Neurological complications of ankylosing spondylitis

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Thirty-three patients with ankylosing spondylitis with or without spinal neurological symptoms requiring surgical intervention were identified in a retrospective review of all cases of spinal disorders treated at the Mayo Clinic during the period from 1984 to 1989. Spinal fractures (traumatic or pseudarthroses), progressive spinal deformity, rotary instability secondary to atlanto-occipital or atlantoaxial subluxation, and spinal stenosis with associated neurological deficit, pain, or spinal instability were the most common indications for surgery. Of the 41 operations performed, there were 17 cervical, 14 thoracic, and 10 lumbar procedures. Eight patients had two or more disorders at separate spinal levels that required surgery. Preoperative neurological deficits were recorded in 13 patients; these improved following surgery in nine and stabilized in four.

This review emphasizes the fragility of the ankylosed spinal column and its inherent susceptibility to fracture with attendant neurological compromise. It also identifies the variety of spinal disorders observed in these patients and discusses their surgical management. The experience with this series suggests that thorough radiological evaluation, extreme caution with endotracheal intubation and with halo-vest applications, early surgical spinal immobilization, and aggressive postoperative mobilization of patients are necessary for a successful outcome. It is concluded that outcomes in these patients can be favorable when managed appropriately.

Key Words • ankylosing spondylitis • spinal stabilization • surgical complication • halo vest

Ankylosing spondylitis is a common rheumatological disorder of adults, with an estimated incidence of 0.4% in the general population. This condition results in disease-specific inflammation at the site of ligamentous insertion into bone. The sacroiliac joints and the lumbar spine are most commonly affected, but with advanced disease the entire spinal column may be involved. Diarthrodial spinal facet joints may also develop synovitis. For unknown reasons, this inflammation leads to a profound tendency for ossification of involved ligaments and joints, with fusion of the spinal column which then functions as a long bone housing the spinal cord. Movement and elasticity of the spinal column are lost. Altered biomechanics of the immobile ankylosed spine render it susceptible to a variety of disorders including fracture/dislocation, progressive spinal deformity, atlanto-occipital or atlantoaxial subluxation, and spinal stenosis. In addition, the rigid spine may secondarily develop osteoporosis, further increasing the risk of spinal fracture.

This study reviews patients with ankylosing spondylitis who were surgically treated for complications of their disease in order to identify the different spinal disorders observed and to better define their management. Criteria for selecting external immobilization versus surgical stabilization with or without decompression are discussed. In addition, the surgical techniques, intraoperative difficulties, and complications observed in these complex cases are reviewed.

Clinical Material and Methods

Patient Population

During the period between 1984 and 1989, 33 patients with neurological complications of ankylosing spondylitis were treated at our institution. All medical and surgical records and the radiological studies were examined for this review. There were 27 men and six women, with an average age of 53 years (range 22 to 83 years). Follow-up evaluations were available for all patients, with intervals ranging from 12 to 77 months (mean 39.1 months). In eight of the 33 patients, two or more spinal disorders at separate spinal levels were diagnosed during the study period. The 41 procedures performed included 17 cervical, 14 thoracic, and 10 lumbar operations.

Preoperative Radiological Evaluation

All patients had plain spine films performed initially (Fig. 1); when necessary to better define the spinal disorder, pluridirectional tomography and/or comput-
M. W. Fox, B. M. Onofrio, and J. E. Kilgore

Plain lateral cervical spine x-ray film of an acute C4-5 fracture/dislocation through both anterior and posterior elements (arrow) following emergency endotracheal intubation. There is fusion of the facet joints and ossification across the disc spaces and the anterior longitudinal ligament.

Computerized tomography were also obtained. Magnetic resonance imaging was performed in four patients with suspected thoracic pseudoarthroses. Computerized tomography with or without myelography was used in all patients with neurological deficits due to its superiority in the assessment of ankylosed bones and ossified supporting structures and their relationship to the spinal canal and thecal sac (Fig. 2).

