Quadriplegia due to pachymeningitis, vasculitis and sepsis in a patient with rheumatoid arthritis: a case report

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Abstract. – We report the case of a 84-year-old man, with history of rheumatoid arthritis, admitted the Hospital for a fall and complaining of dysaesthesia and pain located to the cervical spine and arms. Within a few hours after admission, fever and acute, progressive, ascendant quadriplegia became evident. Magnetic resonance imaging (MRI) of cervical spine showed spinal canal stenosis between C4-C6 with spinal cord compression. Hemocultures resulted positive for Staphylococcus aureus. The clinical picture rapidly evolved to sepsis with a fatal multi-organ failure. An autopsy found a osteosclerosis narrowing the neurocanal at the level of C3-C6, and recent cervical medulla infarction. A histological exam revealed the presence of a suppurative pachymeningitis with local phenomenas of periradicularitis, vasculitis and thrombosis of the anterior or medullar artery, associated with coagulative necrosis of the neural tissue.

Key Words: Sepsis, Pachymeningitis, Quadriplegia, Vasculitis, Rheumatoid arthritis.

Introduction

Rheumatoid arthritis (RA) can involve cervical spine as a result of joints, ligaments and bone severe damage secondary to synovitis. Thus, subluxations are often consequent, and concomitant involvement of the basilar-vertebral vascular system is frequent. Patients affected by RA commonly suffer from pain and neurological signs, and cases of sudden death have also been described. Neck pain radiating to occiput is an early and common symptom of cervical spine damage. Two other clinical manifestations can be observed when subluxation is present: (1) slowly progressive spastic tetraparesis, frequently associated with hypoesthesia of the hands, due to spinal cord compression and (2) possible episodes of medullar dysfunction and vertebral artery compression due to the subluxation of the odontoid process.

Pachymeningitis is a rare illness which can be associated with an infectious, malignant, or rheumatic systematic disease. Typical symptoms are chronic headaches, facial pain, cerebella ataxia, cranial nerve failure, neuro-ophthalmic complications.

We present a case of a patient with history of RA admitted for medullar infarction due to sepsis with pachymeningitis and vasculitis.

Case Report

An 84-year-old man was urgently admitted to the Emergency Room at 7 am for a fall (about 5 hrs before) with consequent bruises and grazes to the elbows. He had a history of hypertension, angina, moderate chronic renal failure, and rheumatoid arthritis (for about 40 yrs) which were treated with corticosteroids and non-steroidal anti-inflammatory drugs (NSAIDs). A few weeks before hospital admission the patient began to suffer leg hyposthenia with a tendency to fall. 10 days before being admitted, he fell suffering bruises and grazes to the legs. Two days before, he complained of pain localized to the dorsal spine and arms with dysaesthesia; these symptoms were partially resolved with NSAIDs. He was a smoker (7-10 cigarettes/day) and denied alcohol consumption. At the moment of observation he was taking methylprednisolone 8 mg/day, long-acting nifedipine 30 mg/day, furosemide 25 mg/day and NSAIDs if necessary.

At the first clinical examination, the patient was conscious, compliant but anxious; his pulse, blood pressure, respiration rate and axillary temperature were 96 beats/min, 120/90 mmHg, 26 breaths/min. The patient was smoker (7-10 cigarettes/day) and denied alcohol consumption. At the moment of observation he was taking methylprednisolone 8 mg/day, long-acting nifedipine 30 mg/day, furosemide 25 mg/day and NSAIDs if necessary. The patient was conscious, compliant but anxious; his pulse, blood pressure, respiration rate and axillary temperature were 96 beats/min, 120/90 mmHg, 26 breaths/min.
and 36.6°C respectively. Chest and abdomen examinations were normal. A systolic ejection murmur (2/6 Levine scale) elective to the mesocardium was detected. The patient complained of severe pain at the cervical spine with irradiation to the arms which was exacerbated by digital pression on the cervical spinous processes (C3-C6). Subcutaneous rheumatoid nodules on the elbows and knees, and morphological alterations of the small joints of the fingers and toes and extrarotated attitude (swan-neck deformities of the phalanges) were present. Furthermore, a physical examination showed atrophy of the arm and leg muscles, prevalent on the right side. Deep tendon reflexes of the arms were normal, and the adductum, patellar and Achilles reflexes were symmetric but reduced. Babinski sign was bilaterally absent, and no deficit of strength or alterations of tactile and pain sensibility were detected.

