of vision or mouth opening. Substantial correction can be achieved safely, however, particularly in the wake of an acute spinal cord injury, the correction of spinal deformity must be approached cautiously. Final measurements of angulation in the present series indicate that the spine in a majority of our patients was corrected to a position of slight extension, simulating the normal lordotic curve of the cervical spine, without worsening neurological deficit (Fig. 1 right). Correction should be achieved gradually by incremental adjustment of the halo vest, and distraction and dislocation must be rigorously avoided. The correction of flexion deformities has highly salutary effects on the patient's daily activities, and we would encourage others to take the opportunity presented by a fracture through all of the anterior and posterior elements to attempt some gentle correction of the flexion deformity. Good results achievable by conservative treatment are once again acknowledged, with the corollary that these very unstable spines may be successfully managed by halo-vest immobilization in the supine position for longer if necessary, or possibly by halo-pelvic fixation. Even this measure may not suffice to arrest neurological deterioration in all instances, leaving some patients who will require surgical stabilization despite the apparent risks.

References


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