Spinal Fractures

Twenty-eight spinal fractures were observed in 23 patients. Of these, 11 (nine traumatic and two stress fractures) occurred in the cervical spine, 14 (three traumatic and 11 stress fractures) were found in the thoracic spine, and three stress fractures were diagnosed in the lumbar spine. In two patients multiple fractures were diagnosed concurrently, while three patients had multiple fractures identified at different times.

The 12 traumatic fractures occurred in 11 patients: six resulted from falls while standing, walking, or from a height of less than 4 feet; three were sustained in motor-vehicle accidents; one was incurred during heavy lifting; and two occurred during emergency endotracheal intubation. Seven of these patients presented acutely, while four had delayed presentation ranging from 1 month to 3 years following injury. Three of these patients had a pseudoarthrosis at the fracture site. The average age of patients with acute-presentation fractures was 67 years (range 35 to 83 years).

Twelve patients with 16 stress fractures or pseudoarthroses presented with long-standing pain or progressive spinal deformity but no history of previous neck or back trauma. Patients described symptoms of new neck or back pain and/or a progressive bent-over posture for periods ranging from 2 months to many years prior to presentation. These patients ranged in age from 33 to 71 years (mean 54 years).

Other Spinal Disorders

Six patients were identified with progressive spinal kyphosis without fracture or pseudoarthrosis. All had long-standing low-back pain and deformity, which disallowed vision above a horizontal view line. One patient had respiratory compromise resulting from his deformity. The average age of these patients was 38 years (range 35 to 41 years).

Rotary instability secondary to atlanto-occipital subluxation without fracture was observed in three patients; two of these had atlantoaxial subluxation as well. Two additional patients had atlantoaxial instability only. The average age of these patients was 49 years (range 22 to 66 years).

Cervical spinal stenosis was diagnosed in three patients with progressive myelopathies, and lumbar stenosis was found in one patient presenting with pseudoclaudication. These patients were aged 35, 47, 66, and 82 years, respectively.

Thirteen patients had neurological compromise prior to surgical intervention. Five patients developed deficits following traumatic fractures, including cervical myelopathy in four and a unilateral cervical radiculopathy in one. One patient suffered moderate upper-extremity paresis and T-12 level paraplegia after sustaining a C4-5 fracture/dislocation and displacement of a missed L-1 fracture following emergency endotracheal intubation after a motor-vehicle accident. One additional patient became quadriplegic following emergency intubation for respiratory distress resulting from a motor-vehicle accident.

Two patients with thoracic stress fractures had long-
Neurological complications of anklyosing spondylitis

**Fig. 3.** A: Illustration of a cervical spine fracture (acute or stress) with no malalignment or associated neurological deficit. B: The suggested management includes spinous process wiring with bone grafting and halo-vest immobilization.

**Fig. 4.** A: Illustration of a stable thoracic pseudoarthrosis without neurological deficit. B: The suggested management involves posterior rod and bone fusion.

**Fig. 5.** A: Illustration of an unstable thoracic pseudoarthrosis without neurological deficit. B: The suggested management includes excision of the pseudoarthrosis with strut grafting followed by posterior rod and bone fusion.

**Fig. 6.** A: Illustration of a displaced acute cervical spine fracture with associated neurological deficit. Note the anterior dural deformity. B: The suggested management includes partial corpectomies and strut grafting, followed by a posterior spinous process wiring and bone fusion.

Standing mild cauda equina syndromes preoperatively. All three patients with cervical spinal stenosis had mild myelopathies prior to surgery. The single patient with lumbar spinal stenosis had both unilateral radiculopathy and pseudoclaudication.

**Surgical Management**

**Fractures.** Two patients with cervical spine fractures were initially managed with halo-vest immobilization. One patient with an acute-presentation C3–4 fracture was left quadriplegic after emergency intubation following a motor-vehicle accident. He was transferred to our institution in critical condition and died from pulmonary complications while in halo traction prior to surgical intervention. One other patient, who underwent fusion of an acute T9–10 interspace fracture suffered during a motor-vehicle accident, also had a questionable C6–7 fracture with no neck pain and was placed in a halo vest postoperatively.

