Craniocervical instability associated with rheumatoid arthritis: a case report and brief review

Eric Chun-Pu Chu[1]^ , Arnold Yu-Lok Wong[2]^, Linda Yin-King Lee[3]^

[1]New York Chiropractic & Physiotherapy Centre, New York Medical Group, Hong Kong, China; [2]Department of Rehabilitation Sciences, The Hong Kong Polytechnic University, Hong Kong, China; [3]School of Nursing and Health Studies, The Open University of Hong Kong, Hong Kong, China

Correspondence to: Eric Chun Pu Chu, BSc, DC, FRCC. New York Chiropractic and Physiotherapy Centre, 41/F Langham Place Office Tower, 8 Argyle Street, Hong Kong, China. Email: eric@nymg.com.hk.

Abstract: Rheumatoid arthritis (RA) is an autoimmune disease that affects the synovial tissue which lines joints and tendons. The craniocervical junction is made up exclusively of synovial joints and ligaments and especially vulnerable to the inflammatory process of RA. The chronic inflammation of RA leads to loss of ligamentous restriction and erosion of the bony structures and results in craniocervical instability (CCI). This is a case report of an 80-year-old woman who had been diagnosed with seropositive RA two decades ago presented with head dropping and losing balance while walking for several months. Radiographic images of the cervical spine showed RA-related features of instability in the form of atlantoaxial instability, cranial settling and subaxial subluxation. Since physical therapy and acupuncture previously failed to provide a substantial, long-lasting outcome, the patient sought chiropractic care for her condition. The chiropractic regimen consisted of upper thoracic spine mobilization/adjustment, electrical muscle stimulation of the cervical extensors, home exercises and neck bracing. She regained substantial neck muscle strength, gaze angle and walking balance following a 4-month chiropractic treatment, although cervical kyphosis persisted. The current study aims to provide basic knowledge of CCI associated with RA and ability to modify a treatment program to accommodate the needs of patients with coexisting red flags.

Keywords: Cervical spine; chiropractic; craniocervical instability (CCI); head dropping; rheumatoid arthritis (RA)

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Introduction

Craniocervical instability (CCI) is a pathological condition in which ligamentous connections from the skull to the spine are incompetent (1). Rheumatoid arthritis (RA) is an immune-mediated inflammation that primarily affects the lining of the synovial joints and tendons (2). The craniocervical junction consists exclusively of synovial articulations between the occipital condyles, atlas (C1) and axis (C2). The characteristics of the craniocervical articulation make it especially vulnerable to the involvement of RA (3). Recent estimates suggest that up to 86% of RA patients have radiographic evidence of cervical spine involvement (4) even within 2 years after the initial diagnosis of RA (5). Insidious synovitis may cause erosion or incompetence of the involved ligaments, bones, or joints, and resultant CCI (3). The consequence of CCI takes the form of atlantoaxial subluxation, cranial settling

^ ORCID: Eric Chun-Pu Chu, 0000-0002-0893-556X; Arnold Yu-Lok Wong, 0000-0002-5911-5756; Linda Yin-King Lee, 0000-0002-8588-3556.

and subaxial subluxation. Patients may report a sensation of the head dropping forward during cervical flexion. Neurological deficits, spinal stenosis and myelopathy can occur depending on the degree of damage to the articulation (5). Conservative treatment is the mainstay of initial treatment for CCI without progressive neurological deficits (6). Surgery is aimed at halting further neurological deterioration. However, a multidisciplinary approach involving rheumatology, rehabilitation and surgery is beneficial to optimize outcomes (3). Familiarity with the pathogenic processes of CCI associated with RA, the imaging features, and a basic knowledge of therapeutic alternatives are prerequisites for a personalized plan of treatment.

We present the following article in accordance with the CARE reporting checklist (available at http://dx.doi.org/10.21037/acr-20-131).

