Spinal Cord (2000) 38, 327–329

www.nature.com/sc

Case Report

Slow ascending myelopathy, tetraplegia, carcinoma of the bladder and amyloidosis in a patient with ankylosing spondylitis

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Objective: We report a case of slow ascending myelopathy in a patient with ankylosing spondylitis (AS).
Design: Case report of a 60-year-old patient suffering from AS, who developed over a period of 39 years a slow ascending myelopathy leading to tetraplegia, squamous cell carcinoma of the bladder and amyloidosis of the small intestine secondary to neuropathic bladder and bowel.
Setting: Department and Outpatient's Department of Neurological Rehabilitation Sheba Medical Center, Tel Hashomer, Israel.
Subject: Single patient case report.
Main outcome measure: Clinical follow-up of the patient between the years 1959–1998.
Results: Physical examination disclosed deteriorating incomplete tetraplegia with hypotonia and hyporreflexia. Neurogenic bladder and bowel complicated to squamous cell carcinoma and amyloidosis.
Conclusion: To our knowledge, flaccid tetraplegia associated with AS, has never been reported in the literature. The possibility of vascular compression by the ankylosed spine causing the clinical picture of flaccid tetraplegia in this patient is discussed.
Spinal Cord (2000) **38,** 327–329

Keywords: slow ascending myelopathy; tetraplegia; amyloidosis; squamous carcinoma

Introduction

Ankylosing spondylitis (AS), also known as von Bechterew's disease or Marie-Strumpell's disease, is a chronic inflammatory illness that primarily affects the sacro-iliac joints and the spine. It is characterized by pain and stiffness of the back. The primary pathological site of AS, enthesopathy, lies at the insertion of ligaments and capsules into bone. In addition to spinal symptomatology, some patients may exhibit a peripheral arthropathy and regions of insertional tendinitis. Non musculo-skeletal manifestations of the disease include uveitis, pulmonary fibrosis, cardiac valvular disease and amyloidosis.

Neurological complications are rare in AS and consist of solitary nerve root lesions, myelopathy[1,2] associated with spontaneous atlanto-axial subluxation[3–5] and cauda equina[6] compression after traumatic fractures of the multi-level fused vertebral column.[7–9] Some reports[10,11] suggest an association with multiple sclerosis. In the following case report we describe a patient suffering from AS, who developed progressive spinal cord damage over 39 years. The clinical neurological picture of flaccid tetraplegia in our case strongly suggests, that damage to the entire spinal cord occurred most probably due to vascular compression by the ankylosed spine.

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Case report

S.Y., male, born in Poland in 1938, was first admitted to our center in 1959 when he complained of lower limb spasm occurring mainly at rest. The neurological picture developed to include permanent leg pain, both during exercise and at rest, mild sensory loss and some sphincter disturbances. He was diagnosed as having an incomplete cauda equina syndrome of unknown etiology. All laboratory investigations at that time were normal. Lumbar puncture revealed opening pressure of 18 cm/water with normal fluid analysis. Spinal column X-rays showed some flattening of lumbar lordosis. Myelography was reported as showing 'a defect in the dural sac at the D_{12} level

and some changes compatible with an arterio-venous malformation of the spinal cord' but review of these films has not confirmed this. The etiology remained uncertain, with the differential diagnosis including Guillain-Barre's syndrome or arterio-venous malformation. No pathological ocular signs were noted. Metabolic or chemical causes (heavy metals, porphyria, etc.) of any spinal cord disease were eliminated. No treatment was suggested.

In 1961 further neurological deterioration was observed, with flaccid paraplegia below the level of D_7. In 1962 the level of paralysis was noted at D_8, with a lower motor neuron neuropathic bladder. The patient could ambulate with crutches and long leg braces.

In 1964 he was hospitalized in our rehabilitation center for walking training, but was discharged in a wheelchair. In 1968 and 1969 he underwent surgical closure of decubitus ulcerations. In 1971 he suffered from sudden acute pleuro-carditis of suspected viral etiology. Tuberculosis and bacterial infection were excluded. After traumatic insertion of a Foley catheter during this illness he developed a urethral diverticulum. Surgical correction attempts failed and a permanent supra-pubic catheter was inserted. His renal function was normal after that.

It was only in 1972 that the first spondylytic changes in the spine and the disappearance of the sacro-iliac joints were noted. In 1977 the neurological status was flaccid quadriplegia below the C_7-C_8 level, with a rigid spine and only a 30° rotation of the neck. HLA-B27 was positive at that time. In 1979 the sensory disturbance ascended to the C_3 level, with the motor level remaining constant.

A CT scan of the entire spine performed in 1982 revealed severe osteoporosis, posterior ligament ossification, facet hypertrophy, but no sign of any vascular lesion. At that time MRI studies were not available at our hospital. The classical picture of a bamboo spine was found involving the thoraco-lumbar spine. The patient was only partially independent in activities of daily living, moving around in a powered wheelchair.

He developed severe complications in 1994: osteomyelitis of the left trochanter, perineal urinary fistulae, hypertension, sacral decubiti ulcers and sepsis. During a long hospitalization, his decubiti were surgically closed and he received prolonged antibiotic treatments.

In February 1997 he developed gall bladder empyema, which necessitated laparoscopic cholecystectomy. In December of the same year he had recurrent urinary tract infections with renal failure. In January 1998 a massive bleeding occurred around the cystectomy and renal function further deteriorated. A month later he was readmitted in order to perform a radical cystectomy with an ilial conduit. Squamous cell carcinoma of the urinary bladder and amyloid deposits of the small bowel were found. The post-operative course included respiratory insufficiency, sepsis and cardiac arrest, which led to his death. Prior to his last hospitalization he was found to suffer from complete tetraplegia below C_6.

Discussion

Our patient showed a very slow ascending neurological deterioration, progressing over 39 years. AS was diagnosed 13 years after the first symptoms occurred. A search of the available literature has not yielded any description of slowly ascending myelopathy with eventual tetraplegia associated with AS. Neurological symptoms first appeared as a cauda equina syndrome (CES). This syndrome, first described by Bowie and Glasgow in patients with AS, is a rare complication of long standing disease (usually more than 15 years), when the disease is inactive. Myelography revealed defects in the dural sac. The typical myelographic image in CES with AS shows a voluminous sac and a dorsal arachnoid diverticula. The cause of CES is still unknown, but it was observed that spinal arachnoiditis results in damage of nerve roots. The possibility of a concomitant arterio-venous malformation of the spinal cord was eliminated by a negative CT scan searching for such a vascular malformation.

At the time when the diagnosis of AS was reached, clinically based on a rigid spine with loss of lumbar lordosis and radiographic signs of sclerosis of the sacro-iliac joints and ankylosis of the lumbar spine, the patient had flaccid paraplegia.[12] It is well known that paraplegics may develop bilateral blurring of the sacro-iliac joints, bone sclerosis and spinal column changes very similar to those seen in patients with AS.[13] The additional association with the HLA-B27 gene and severe pleuro-pericarditis in our patient make the diagnosis of AS very likely.

A decade later the clinical picture developed towards flaccid tetraplegia, stopping at the neurological motor level C_6 and sensory C_3.[14]

Eventually the patient developed bladder carcinoma, small bowel amyloidosis and renal failure.

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