Four different surgical procedures were used for stabilization of fractures in patients with anklyosing spondylitis. Patients who had acute-presentation fractures or pseudoarthroses without spinal malalignment or neurological deficit underwent posterior fusion (Figs. 3 and 4). Fractures or pseudoarthroses associated with severe spinal instability but no neurological deficit were fused both anteriorly and posteriorly. For example, thoracic pseudoarthroses causing instability were excised anteriorly, then fused with an iliac crest, rib, or fibular strut graft followed by posterior fusion (Fig. 5). The surgical procedure used for patients with a neurological deficit was based on the alignment of the spine and the location of thecal sac compression. If the patient had anterior cord compression, an anterior decompression was performed followed by strut grafting and posterior fusion (Fig. 6). If there was no evidence
of anterior cord compression, then a decompressive laminectomy was undertaken followed by posterior fusion. Patients with cervical spine fractures or pseudoarthroses were maintained in a halo vest for 3 to 6 months until radiologically verifiable fusion could be demonstrated. Patients with thoracolumbar fractures wore custom-fitted polypropylene spinal orthoses for 6 to 12 months. Table 1 summarizes the surgical procedures used in patients with spinal fractures in our series.

**Spinal Deformity.** Five patients underwent posterior wedge osteotomy at the L2-3 level, Harrington rod instrumentation, and iliac crest bone grafting for progressive spinal kyphosis (Fig. 7). One patient underwent both anterior and posterior osteotomies and fusions for correction of a severe spinal deformity. All patients were kept in a spinal orthosis until fusion was demonstrated.

**Rotary Instability.** There were five patients with a combination of seven atlanto-occipital or atlantoaxial subluxations without fracture. All patients were neurologically normal. One patient underwent C1–2 spinous process wiring and iliac crest bone grafting. Two patients with atlanto-occipital instability and both patients with combined atlanto-occipital and atlantoaxial instability were managed with occiput to C-2 spinous process wiring and bone grafting. All patients were immobilized in halo vests for at least 3 months postoperatively.

**Spinal Stenosis.** All patients with spinal stenosis underwent standard decompressive laminectomies. Facet wiring and bone grafting were performed in one patient following C4–7 laminectomy for an associated C-5 on C-6 pseudoarthrosis. This patient was kept in a halo vest for 3 months.

**Illustrative Case**

This 66-year-old man had a 25-year history of ankylosing spondylitis with known mild cauda equina syndrome for 9 years manifesting as bowel and bladder incontinence, bilateral S-1 weakness, and sacral numbness. The patient presented with a 4-month history of new thoracic back pain which was exacerbated by movement. No additional neurological deficits were noted and he had no history of back trauma. X-ray films revealed a T11–12 pseudoarthrosis (Fig. 8). External immobilization for 2 months did not result in pain relief or fusion. The patient then underwent excision of the pseudoarthrosis and adjacent endplates, followed by anterior and posterior fusions from T-9 through L-1. Postoperatively, the patient achieved stable fusion and pain relief. His neurological status was unchanged.

**Results**

**Surgical Outcome**

Twenty-three patients underwent surgical stabilization for 28 different fractures/dislocations. One patient died from pulmonary complications following a trans-
Neurological complications of ankylosing spondylitis

FIG. 8. X-ray films, anteroposterior (left) and lateral (right) views, of the spine demonstrating a T11-12 pseudoarthrosis (arrows) in a patient with long-standing cauda equina syndrome and new neck pain of 4-month duration. Note the sclerotic endplates.

Fusion reaction during the early postoperative period. Another patient with a cervical spine fracture died from pulmonary complications prior to surgery while immobilized in halo traction. Of the remaining 21 patients, all had postoperative radiographs demonstrating solid fusion at follow-up periods ranging from 12 to 77 months (mean 39.1 months). All six patients with progressive spinal deformity achieved partial correction following surgery. All patients with rotary instability achieved stable fusion. Patients with decompressive surgery for stenosis showed no instability on follow-up x-ray studies.

Neurological Outcome

Thirteen patients had preoperative neurological deficits which improved in nine patients and were stabilized in four patients following surgery. Five patients with acute cervical spine fractures had mild improvement of their preoperative myelopathies, one showed no improvement, and one patient died prior to surgery. Neither patient with a preoperative cauda equina syndrome and thoracolumbar pseudoarthrosis improved following decompression and fusion. Partial improvement was observed in all four patients who underwent spinal stenosis surgery.

