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Case report

Cervical instability in a patient with rheumatoid arthritis

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Abstract

Introduction: Rheumatoid arthritis (RA) is a chronic autoimmune connective tissue disease characterized by symmetrical arthritis associated with extra-articular changes. Although peripheral joint involvement is the dominant symptom of RA, many patients develop cervical spine involvement in the course of the disease, manifesting as cervical instability.

Aim: The aim of this study is to describe a case of an RA patient with spinal myelopathy to increase awareness of this complication, hoping that its early diagnosis may prevent further serious consequences.

Case study: A 63-year-old patient, who was diagnosed with RA 18 years ago, was admitted to the Rheumatology Clinic due to suspected exacerbation of rheumatic disease. Functional X-Ray and MRI was performed, which showed instability in the C3–C4 segment with spinal cord compression. Subsequently, the patient underwent cervical spine surgery. After the surgery and rehabilitation, the patient demonstrated neurological improvement.

Results and discussion: Every patient diagnosed with RA should be educated about the possibility of a complication of cervical instability and be familiar with the neurological symptoms that may result from it. If cervical instability and subsequent cervical myelopathy are detected early, the symptoms may be reversible or significantly reduced by surgical spinal cord decompression and cervical stabilization.

Conclusions: It is very important to perform a functional X-ray of the cervical spine to exclude instability, especially before rehabilitation treatment. Neck pain in patients diagnosed with RA may indicate cervical instability that requires more thorough neurological examination to exclude cervical myelopathy.

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1. INTRODUCTION

Rheumatoid arthritis (RA) is a chronic autoimmune connective tissue disease characterized by symmetrical arthritis associated with extra-articular changes and systemic complications. It is the most common inflammatory rheumatic disease and affects 1% of the adult population.[1] Up to 86% of patients develop cervical spine involvement in the course of the disease, manifesting as cervical instability.[2] Cervical spine involvement is a consequence of intense chronic synovitis occurring in the joints, leading to bone erosion and subsequent sagging ligaments, and finally clinical and radiological instability.[2] Cervical instability, that can lead to cervical myelopathy, may manifest itself as neck pain, occipital headache, limited range of motion in the cervical spine, numbness in hands, weak hand grip, impaired fine motor skills, weakening of muscle strength in upper and lower limbs, sensory disturbance or gait disturbance. The spinal cord is then flattened in the anteroposterior dimension, which can lead to cell, nerve and spinal cord tracts damage.

It is important to educate patients and doctors about the symptoms of neck instability. Given that cervical instability does not always manifest with neurological symptoms, screening functional radiographs of the cervical spine should be taken after the diagnosis and before rehabilitation to avoid subsequent complications.

2. AIM

The purpose of this report is to describe a case of patient with RA with spinal myelopathy to increase awareness of this complication among both doctors and patients, hoping that its early diagnosis may prevent further serious consequences.

3. CASE STUDY

A 63-year-old female nurse with a BMI of 25, who was diagnosed with RA 18 years ago, was admitted to the Rheumatology Clinic due to suspected exacerbation of rheumatic disease. According to the patient, she was bedridden for 4 months. She couldn't transport herself and was looked after by her son with alcohol problems. For 7 months before admission to hospital, she had been experiencing pain and progressive weakness in her legs, wrists, arms, shoulders and neck, with pain radiating to the upper limbs, as well as reduced handgrip strength. She had no need of any assistive devices before the collapse in physical function.