Case presentation

An 80-year-old Chinese woman complained of an inability to hold the head in an upright position and losing balance while walking for several months. Her medical history was notable for chronic stiffness and pain which began in the finger joints, mild pain in the neck and shoulders, and insidious development of cervical kyphosis. The elbows and knees were not affected. She had previously been given a diagnosis of seropositive RA two decades before. Due to her concerns about drug safety and side effects, she only took non-steroidal anti-inflammatory drugs for pain relief as needed. Her past medical history included hypertension, hyperlipidemia and sacral fracture. Recent workup by a neurosurgeon had ruled out cranial and neurological diseases. Her cervical magnetic resonance images showed widespread spondylotic changes and multi-level vertebral wedging. In the past few months, she had been treated with physical therapy and acupuncture but achieved minimal improvement of her head and neck posture. As such, she sought chiropractic care for her neck condition.

Upon examination, the patient was in a head dropped posture and ambulated with a cane to maintain balance. Swan-neck deformity of fingers in both hands was noted and the tendons on the dorsum of the hands were prominent and tight. Pain intensity of the neck and figure joints was rated 3–4/10 on a numeric pain score. The dropped head position was correctable by lying in a supine position. Reduced muscle tone and grating sensation were noticed on nuchal palpation. Passive ranges of neck motion (performed by the tester) were full, while active neck extension (performed by the patient) was limited due to weakness of cervical extensors to raise the head. Manual muscle testing of the bilateral upper trapezius and cervical extensors were grade 1 out of 5, where 0 means no voluntary contraction and 5 means normal muscle function. Pre-treatment EOS® imaging (*Figures 1,2A*) demonstrated reversed cervical kyphosis, superimposition of landmarks of the basal occiput upon the C1 vertebra, anterior subluxation of the C1 (dotted white arrow) on C2 (white arrow) (atlanto-dens interval 3.7 mm), broken spinolaminar line (red curved line) at C2–C3 (anterolisthesis of the C2 upon C3 by 3 mm), and multi-level disc narrowing, multi-level vertebral wedging with osteophytosis. It was difficult to identify the tip of the dens. If the dens extends >4.5 mm above the McGregor line (from posterior hard palate to base of occiput) basilar invagination is present. It was speculated that the loss of head balance and walking imbalance was a result of CCI associated with RA accompanied with anterior vertebral wedging at the middle and lower cervical spine.

In the absence of neurological deficits, the first phase of treatment aimed to improve neck muscle strength. Gentle mobilization of the upper thoracic spine, electrical muscle stimulation (EMS) of the cervical extensors, and home-based neck exercises for motor strengthening were prescribed. Home-based exercises included shoulder shrugs (slowly raise both shoulders up, and hold for 5 seconds), head retraction (use a couple of fingers to tuck the chin gently backwards against the neck, and hold for 5 seconds), and neck glide (slowly slide the chin forward, and hold for 5 seconds). Each exercise repeated for about 5–10 repetitions. The patient did the exercises for at least four rounds a day and wrote them down to make sure the training was done properly. In addition, a cervical brace was prescribed to maintain an upright head posture during daily activity. After 2 weeks of treatment 5 times per week, improved head posture and walking balance were observed. The neck pain score declined from 3–4/10 to 1/10 on the numeric rating scale. Neck muscle strength improved to grade 2 out of 5. Subsequently, the second-phase treatment focused on structural correction. Frequency of treatment was reduced to three times weekly for 2 months. Chiropractic manipulation was applied to the upper thoracic segment by using a segmental drop table to perform a high velocity, low amplitude adjustment with low force. EMS was included in all treatment sessions. Upon completion of the treatment program, the patient reported a relief of neck and shoulder pain and the finger joints still were mildly painful because