Complications

Missed Fractures. Three fractures were missed at initial radiological evaluation for new neck or back pain. Two fractures were subsequently identified due to persistent pain despite external immobilization with Philadelphia collars. Both patients underwent surgical fusion without complication. The third patient became paraplegic after he was moved from his bed to a gurney with displacement of a missed L-1 fracture. He had no recovery of function following decompressive surgery and stabilization.

Postoperative Neurological Deficit. Two patients developed new neurological deficits following surgery. One patient suffered progressive paraparesis 2 weeks after L2-3 anterior and posterior osteotomies with Harrington rod instrumentation and bone grafting for correction of a spinal deformity. A decompressive L2-3 laminectomy was performed with replacement of the grafting materials, resulting in complete resolution of symptoms. Another patient experienced new pain of the right arm and shoulder with a mild right C-6 radiculopathy following occiput-to-C-2 fusion for combined atlanto-occipital and atlantoaxial subluxation. X-ray films revealed a new C5-6 fracture without subluxation. It was suspected that this had occurred during halo-vest application at the time of surgery. The patient was managed conservatively in halo-vest immobilization with eventual resolution of pain and nearly complete recovery of the C-6 weakness.

One additional patient underwent a decompressive laminectomy and posterior fusion for an acute displaced T9-10 fracture and halo-vest application for a questionable C6-7 fracture with no associated neck pain after a motor-vehicle accident. She was neurologically intact prior to surgery and upon awakening from anesthesia. Several hours later, she developed respiratory distress requiring emergency reintubation. Following this event, the patient was found to be quadriplegic. X-ray films showed dislocation of a C6-7 fracture. Emergency decompressive laminectomy and fusion were performed without resolution of her deficit.

Other Problems

One patient suffered displacement of a Harrington rod prior to bone fusion necessitating repeat surgery. Another patient developed a deep posterior wound infection requiring debridement and prolonged halo-vest immobilization, followed by successful anterior fusion once the infection had cleared.

Discussion

Natural History

The natural history of ankylosing spondylitis is benign in most cases. In a study of patients followed over 35 years, 92% remained functionally active while 68% had lifelong back problems; in 41%, there was some degree of spinal immobility. This study also suggests that the course of the disease can be predicted by the pattern during the first 10 years after initial diagnosis. Early rapidly progressive symptoms heralded a poorer prognosis.

In general, patients at greatest risk for spinal column injury are those with total spinal involvement. As the spine approaches total fusion, lifelong back and neck pain may be replaced with stiffness and immobility of the spinal column. This immobility may give rise to vertebral osteoporosis. In this group of patients, the new appearance or exacerbation of previous back or neck pain warrants an exhaustive search for a fracture. Fractures may be multiple as was observed in two patients in our series, and are most commonly associated with hyperextension injuries. Total spine imaging must be performed and should not be limited.
to the symptomatic region.19 Three fractures were missed in our series at initial presentation due to inadequate imaging.

Acute Fractures

Chiropractic manipulation,27 cardiopulmonary resuscitation,13 intubation,12 patient transfer,26 and improperly applied traction devices for reduction of fixed cervical spine flexion deformity12,23,25 have all been implicated as causes for acute traumatic fractures of the spine in patients with ankylosing spondylitis. One patient in our series suffered displacement of a missed L-1 fracture after being moved from his bed to a gurney, with resulting paraplegia. Three other patients in our series became acutely quadriplegic following emergency endotracheal intubation.

When a spinal fracture occurs, it may be extremely unstable with an associated high risk of neurological deficit. Patients with ankylosing spondylitis have a risk of sustaining a cervical spine fracture 3 to 10 times greater than in the general population.12,23,25 and the risk for neurological deficit has been reported in up to 57% of patients with ankylosing spondylitis which, is 3 times more frequent than that for patients without this condition.12,16,23,24,34,37,38 Neurological deficit may occur acutely or be delayed due to spinal instability.3 The lower cervical spine is at greatest risk for acute injury.12,16,17,19,23,37,38 Seven of our nine patients with acute cervical spine fracture had neurological deficits prior to surgical intervention: five of these patients had partial resolution postoperatively.