Upon admission to the Rheumatology Clinic, the patient's primary complaint was pain in the cervical and lumbar spine, as well as her shoulders and wrists. In addition, she had a severe spastic paresis of both lower limbs, expressed more distally and on the right rather than the left side. The spasticity in the lower limbs was measured at Grade 3 according to the modified Ashworth scale (MAS) ranging from 0 to 4. The muscle strength of knee extensors and dorsal flexors of both feet were rated 2 on the Medical Research Council (MRC) scale (rated from Grade 0 to 5, denoting complete paralysis and full strength correspondingly). The muscle strength of the plantar flexors of the left foot was rated 2 on the MRC scale, the right foot was rated 1. The strenght of the rest major muscle group is listed in Table 1. The sensibility in lower extremities was diminished with both superficial light touch and deep sensation disorder, more pronounced on the right leg. Moreover, the presence of pathological reflexes was detected with Hoffman's sign and inverted radial reflex. The response of biceps, triceps and brachioradialis was symmetrically strong. Babinski's sign was not observed.

Other concurrent illnesses were hypertension, stable coronary artery disease, and type 2 diabetes treated with insulin since 2007, methylprednisolonum, bisoprolol, spironolakton, acetylsalicylic acid.

Table 1. Strength of the major muscle group.

	Pre-surgery	Post-surgery	At rehab center discharge
Upper extremity			
Elbow flexors			
Right	4	4	5
Left	4	4	5
Wrist extensors			
Right	4	5	5
Left	4	4	5
Elbow extensors			
Right	3	4	4
Left	3	4	4
Finger flexors			
Right	3	4	5
Left	3	3	5
Finger abductors			
Right	3	4	4
Left	3	4	4
Lower extremity			
Hip flexors			
Right	3	4	5
Left	3	4	5
Knee extensors			
Right	2	4	2
Left	2	4	5
Ankle dorsiflexors			
Right	2	4	5
Left	2	4	5
Long toe extensors			
Right	1	4	5
Left	2	4	5
Ankle plantar flexors			
Right	2	2	4
Left	2	3	5

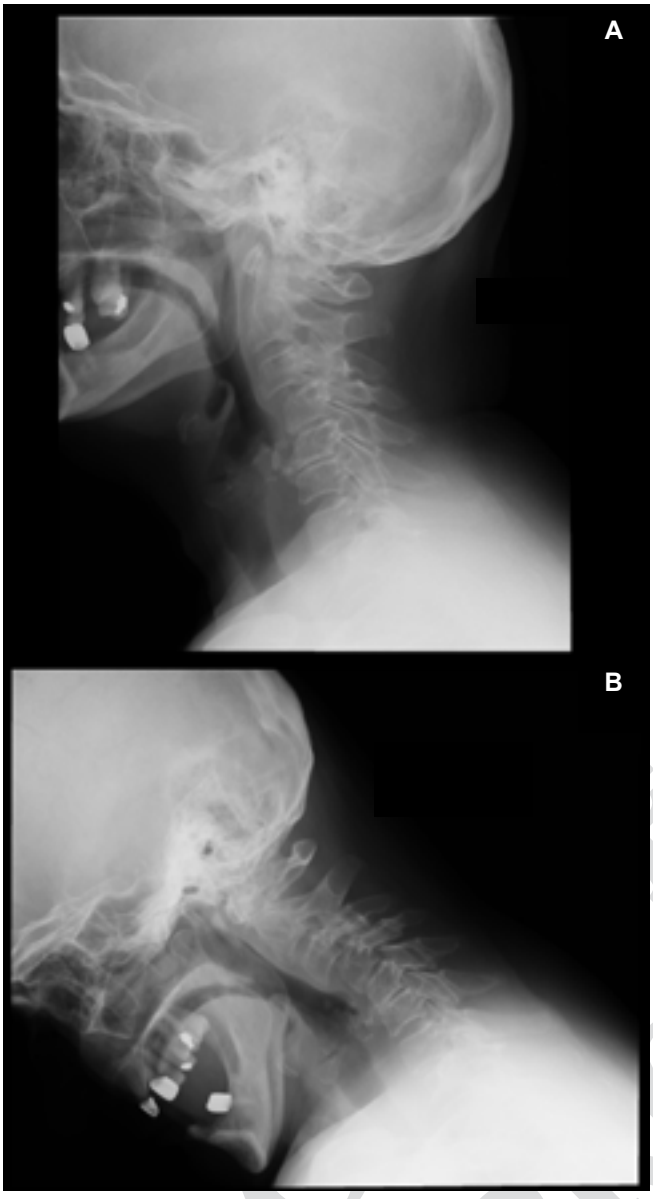

Figure 1. X-radiograph of the cervical segment before cervical spine stabilization surgery: neutral position (A), flexion (B). Date of examination: November 19, 2018.