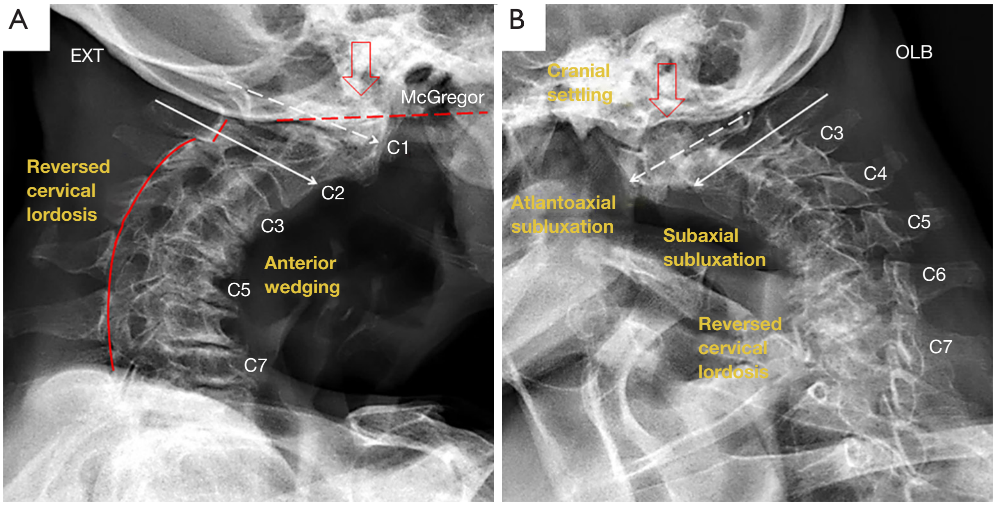

Figure 1 Pre-treatment extension (A) and oblique (B) cervical EOS® images demonstrated vertebral instability consisting of reversed cervical lordosis, superimposition of landmarks of the occipital base upon the C1 vertebra, anterior subluxation of the C1 (dashed white arrow) on C2 (white arrow), broken spinolaminar line (red curved line) at C2–C3, multi-level disc narrowing, and multi-level vertebral wedging with osteophytosis. It was difficult to identify the tip of the dens. If the dens extends >4.5 mm above the McGregor line (from posterior hard palate to base of occiput) basilar invagination is present. Red hollow arrow stands for the force of the gravity of the head. McGregor line (dashed red line) is used in the assessment of basilar invagination. EXT, extension view; OLB, oblique view.