Epidual hematomas have been observed in some patients with traumatic fractures.14,17 Detwiller, et al.,12 reported an epidural hematoma in one of 11 patients with acute cervical spine injury. Farhat, et al.,13 proposed that cancellous bone in patients with ankylosing spondylitis may be more likely to bleed following a fracture, resulting in an epidural hematoma. No patients in our series were found to have an epidural hematoma, nor was excessive bone bleeding noted at the time of surgery.

Controversy exists as to whether patients with acute fracture/dislocation of the cervical spine should be managed nonsurgically with halo-vest immobilization or by surgical fusion. Grisolia, et al.,18 and Weinstein, et al.,26 advocated conservative therapy for patients without neurological deficit, persistent vertebral dislocation, or fracture fragments within the spinal canal, based on the high morbidity rate associated with surgery in ankylosing spondylitis patients. Detwiller, et al.,12 suggested that surgical fusion with postoperative external immobilization produced a satisfactory outcome and allowed earlier patient mobilization, thereby reducing the risk of pulmonary complications common in these patients. In addition, they noted that halo-vest immobilization alone is not without complications.25,40 Fractures commonly involve both anterior and posterior bone elements and ossified ligamentous structures, which may lead to such severe instability that halo-vest immobilization will be inadequate. Others believe that healing of spinal fractures will occur without surgical fusion if external immobilization can be adequately maintained.16,17,39 Fusion failure is generally related to inadequate immobilization in cases treated both surgically and nonsurgically.1

Following realignment or reduction of cervical fractures, we advocate surgical stabilization, most commonly via a posterior approach, with postoperative halo immobilization until fusion can be radiologically documented. We believe that external immobilization alone is inadequate for these patients. In the cervical region when anterior dural deformity is present with neurological deficit, anterior corpectomy straddling the fracture site with strut grafting is performed initially, followed by posterior spinous-process wiring with onlay laminar grafting. Surgical decompression is warranted for patients with progressive neurological deficit associated with radiologically defined spinal deformity or subluxation.16,17,37 Early mobilization in a halo vest is mandatory to minimize respiratory complications inherent with prolonged bed rest.

Pseudoarthroses

Pseudoarthroses or stress fractures are most common in the low thoracic spine and are situated at the level of the intervertebral disc space. These are believed to be true pathological fractures rather than inflammatory erosions of the disc and adjacent endplates.3,18 This finding suggests that the ossified disc space is the weakest area of the fused spinal column and thus is most prone to fracture.3,39 Patients generally present with pain and can be managed conservatively if the posterior elements are intact and no neurological deficit is present.3,22,41 External immobilization with custom-fitted thoracolumbar spinal orthoses may result in eventual fusion and pain relief, although no patient in our series experienced successful spontaneous healing in this manner. Persistent pain, continued pseudoarthrosis, and progressive subluxation are all indications for Harrington rod fixation or spinous-process wiring with onlay bone grafting. Decompressive procedures are performed only for patients with progressive neurological deficit. Anterior fusion alone or in combination with posterior fusion is used when severe instability exists based on anterior and posterior element disruption. For cases approached anteriorly, the pseudoarthrosis is excised and bone-tissue grafting is used to fill the defect. Thoracolumbar orthoses are worn for 3 to 6 months postoperatively until fusion can be documented. All of our patients achieved satisfactory pain relief and fusion of their pseudoarthroses.

Other Spinal Disorders

Spinal Deformity. Patients with progressive spinal deformity should be evaluated by a surgeon versed in spinal stabilization for consideration of wedge osteotomy and stabilization.31,32 The usual indication for this corrective surgery is progressive spinal kyphosis resulting in an inability to see the ground ahead while walking, making the patient more prone to falls or other injuries.37 Most commonly a dorsal wedge osteotomy followed by Harrington rod instrumentation and bone grafting is performed.