At 9 am the patient was suffering from intense pain to the neck and back. A clinical re-evaluation showed that he was not able to move his hands or arms, while he was still able to raise his shoulders. Deep tendon reflexes of the arms were absent, as well as the patellar and adductum reflexes, while the Achilles reflex was less evocable and asymmetrical (the left more than the right). Babinski sign was bilaterally absent. Moreover, there was a tactile and painful hypoaesthesia of the legs and trunk beginning from the base of the chest. Fever was present (T: 38.4°C). A treatment with ceftriaxone 2 g intravenously/day, omeprazole 20 mg/day, nifedipine 30 mg/day, furosemide 25 mg/day and methylprednisolone 8 mg/day was performed. To reduce the pain, 200 mg of ketoprofen IV and 100 mg of tramadol intramuscular (IM) were administered without results, followed, at 11.30 AM, by 10 mg of chloridrate morphine used of antipyretic drugs. A peripheral blood sample revealed after 24-36 hours leucocytosis (11.9 × 10^9/L), platelets count 110 × 10^9/L (150-450 × 10^9/L), erythrocyte sedimentation rate 38 mm/hr (0-10 mm/hr), fibrinogen 8.18 g/L (1.5-4.0 g/L), creatinine 141.4 µmol/L (53-123.7 µmol/L), gamma glutamyl transferase 61 U/L (11-50 U/L), lactate dehydrogenase 617 U/L (230-460 U/L), creatine-phosphokinase 394 U/L (38-174 U/L), iron 1.074 µmol/L (10.74-28.64 µmol/L), prostatic specific antigen 48.6 µg/L (0-4.0 µg/L).

Hence, on the basis of both clinical and radiological findings two interpretative possibilities were hypothesized: (1) an osteophytic spondylosis with secondary compressive myelopathy; (2) an RA involvement of vertebral spine, with partial medullary compression. Thus, an “acute” recent phenomenon, e.g. a vertebral fracture, could have “unbalanced” an already unstable situation. Rheumatologic and orthopedic consultants, agreed with the above-mentioned hypotheses, recommending immobilization of the patient and pain control.

In the following hours the patient presented a progressive worsening conscious state and of clinical condition, with fever (39.5°C). At 5 pm he was drowsy, opened his eyes if stimulated but he was not able to answer questions; was able to swallow but not to cough, and the Glasgow Coma Scale (GCS) score was 9. Moreover, a flaccid quadriplegia was present. Brain computed tomography (CT) scan was normal. Microbiologic studies (blood and urine cultures for bacteria, fungi, and mycobacterium tuberculosis) were performed, and a positive hemoculture was found for Staphylococcus aureus. At 9 pm the clinical status of patient worsened to a deep coma, with miotic and poorly reacting pupils and frequent and superficial breath. GSC score was 5. Magnetic resonance imaging (MRI) of cervical spine showed a stenosis of neurocanal between C4 and C6 with concomitant spinal cord compression. Vertebral bodies C4, C5 and C6s showed degenerative phenomena associated to synovial cloth in correspondence to the odontoid (Figure 1).

In the following hours, the patient’s condition progressively worsened, with deepening of coma, miosis, frequent and superficial breath. GCS score was 3. High fever was present, despite the use of antipyretic drugs. A peripheral blood sample revealed after 24-36 hours leucocytosis (11.9 × 10^9/L), platelets count 93 × 10^9/L, fibrinogen 17 g/L, creatinine 592.2 µmol/L, potassium 7 mmol/L. Despite the above-mentioned therapy the clinical condition worsened progressively and death occurred 84 hours after admission.

The autopsy revealed the presence of severe ulcer-cerous-thrombotic lipohyaline and calcific arteriosclerosis of the aorta, coronary tree (circumflex branch and of the right coronary showed stenosis of the 70%), cerebral, and renal vessels as well; a moderate left ventricular myocardial hypertrophy with dilatation; an osteosclerotic narrowing of the cervical canal at the level of C3-C6 associated with recent cervical medulla infarction due to thrombosis of the anterior spinal artery. Multiple infarctions within the limits of a prostatic multin-
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Figure 1. Cervical Magnetic Resonance image: The odontoid process is in axis and shows normal articul ar intercourse with the anterior arc of the atlas, and is surrounded by hypointense pathological tissue. The vertebral bodies of C4, C5 and C6 and the discs between them are considerably altered and reduced in height. Slight anteroposterior slip of C2 on C3 is present. The spinal canal presents a reduced diameter between C4 and C6 where the cervical spinal narrow appears bulgy, with diffused alteration of the signal and complete obliteration of the subarachnoid perimedullar spaces.

odular hyperplasia were found. Finally, ulcerative bulbar duodenitis with light hemorrhage and a multiple organ acute congestion were detected.