Figure 2. MRI of the cervical spine showed the spinal cord compression at C3–C4. At the level of C3–C5, the spinal cord signal intensity was slightly changed, as is the case with myelopathic changes visible myelopathy at C3–C4 which is more visible on figure B. Date of examination: November 28, 2018.

Prior to admission to the hospital, the patient had no imaging performed. Later, due to spastic paresis, functional X-ray (Figure 1) and MRI of the cervical spine (Figure 2) were performed. The functional X-ray showed multilevel discopathic changes in the C3–C4, C4–C5, C5–C6 cervical segment, instability in the C3–C4 segment with 7.5 mm anterior horizontal displacement of C3 in relation to C4 vertebra at the flexion position. MRI showed the spinal cord compression at C3–C4. At the level of C3–C5, the spinal cord signal was slightly changed, as is the case with myelopathic changes.

Given the history and physical examination findings and concern over progressive cervical myelopathy the patient underwent a cervical spine surgery over the first three weeks after diagnosis. The stabilization operation involved a C3–C4–C4–C5 discectomy, dislocation reposition and stabilization with interbody graft at C3–C4, intervertebral disc implants at C4–C5 and C5–C6, and anterior cervical plate from C3 to C6 (Figure 3). It was performed within a week of the neuro-orthopedic consultation. A day after surgery, the patient demonstrated a reduction of pain both in the cervical spine and upper limbs. In the clinical study, there was a slight improvement in the muscle strength in all major muscle groups of the upper and lower extremities observed (Table 1), which indicated correct decompression of the spinal canal. The response of biceps, triceps and brachioradialis was symmetrically strong, the reflexes from the patellar tendon were symmetrical and normal. The Achilles tendon