M. W. Fox, B. M. Onofrio, and J. E. Kilgore
Neurological complications of ankylosing spondylitis

Rotary Instability. Patients with nontraumatic atlanto-occipital or atlantoaxial subluxation should be managed with posterior fusion. Spinal fusion in patients with ankylosing spondylitis begins in the lumbar spine and ascends. As the enthesopathic process reaches the cervical spine, upper cervical spine instability may occur because these are the only remaining movable spinal joints. In addition, chronic synovitis can cause laxity or rupture of the transverse ligament. Basilar invagination can also occur. Sharp and Purser reported neurological deficits in seven of 22 patients with C1–2 subluxation. It is unknown whether these subluxations will heal with external immobilization alone. All of our patients with atlanto-occipital or atlantoaxial subluxation were neurologically intact and underwent posterior fusion for severe occipital pain and instability. Each was maintained in a halo vest for at least 3 months until stability was obtained.

Spinal Stenosis. Patients with symptomatic spinal stenosis are candidates for decompressive laminectomy. Spinal stenosis may arise secondary to the proliferative processes of ankylosing spondylitis with subsequent canal narrowing or may simply be coincidental. All patients with ankylosing spondylitis should be suspected of having some component of spinal stenosis, and caution should be used when performing laminectomies. In our series, decompressive laminectomies were performed in three patients with cervical myelopathy and one patient with pseudoclaudication. All patients had partial resolution of their symptoms.

Operative Difficulties

Spine surgery in patients with ankylosing spondylitis is more difficult due to the fragility of the spinal column. Extreme care must be taken when moving or lifting patients or during positioning for surgery to maintain fixed spinal kyphosis and to prevent fracture or dislocation, as was observed in one patient in our study. Halo-vest application can be hazardous, as demonstrated in one of our patients. Endotracheal intubations must be skillfully performed without excessive neck extension to avoid cervical fracture, which was observed in three of our patients.

The inability to flex or extend the rigid spinal column can make surgical exposure more difficult. The dense ossification of the supporting ligaments and joints, including interlaminar and yellow ligaments, may make access to the spinal canal both difficult and dangerous. Additionally, unsuspected spinal stenosis may lead to an increased likelihood of neurological compromise following surgery. Advanced osteoporosis may limit the use of strut grafts or stabilization devices.

We had one patient with multiple arachnoid diverticula resulting in extremely thin dura and lamina in the thoracic region. Although cerebrospinal fluid leakage was avoided, a dural onlay graft straddling the involved lamina was used to decrease the risk of undetected leaks. Arachnoid diverticula in patients with ankylosing spondylitis has been observed previously. The cysts are generally asymptomatic, as in our case, but may cause neurological conditions including a cauda equina syndrome. Erosion through involved laminae or into the interlaminar space can be an additional hazard at surgery.

Pulmonary Complications

The pulmonary system is at great risk in patients with advanced disease due to fibrotic upper lung fields, ankylosed ribs, and exaggerated thoracic kyphosis, resulting in a fixed rib cage restricting pulmonary and diaphragmatic expansion. Mortality rates from 35% to 50% have been reported in elderly patients with ankylosing spondylitis who suffer complete cervical cord injuries due to pulmonary complications. In our series, two patients died from respiratory complications and one patient developed severe postoperative pneumonia. These data emphasize the necessity of early postoperative mobilization to help prevent these complications.

Conclusions

This study of 33 patients with neurological complications of ankylosing spondylitis has shown the diversity of spinal disorders seen associated with this disease. Aggressive radiological evaluation, cautious endotracheal intubation and halo-vest application, and early surgical stabilization procedures with postoperative external orthoses are all important to prevent additional spinal cord injury. Decompressive surgery is indicated for those patients with progressive neurological deficit. Although surgery in these complex cases can be more difficult, we have demonstrated that solid fusion can be achieved in these patients, indicating intact bone reparative processes. Early mobilization is mandatory to reduce the risk of pulmonary complications common to these patients and to those with prolonged bed rest. The early involvement of a spinal rehabilitation team is essential. We conclude that outcome in these patients can be favorable if early diagnosis and appropriate management principles are achieved.

Addendum

All operative stabilization procedures described in the text were performed by members of the Mayo Clinic Orthopedic Department.

References


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