Case Report

Upper cervical instability associated with rheumatoid arthritis: a case report

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Rheumatoid arthritis (RA) affects between 1 and 2 million individuals in the United States. Atlantoaxial instability (AAI) has been shown to have 40–85% prevalence among individuals with RA. Despite the high incidence of craniovertebral involvement, overt symptoms of instability are rare. The high risk of AAI and limited symptomology should increase therapist suspicion of potential contraindications and precautions to initiation of therapy for the cervical spine without prior diagnostic imaging. The purpose of this case study is to describe the historical, clinical, and diagnostic imaging complexity of AAI associated with RA, and to illustrate the use of these factors in the clinical reasoning within a patient case.

Keywords: Cervical instability, Atlantoaxial instability, Rheumatoid arthritis, Physical therapy

Introduction

Rheumatoid arthritis (RA) is a multisystem disorder distinguished by synovitis that affects between 1 and 2 million individuals in the United States.^{1,2} The musculoskeletal features of synovitis consist of symmetrical inflammation and resultant deformity of smaller joints including those of the hands, wrists, elbows, feet, ankles, and knees.³ In addition to small joint deformities, atlantoaxial instability (AAI) has been shown to have 40–85% prevalence among individuals diagnosed with RA.^{4–10} Despite the high incidence of craniovertebral involvement due to weakening of the transverse ligament, there is not a consistent presentation of AAI in this population.¹¹ Due to the absence of reliable symptoms of AAI, physical therapists should routinely screen patients diagnosed with RA for cervical instability prior to treatment of the cervical spine.¹² The purpose of this case study is to describe the historical, clinical, and diagnostic imaging considerations associated with AAI in patients with RA and to illustrate the influence of these factors on the clinical reasoning process within a patient case.

Background

Atlantoaxial instability is defined by hypermobility between C1 and C2. An atlantoaxial distance greater than 5 mm, demonstrated by lateral radiographs, is indicative of AAI.⁶ Dynamic lateral views of the cervical spine may be more useful than static, neutral views in detecting atlantoaxial subluxation. Upper cervical instability can be determined by a

displacement of 3.5 mm on functional flexion-extension films.⁶ Although the radiographic findings are well-defined, there are multiple misconceptions regarding the signs and symptoms related to the diagnosis of AAI, including the belief that suboccipital pain is consistently present with instability. Suboccipital pain is the most common symptom of AAI, however, it may be absent in more than half of the individuals with known instability.⁷ Disease duration has also been traditionally associated with increased risk of cervical instability.^{8,9} In contrast to beliefs of increased subluxation risk with disease progression, a prospective study demonstrated that 83% of atlantoaxial subluxations occur within the first 2 years following the diagnosis of RA.¹¹ Thus, it would be incorrect for a physical therapist to lower the suspicion of AAI based on the belief that instability is a sequela of later stages of the disease process. There is also a significant discrepancy in the literature regarding the presence of neurological signs and symptoms with AAI.¹³ Neurological signs have been described as hyper-reflexia, spasticity, pareses, paresthesia, overt loss of balance with head movement, nystagmus, and poor coordination with ambulation. Neurological symptoms that have been reported include headache, dizziness, tinnitus, and dysphagia.^{13,14} In contrast, it has been shown that neurological signs and symptoms may not be present in subluxations of up to 10 mm.^{6,15} Therefore, conventional signs and symptoms related to AAI including suboccipital pain, length of time since diagnosis with RA, and the presence of overt neurological symptoms cannot be used as reliable predictors of upper cervical instability.