Histopathologic features confirmed the presence of collagenous necrosis of occipitoatlantid ligaments with a poorly neutrophil inflammatory reaction; pachymeningitis with suppurative aspects, vasculitis, thrombosis and periradicularitis were present at the level of spinal marrow and relative meningeal membranes (Figure 2); coagulative necrosis of cervical spinal cord nervous tissue was also found. The prostatic gland showed multiple infarctions within the limits of a multinodular hyperplasia.

Conclusive diagnosis was suppurative pachymeningitis due to *Staphylococcus aureus* sepsis (the probable source was from limb injuries) with vasculitis and occlusion of the anterior spinal artery; neurocanal narrowing following a severe spondylopathy (C3-C4) in subject with rheumatoid arthritis.

Discussion

RA frequently involves the cervical spine and may lead to neurologic impairment by bone compression, and/or cartilage, bone and ligaments destruction. Abnormal mobility in the C1-C2 region is most common, but lower levels may also be involved. If epistropheus dens moves up through the foramen magnum, producing basilar invagination and threatening the upper cervical cord and medulla, a basilar-vertebral insufficiency may also occur, with syncope and sometimes sudden death. RA localized to the apophyseal joints and disc spaces may cause subluxation at different levels, which may slowly lead to quadriplegias.

The clinical manifestations of cervical RA are pain, typically localized in the neck and occipital area, neurological disturbance and, potentially, death. Basilar-vertebral insufficiency may cause loss of equilibrium, tinnitus, vertigo, visual disturbance and diplopia. Subjective sensations may be altered, yielding paresthesias, numbness and sensations of hot and cold. Bulbar compromise could be paroxysmal and could determine sudden death.

Inflammatory central nervous system (CNS) involvement in RA occurs in the setting of longstanding, active, erosive articular disease and is accompanied by extracranial and extraspinal nodules and vasculitis. CNS rheumatoid nodules are commonly asymptomatic, in contrast to CNS vasculitis.

Pachymeningitis is an inflammatory process of the dura, usually associated to infections of neighboring structures, epidural or subdural. The spinal isolated pachymeningitis is altogether a fairly rare pathology. Rheumatoid pachymeningitis could be a rare complication of RA and can occur either very early or after many years of disease onset.

*Staphylococcus aureus* is the micro-organism most often implicated, and weakening diseases and immunodeficiency syndromes can be predisposing conditions.
The peculiarity of the present case is the concurrence of several unfavorable conditions concurring at the clinical state. First, the patient was old and presented a RA which determined a serious alteration of the cervical spine through the years, although the clinical status was apparently stable. Here the marked narrowing of the spinal canal also produced a vascular compromise. Hypothenia of the legs with a tendency to fall, already referred in the patient’s recent history, could be attributed to a “medullar angina”, with a clear-cut worsening in the days before admission. In fact, about 10 days before, he accidentally fell, suffering bruises and grazes to the legs, and two days before that he complained of the onset of back pain localized at the dorsal spinal column which radiated to the arms with dysaesthesia. *Staphylococcus aureus* sepsis may determine, on one hand, a pachymeningitis and on the other, the vasculitis phenomena.

In the present case, the pachymeningitis confirmed only by a histopathologic study has the extremely rare peculiarity to be located essentially at the level of the dura and the cervical spine only, without involvement of arachnoid and pia mater. Such an inflammatory process produced a rupture of the precarious vascular equilibrium, determining a vasculitis at the level of the small arterial vessels contained in the same dura, the occlusion of the anterior spinal artery and, therefore, the medullar infarction. The pachymeningeal involvement was essentially inflammatory, and there were not rheumatoid nodules-like granulomas at the level of dura.

The above-reported clinical investigations did not allow the determining of the original source of infection. On the basis of multiple hemocultures positive for *Staphylococcus aureus*, we can reasonably suppose in this case, that this bacterial organism, a skin saprophytic germ, was the cause of sepsis. It’s likely that the grazes suffered to the legs some days before admission could have been the microorganism’s way in. The severity of the infection could be explained by the likely compromise of the immune system of the patient who was old and had been treated with steroids, though at relatively low doses, for many years.

**References**


