Atlantoaxial Subluxation in a Patient with Ankylosing Spondylitis Who Presented with Progressive Quadriplegia After Minor Trauma

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Atlantoaxial subluxation due to minor trauma manifesting with progressive quadriplegia is rare. We present a case in a 38-year-old man who had ankylosing spondylitis for 10 years. The patient initially had right arm weakness, which progressed to bilateral upper and lower extremity weakness within one month. Ten days prior to referral to the emergency department (ED), he fell and was totally bed-ridden with quadriplegia. Two-dimensional computed tomographic (CT) scan of the C-spine performed in the ED demonstrated atlantoaxial subluxation. Conservative supportive treatment was suggested and he was discharged in stable condition 46 days later.

Key words: ankylosing spondylitis, quadriplegia, atlantoaxial subluxation

Introduction

Ankylosing spondylitis (AS) is a group of immune-mediated disorders characterized by chronic inflammation of the axial skeleton that may progress to a rigid, osteoporotic spine. Neck pain and stiffness is one of the characteristics of this advanced disease. It usually begins in the second decade, and is more common in men than women. Patients with advanced AS are susceptible to fracture after trauma, and the fracture tends to involve the lower cervical vertebral area. The most serious complication encountered in AS is spinal fracture, dislocation and atlantoaxial subluxation (AAS). However, AAS is considered an uncommon feature of AS without rheumatoid arthritis. AAS may be symptomless, but it may cause severe pain or neurological symptoms.

Herein, we report a case of AS with minor trauma presenting with progressive quadriplegia after one month.

Case Report

A 38-year-old man was referred to our emergency department (ED) because of paresthesia in the limbs and trunk numbness. He had lower back pain for 10 years without any prior trauma. Ankylosing spondylitis was diagnosed by radiography (squaring of the lumbar vertebrae and bony erosion and sclerotic changes in both sacroiliac joints) and a positive result on a human leucocyte antigen (HLA)-B27 test. He was regularly followed in our orthopedic clinic and had been managed with an oral nonsteroidal-anti-inflammatory drug (diclofenac, 25 mg three times...
daily) and an anti-rheumatic drug (sulfasalazine, 500 mg four times daily). However, he had neck stiffness and neck pain over the past year. The neck pain was mainly related to motion but sometimes was also present at rest. Therefore he was treated with acupuncture and the pain and stiffness subsided. One month before he was referred to our ED, he felt right arm weakness followed by progressive weakness of the bilateral upper and lower extremities. He could not walk without assistance and complained of falling easily. Ten days prior to this ED visit, he fell and was totally bed-ridden because of quadriplegia. Difficulty in urination and urine incontinence was noted several days later. Two days before this ED visit, he began to feel shortness of breath, dyspnea and abdominal fullness.

Physical examination showed muscle atrophy, and generalized hyperreflexia in his bilateral upper arms and lower legs. The numbness was most prominent in the distal parts of both upper and lower extremities in a stocking-glove distribution. In addition, the bilateral limbs were flaccid and proprioceptive sensation was poor even though pain and temperature sensations were preserved. Manual muscle-testing revealed generalized muscle weakness (grade 1 of 5) throughout the upper and lower extremities. Spinal cord injuries were thus suspected. In the ED, his body temperature was 36.5°C, blood pressure 131/71 mmHg and the heart rate 134 beats/min. Laboratory tests revealed the following results: leukocytes 17,100/mL, blood urea nitrogen (BUN) 79 mg/dl, creatinine (Cr) 7.0 mg/dl, and potassium 7.7 mEq/L. The urinalysis showed numerous white cells. Acute renal failure, with urinary tract infection and hyperkalemia was diagnosed. Lateral radiographs of the cervical (C)-spine (Fig. 1) and tharaco-lumbar (T-L)-spine (Fig. 2) showed a bamboo spine. Two-dimensional computed tomographic (CT) scan of the C-spine demonstrated atlantoaxial subluxation (AAS) (Fig. 3). Because of the progression of impending respiratory failure, nasal endotracheal intubation was performed in the ED. The patient was then transferred to the surgical intensive care unit (SICU) after intubation. Hydration and empirical antibiotics (ciprofloxacin, 400 mg two times daily) were administrated in the ED. Three days after admission, the antibiotics were changed to vancomycin (500 mg four times daily) in the SICU because of bacteremia (Methicillin-resistant Staphylococcus aureus in the blood culture) and sepsis (leukocytes 20,900/mL). Two weeks later, the sepsis was controlled, and renal function returned to the normal range (BUN 8 mg/dl, Cr 0.8 mg/dl) after hydration. A tracheostomy was performed because of respiratory failure and a cystostomy was performed for neurogenic bladder. Conservative supportive treatment for AAS was suggested because of severe kyphoscoliosis and long-term neurologic impairment in this patient. The patient’s condition was stable and he was discharged 46 days later. He was advised to receive regular follow up in the neurosurgery clinic.