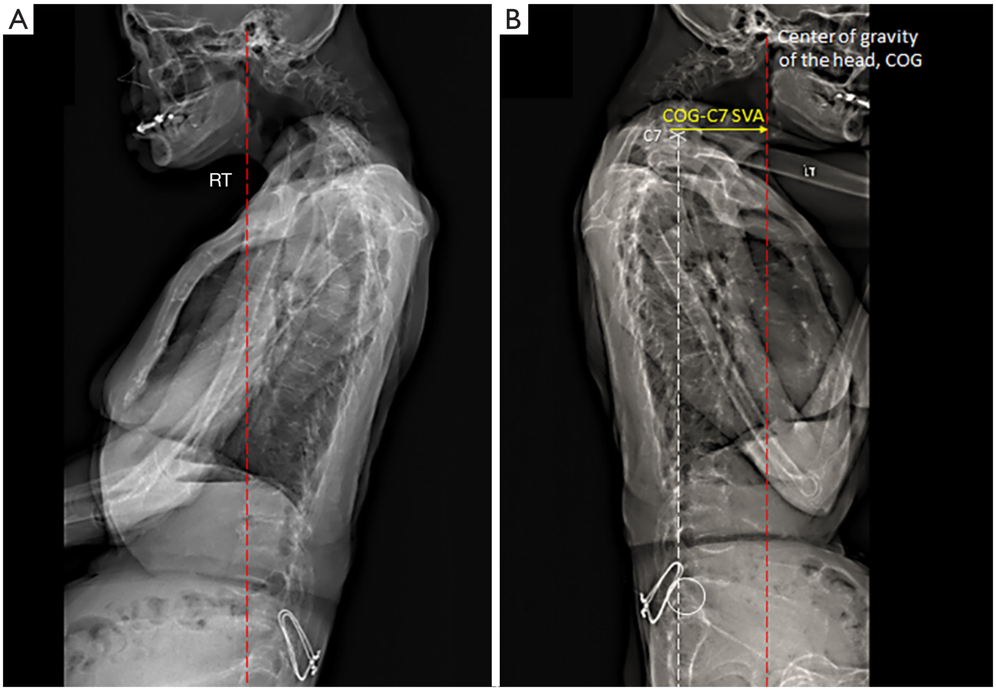

Figure 2 Postural assessment. (A) At initial assessment, the patient could not stand unaided. She supported herself with the right hand holding door handle and the back leaning against the cabin interior. Sagittal EOS® radiograph showed severe cervical kyphosis, degenerative spondylosis and s/p internal fixation of the sacrum. (B) At 4-month follow-up, sagittal EOS® imaging demonstrated cervical imbalance with respect to offset of the center of gravity of the head [COG-C7 sagittal vertical axis (SVA) offset, yellow arrow]. With straightening of the lumbar spine, the C7 plumb line (dashed white line) was tangential to the posterior-superior corner of the upper sacral endplate (white circle), suggestive of a compensated global sagittal balance. In the sagittal plane, the gravity line (dashed red line) passes slightly posterior to the hip joint.

Figure 3 Comparison of the patient's posture. (A) Upon examination, the patient was in a head dropped posture and ambulated with a cane to maintain balance. (B) At the 4-month follow-up, she regained substantial gaze angle along with walking balance, although cervical kyphosis and some weakness of neck extensors persisted.

of rheumatoid changes. Neck muscle strength was further improved from grade 2/5 to 2+/5. After that the use of cervical brace was discontinued. She regained substantial gaze angle along with walking balance at the 4-month follow-up (*Figures 2B,3*), although cervical kyphosis and some weakness of neck extensors persisted. No adverse events had occurred. The patient was encouraged to continue home-based neck exercises to battle head dropping.

All procedures performed in this study were in accordance with the ethical standards of the institutional and national research committees and with the Helsinki Declaration (as revised in 2013). A written informed consent was obtained from the patient.

Discussion

The pathogenesis of RA is not completely understood. Recent evidence suggests a potent bidirectional interaction between the T cells (T lymphocytes) and synovial fibroblasts leads activation of the proinflammatory cytokines (tumor necrosis factor-α, interleukin-1 and interleukin-6) that drive synovial inflammation (7). During the course of RA, the synovium transforms into a hyperplastic invasive tissue that leads to destruction of cartilage, subchondral bone and periarticular structures of the affected joint (8). The craniocervical junction is composed exclusively of synovial articulations and vulnerable to the aforementioned effects of RA.

Firstly, synovial dendritic cells are the key professional antigen-presenting cells able to attract a cluster of T-cells to the affected joints and activate antigen-specific T-cell responses (9). The interactions between T cells and synovial fibroblasts from the lining layer could contribute to the synovial hyperplasia and formation of the synovial pannus that compromise the bony structures and ligamentous integrity of the craniocervical junction (2). In such a situation, the head pulls the atlas (C1) anteriorly from the axis (C2) during neck flexion, and an anterior atlantoaxial subluxation takes place (5). A recent review found that atlantoaxial subluxation accounts for 65% of cervical complications associated with RA (10). Secondly, as the inflammatory activity continues, lateral atlantoaxial facet joints may be damaged, and the weight of the head will press the atlas downward around the axis. When articular structures between the skull base, atlas, and axis are disrupted cranial settling (i.e., downward subluxation, atlantoaxial impaction) occurs (5). Thirdly, the transmission of axial load down the subaxial area can cause degenerative discitis, vertebral erosion and fragility fracture, which may result in multiple subaxial subluxations and subaxial ankylosis (11).