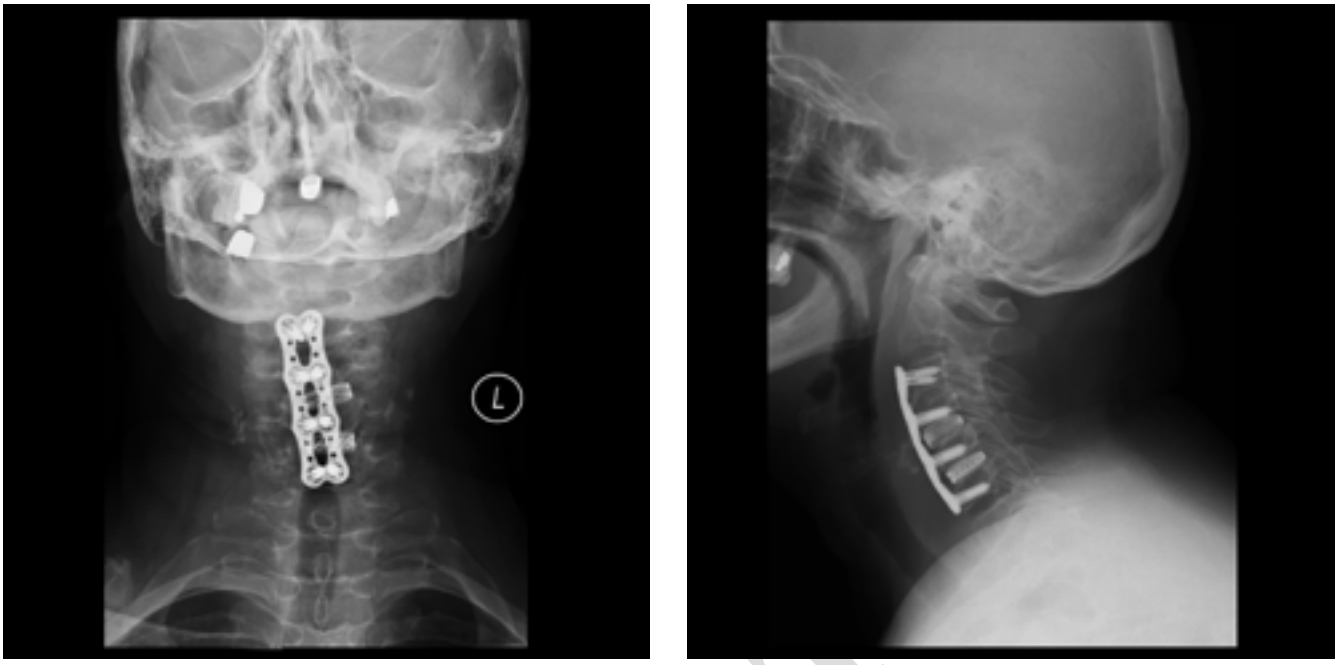

Figure 3. X-radiograph of cervical segment post-surgery: sitting position, AP position (A), lateral position (B). Date of examination: December 27, 2018.

response seemed weakened, and the plantar reflex was normal. Babinski's sign was not observed.

The patient was admitted directly from our Neuroorthopedics and Neurology Clinic in 7 days post-surgery. At the time of admission to the rehabilitation clinic, the patient was non-ambulatory. She could only change positions of her body in bed without being able to sit alone. She had a quadriparesis that was most pronounced in the plantar flexors of the feet, and diminished superficial and proprioceptive sensation, more pronounced on the right.

A 6-week comprehensive rehabilitation was planned, including breathing exercises, isotonic and isometric exercises for the lower limb muscles, verticalization with forearm platform, transfer and ambulation training, closed kinetic chain upper extremity and shoulder girdle exercises, and occupational therapy (with the focus on daily routine actions). During the stay, the patient's functional state progressively improved: her muscle strength increased, she could move around with a walker and change the position of the body freely.

After 6 weeks of rehabilitation, the patient was able to walk on her own using a forearm platform rolling walker for a distance of 60 m. We observed muscle strength improvement in major upper limb muscle groups, primarily in elbow flexors, left limb wrist extensors and fingers flexors. As for the lower limb muscle strength, there was a significant improvement in hip flexors, dorsal and plantar feet flexors and long extensors of the toes (Table 1).

4. RESULTS AND DISCUSSION

According to some authors,[3] 30%–69% of patients with RA have limited mobility in the cervical spine, which is why it is so difficult to examine a patient with RA for cervical instability. Due to joint deformities in the course of the disease, it might be difficult to interpret a neurological examination (for example, in the case of hallux rigidus, despite the damage to the pyramidal tract, the Babinski reflex cannot be evoked). The most common cervical myelopathy symptoms include neck pain, gait disturbance, weakness and instability of the lower limbs, reduced hand function precision and hand numbness. Here, we describe a case of a patient with neurological symptoms of cervical myelopathy developed in the course of cervical spine instability. Unaware of these possible complications, she associated her symptoms with RA, i.e. her underlying disease. Prior to admission to the hospital, the patient observed increasing pain and weakness in the lower limbs. Her major concern was weakness in the upper limbs and weak hand grip. The neurological deterioration was gradual, which is typical in most cervical myelopathy patients.