**Discussion**

Spinal cord injuries are not uncommon in the ED especially in patients suffering traumatic injuries such as motor vehicle accidents, falls, gunshot wounds, and sports injuries. Clinical presentation with limb weakness, neck pain, urinary incontinence and dyspnea can be seen in patients with spinal cord injuries. Neurological deficits worsen quickly after injury. A detailed evaluation should be performed without delay, especially in patients with multiple trauma. Patients with ankylosing spondylitis are more predisposed to spinal trauma and cord injury than the healthy population\(^{(5,6)}\). Injuries to the lower cervical spine (C5-C7) occur frequently in ankylosing spondylitis, whereas, fracture of the upper cervical
spine in such patients is rare\(^{(3,4)}\). Our patient had ankylosing spondylitis for 10 years and suffered from progressive quadriplegia due to atlantoaxial subluxation. The diagnosis of atlantoaxial subluxation may be delayed because when trauma is minor. The physician should be alert to the patient with ankylosing spondylitis and a detailed neurological examination should be performed to rule out spinal cord injuries even in cases of minor trauma.

Spinal cord injury is a true emergency in the emergency department. A neck collar should be applied in patients suspected to have cervical spine injury to prevent further injury. In our case,
a neck collar was used by the ambulance crew and the cervical spine was immobilized in the neutral position when the patient was transferred to our ED. The neck collar was removed by the family because of patient discomfort. After relief from the neck collar, the pain lessened. In our case, neck deviation to the right side had already been found one year previously. Changes to the normal position by the hard collar were equivocal because of this procedure which may have increased the neck pain and induced further spinal cord injury. Papadopoulos et al\cite{7} reported a case of AS in which a hard cervical collar was used to normalize a fixed flexion deformity of the neck. Unfortunately, the hard cervical collar increased the neck pain and caused paraesthesia, followed by quadriplegia, shortness of breath, hypotension and bradycardia. They suggested immobilization in a flexed position by placing sandbags under the occiput which may reduce cervical fracture and prevent further cord damage\cite{7}.

ankylosing spondylitis is a complex and potentially debilitating disease that is insidious from the onset, and progresses to radiological sacroilitis over several years\cite{1,2}. Early diagnosis is not easy. The initial symptom is pain generally felt deep in the buttock or in the lower lumbar regions\cite{1,2}. Even minor trauma to the rigid, fragile spinal column can cause severe damage\cite{1,3}. The mechanisms explaining AAS include transverse ligament damage by a periodontoidal proliferative pannus or sequela of ossification of the anterior and posterior longitudinal ligaments, associated inflammatory lesions (cervical spine osteoarthritis, atlantodental synovitis, erosions of the dens and adjacent ligaments), and physical stress (kyphosis of the dorsal spine and weight of the head at the C1-C2 level)\cite{2,8-10}. Osteoporosis is another possible mechanism of AAS since reduced mineral content may result in more fragile bones. Patients often have a history of a hyperextension injury and prior alcohol use.

There are no established guidelines for the management of AAS in patients with AS. Ramos-Remus et al concluded that surgical stabilization should be recommended when displacement between the anterior aspect of the odontoid and the posterior aspect of the anterior arch of the atlas is
greater than 5mm in the lateral maximal flexion view\(^{(11)}\). Also, severe pain cannot be controlled by a collar if displacement of the sagittal diameter of the spinal canal is 30% or greater or there are neurologic symptoms or signs. In our case, because of severe kyphoscoliosis and long-term neurologic impairment, the patient received rehabilitation and conservative treatment only.

**Conclusion**

In conclusion, the risk of spinal cord injury in AS is higher than in the healthy population. Careful neurologic and radiographic evaluation of patients with this disease should be performed to monitor possible myelopathy. A detailed examination of the entire spine should be performed to rule out spinal cord injuries in all patients with underlying disease of the spine even if only minor trauma is noted. The present report illustrates the importance of appropriate neurologic and radiographic evaluation of patients with AS to help prevent severe neurological complications.

**References**

僵直性脊椎炎因轻微外伤导致寰椎与枢椎的半脱位
以渐进性的四肢麻痹来表现：一病例报告

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脊椎损伤在急诊部门并非罕见的急症，因为轻微外伤导致寰椎与枢椎的半脱位而以渐进性的四肢麻痹来表现则相当少见。我们报告一位38岁僵直性脊椎炎的男性患者，一開始感觉右手臂无力，在一个月后无力的情形恶化且蔓延至四肢。在来我科的急诊室的前10天，病患因无力而跌倒，造成四肢麻痹及卧床在家。因为四肢麻痹，颈部疼痛及尿失禁被转诊至我们的急诊，颈椎的2-D电脑断层显示寰椎与枢椎的半脱位。经过保守及支持性的治疗后，病患状况稳定而於46天后出院。

关键词：僵直性脊椎炎，四肢麻痹，寰椎与枢椎

収件：98年6月3日  接受刊载：98年8月24日
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