The combination of forward flexion and excessive loading forces causes progressive damage to the anterior portion of the cervical vertebrae. Degeneration of the disco-ligamentous structures and osteoporosis with aging are aggravating factors for wedge fracture of the cervical spine. In terms of radiological examinations, a reduction in anterior height ≥20 % or ≥3 mm of the posterior height of the affected vertebra is evidence of a wedge fracture (12). In the absence of any posterior disruption, anterior wedge fracture is generally a mechanically stable fracture. However, radiographs showing anterior wedging ≥50% or multiple adjacent wedge fractures, as seen in the present case (*Figure 1*), are indicators of cervical spine instability (13). Unstable vertebrae can shift and also press on nearby structures such as the nerve roots, spinal cord and vertebral arteries. CCI associated with RA may increase the risk of myelopathy and sudden death (5). Reported mortality rate in RA patients was as high as 50% in the first year of developing myelopathy (5,14).

Although patients with cervical instability and spinal canal stenosis have a tendency to deteriorate, conservative treatment remains the mainstay of management for CCI without progressive neurological deficits (6,11,15). The primary goal of conservative treatment is to improve the quality of controlled motion, to limit progression, to prevent neurologic injury, and to avoid unnecessary surgery. Optimal conservative treatment should be multidisciplinary (3), consisting of disease modifying antirheumatic drugs, immunosuppressive therapies, symptomatic treatments, cervical collars, physical therapy exercises, occupational therapies, and nursing support on promoting optimal physical and psychosocial functioning. The pharmacological strategies can slow the progression of RA, but medications cannot reverse damage that has already occurred. The presence of ligament disruption, joint erosion, bone destruction and vertebral wedging represent aberrant changes in the spinal structures. Surgery is for only those who fail repeated nonsurgical treatments, or in the presence of progressive neurological symptoms of spinal cord compromise (11).

Asymptomatic cervical instability is quite common in people with RA, where up to 50% of patients with anterior atlantoaxial subluxation may be unaware of the abnormality (16). Since there is only limited capacity for subluxation to progress only a few patients will eventually require surgery (15). The main complication of CCI is spinal cord compression, especially if the vertebrae get misaligned. According to the World Health Organization guidelines (17), high-velocity thrust procedures are contraindicated in the anatomical areas of pathology. In the interest of patient's safety, cervical manipulation should not be used in the cases in question. Treatment instead consisting of gentle mobilizations through pain-free ranges of motion and other manual techniques applied to the upper thoracic spine were alternate options for a personalized plan of treatment (18). As seen in the current case, chiropractic care could help by relaxing hypertonic musculature, relieving neural compromise, disrupting periarticular adhesions, and restoring spinal function (19). Subsequent EMS and neck exercises might improve muscle endurance and strength to keep the head upright. Neck bracing might support the weight of the head and keep pressure off any strained neck muscles. Appropriate conditioning and proper control of biomechanical stresses can allow the joint to function properly, although structural changes are irreversible.

Among the limitations of this study, the following should be considered. The first was that a single number of subjects investigated required confirmation of the observed findings in a larger population. Second, the chiropractic regimen consisted of spine mobilization/adjustment, EMS, home exercises and neck bracing. In the absence of control groups, benefits from different treatment options cannot be established. Moreover, radiographic interpretation of the present study was based on EOS® radiographs. Discrepancy between EOS® full spine radiograph and standard radiographs exists due to variations in image projection, patient positioning and weight-bearing conditions of the anatomic structures of interest. However, the strength of the present study was to provide basic knowledge of CCI associated with RA and ability to modify a treatment program to accommodate the needs of patients with coexisting red flags.

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Footnote

Reporting Checklist: The authors have completed the CARE reporting checklist for case reports. Available at http://dx.doi.org/10.21037/acr-20-131

Conflicts of Interest: All authors have completed the ICMJE uniform disclosure form (available at http://dx.doi.org/10.21037/acr-20-131). The authors have no conflicts of interest to declare.

Ethical Statement: The authors are accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved. All procedures performed in studies involving human participants were in accordance with the ethical standards of the institutional and/or national research committee(s) and with the Helsinki Declaration (as revised in 2013). Written informed consent was obtained from the patient.

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