Initial cervical myelopathy symptoms are usually subtle and, in 75% of patients, include dexterity reduction that leads to disruption of precise movement activities, such as writing and buttoning.[4] These disorders were also our patient's first myelopathy symptoms noticed about 4 months prior to surgery. However, she thought they were her underlying disease manifestations. RA and myelopathy patients gradually develop upper limb weakness. They often complain of numbness in their hands and forearms (which was also the case with our patient). Muscle atrophy is observed, too. There are also characteristic pathological symptoms such as inverted radial reflex, Hoffman's sign and increased tendon reflexes in the limbs (also observed in our patient). All these symptoms are indicative of pyramidal tract damage and, when found during a clinical examination, should hint at myelopathy. The characteristic symptoms of myelopathy in the upper limbs also include:

(1) the finger escape sign that is tested by asking patients to hold their fingers extended and adducted for 30 s (if the ulnar digits drift into abduction and flexion, the result is positive),
(2) grip and release test that consists in making and releasing a fist at least 20 times within 10 s (positive when the patient is unable to this without impairment and clumsiness), and
(3) hyperactive pectoralis muscle reflex evoked by tapping the pectoralis tendon in the deltopectoral groove (the test is positive if adduction and internal rotation of the shoulder is observed).[5]

The described patient had superficial and deep sensation (proprioceptive) disturbances, which is also more characteristic of myelopathy rather than of RA.

One of the first symptoms of myelopathy is gait disturbance resulting from damage to the posterior spinal column and from paresis. Patients often have an impaired vibration sensation in the lower limbs. Patients with the posterior spinal column damage have a positive Romberg's test. Unfortunately, we cannot say whether the patient had exactly this type of symptoms, because she could not walk on her own upon admission. The majority of myelopathy patients manifestate upper motor neuron symptoms, which are characterized by hyperreflexia, hypertonia and weakness, especially visible in the iliopsoas muscle and quadriceps muscle. Weakness and sensory disturbances lead to characteristic changes in gait, slower gait speed, decreased step length, longer stride time. Those may also be accompanied by Babinski's symptom and pathological clonus. Neurogenic bladder dysfunction is also observed in severe cases of cervical myelopathy.[5] In patients with aforementioned symptoms, other diagnoses such as polyneuropathy should be excluded. In polyneuropathy, apart from muscle strength weakening and gait disturbances, the deep reflex response becomes weaker compared to cervical myelopathy where it is abnormally strong.

In two separate cohort studies, 7%–21% of patients with RA showed signs of subluxation in the cervical spine during the first year after rheumatic disease diagnosis.[2] Although RA is more common in female patients, severe forms of cervical instability are more common in men.[4] There are three types of instability in the cervical spine: atlantoaxial subluxation (AAS), subaxial subluxation (SAS) and cranial settling (CrS). AAS is the most common cervical instability localized at C1–C2 (in approximately 65% of patients). There are three subtypes:anterior-located between the anterior arch of C1; posterior-located between posterior arch of C1 and the dens of axis and lateral asymmetrical or unilateral changes of the lateral atlantoaxial joint leading to impaired rotation. The second most common instability, manifesting itself in 20% of RA patients, is observed at C3 to C7 levels (SAS). The CrS, intussusception of the odontoid of the second cervical vertebra into the foramen magnum, affects about 15% of patients.[6]

In the natural course of RA, the above-mentioned disorders may also occur, which significantly hinders the early detection of cervical myelopathy symptoms. Therefore, every patient diagnosed with RA should be educated about the possibility of a complication of cervical instability and be familiar with the neurological symptoms that may result from it. Doctors who treat RA patients should remember about a detailed neurological examination that may be hard to perform due to severe hand and feet deformities. Given the high incidence of cervical instability in RA patients, proper diagnosis is crucial for early detection and proper therapeutic management.

In all patient with diagnosed RA, it is sufficient to take a functional X-ray of the cervical segment in 4 positions: neutral, flexion and extension, and in the AP position. To determine the C1–C2 instability, it is necessary to measure the distance between the anterior arch of the C1 and the odontoid of the C2 (ADI) that should not exceed 3 mm. Some authors believe that ADI above 5 mm at the flexion position is a sign of clinically significant instability, and figures above 8 mm are an indication for surgical treatment. Radiographs should also measure the distance between the odontoid and posterior arch of C1 (posterior ADI), which assesses the actual width of the spinal canal and should be above 14 mm.[7] A CT or MRI cervical spine examination is indicated if plain radiographs prove the presence of or hint at any cervical spine disease, or in case of severe cervical pain or any neurological manifestations to visualize soft tissues.[8–11] The described patient had subaxial subluxation, C3–C4–C5–C6, with 7.5 mm anterior horizontal displacement of C3 in relation to C4 vertebra, that caused the spinal canal to narrow to 10 mm leading to symptomatic cervical myelopathy. The spinal cord diameter measured on lateral radiographs from C3 to C7 is 14–23 mm. This measurement is a better predictor of neurological impairment (if less than 14 mm) than the degree of subluxation between the vertebrae.[12,13]

If cervical instability and subsequent cervical myelopathy are detected early, the symptoms may be reversible or significantly reduced by surgical spinal cord decompression and cervical spine stabilization.[2]

If cervical myelopathy in imaging studies occurs in the course of cervical instability in RA patients, surgery is the best treatment. Sunahara et al. studied 21 patients with myelopathy in the course of RA who chose not to undergo a surgical treatment and, as a result, showed no neurological improvement. Of these patients, 76% experienced neurological deterioration, and all patients became bedridden within 3 years.[14] Boden et al., on their part, investigated 73 RA patients with cervical spine involvement over an average of 7.1 years; 48% of those patients had operative treatment, and 71% of them had neurological improvement after surgery. It was also noticed that the fewer neurological symptoms before surgery, the quicker the recovery.[15] Most patients, 2–8 weeks after cervical decompression surgery, undergo in-hospital rehabilitation involving physiotherapy and/or occupational therapy. According to studies,[16] the shorter duration (2 weeks) of cervical spine immobilization after cervical stabilization surgery, compared to longer periods (4 and 8 weeks), reduces neck pain and maintains spinal cervical mo-

bility. Besides, short postoperative immobilization does not cause intervertebral instability.

Rehabilitation treatment aims at muscle strengthening improving motor coordination and gait and returning the patient to their activities of daily living.[17] Rehabilitation should be immediate, because the sooner rehabilitation after surgery starts, the less is the likelihood of complications such as pressure sores, spasticity and urinary incontinence. Besides, appropriate selected orthoses or other supportive devices contribute to greater patient independence.[18] Our patient underwent comprehensive rehabilitation immediately after surgery at the Neuroorthopedics and Neurology Clinic. At the time of admission, she was bedridden. Upon discharge, the patient was able to walk on her own using a forearm walker for a distance of about 60 m. We observed muscle strength improvement in major upper limb muscle groups, primarily in elbow flexors, left limb wrist extensors and fingers flexors. As for the lower limb muscle strength, there was a significant improvement in hip flexors, dorsal and plantar feet flexors and long extensors of the toes.

5. CONCLUSIONS

(1) Given that cervical instability can lead to life-threatening complications arising from spinal cord compression, such as cervical myelopathy or sudden death, it is very important to detect and treat this disease early.
(2) Although peripheral joint involvement is the dominant symptom of RA, many patients develop cervical spine involvement in the course of the disease, manifesting as cervical instability.
(3) It is very important to perform a functional X-ray of the cervical spine to exclude instability, especially after diagnosis and before rehabilitation treatment.
(4) Neck pain in patients diagnosed with RA may indicate cervical instability that requires more thorough neurological examination to exclude cervical myelopathy.
(5) The delay in diagnosis and treatment can cause long-term disability, which is why it is so important to educate the patients and doctors involved in treating RA patients about the symptoms of neck instability, especially those that are similar to the manifestations of the underlying disease.
(6) Early diagnosis of cervical instability and effective treatment before the development of irreversible spinal cord damage is crucial to maintaining patients' quality of life.

Conflict of interest
Authors declare none conflict of interest.

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