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Access to diagnostic imaging of the cervical spine may assist the physical therapist in substantiating the presence of AAI when unobstructed views can be obtained. Cervical radiographs continue to be the most common method of identifying AAI.⁶ The current European League Against Rheumatism (EULAR) guidelines suggest monitoring of functional instability of the cervical spine in patients with clinical symptoms of AAI through lateral radiographs performed in flexion and neutral.¹⁶ However, bone destruction and osteoporotic changes associated with RA can create radiographic images of the cervical spine that may be difficult to interpret.^{6,10} Alternative diagnostic imaging, such as MRI, is more sensitive than radiographs in detecting early structural changes in joints.¹⁷ Nevertheless, cost limits the use of MRI in daily clinical practice and it is currently only recommended when cervical radiographs are positive or neurological signs and symptoms are present.¹⁶

When unobstructed cervical radiographs or MRI films are not available for review, the therapist should consider the common musculoskeletal features of RA. In addition to cervical spine views, utilization of radiographs of the peripheral joints has been shown to assist in the determination of the risk of AAI as a component of the Large Joint Index.^{9,18} Plain radiographs provide optimal documentation of joint degeneration and destruction.^{17,19} The Larsen method is one system of classifying joint degeneration in individuals with RA.¹⁷ The method uses a continuous quantitative scale of measurement that includes a global score of both joint space narrowing and erosion for each joint.¹⁷ Each joint is graded on a score of 0 (normal) to 5 (maximal destruction). The 40 joints in the hands and feet are scored emphasizing the destruction that generally takes place in the small joints.¹⁹ The joints utilized include the proximal interphalangeal joints of digits 1–5 in both the hands and feet, metacarpophalangeal and metatarsophalangeal joints of digits 1–5, and 4 regions in the wrist.¹⁹ The minimal clinically important difference (MCID) for the Larsen method is approximately 2 points on a 200 point scale or 1% of the maximum score.¹⁷ Table 1 provides the scoring guide for the Larsen method.¹⁷

The large joint index utilizes the Larsen method and has been recommended to assist with establishing the risk of AAI in individuals diagnosed with RA.⁷ The large joint index is an 8 point scale based on

radiographs of the shoulders, elbows, hips, and knees. A Larsen grade ≥ 2 at each joint is scored as 1 point. A Large Joint Index ≥ 4 has been suggested as a predictor of AAI with a sensitivity of 86% and specificity of 62%.⁷ Therefore, the degree of peripheral joint involvement can assist the therapist in assessing the patient's risk factors for AAI.

In addition to the severity of upper cervical pathology being related to radiographic evidence of peripheral joint degeneration, multiple studies have explored the relationship between the presence of AAI and the necessity for total joint replacement in individuals diagnosed with RA. In studies of patients with RA scheduled for hip or knee replacement surgery, 44–65% demonstrated AAI or previous surgical cervical fusion on pre-surgical radiographs.^{4,7,8,10,12} The anomaly noted on imaging of these patients was not associated with symptoms of upper cervical instability. Simply, a history of large joint arthroplasty may be an indicator of potential upper cervical instability in individuals diagnosed with RA.

Patient Characteristics

This case study involves a 62-year-old female diagnosed with RA presenting to physical therapy with the primary complaint of left shoulder pain following a difficult commode transfer. She was referred to physical therapy by an internal medicine physician for the treatment of a suspected left rotator cuff tear. The patient was not a surgical candidate due to cardiovascular pathology and, therefore, diagnostic imaging of her shoulder had not been performed. The patient's extensive medical history included her being diagnosed with RA at the age of 42 with a resultant loss of ambulatory status 4 years previously. Her surgical history was significant for bilateral total knee replacement surgery at the age of 55 and a right radial fracture with rod placement at the age of 57. Additional comorbidities included pericarditis leading to pericardial effusion and cardiomyopathy, coronary artery disease, hyperlipidemia, hypothyroidism, uterine cancer, and depression. She was being regularly seen by several specialists including cardiology, rheumatology, and internal medicine.

Examination

The patient was transported into the therapy office via a manual wheelchair propelled by her spouse. On

Table 1 Larsen method of classifying joint degeneration in individuals with rheumatoid arthritis (RA)¹⁷

0	Intact bony outlines and normal joint space
1	Erosion less than 1 mm in diameter or joint space narrowing
2	One or several small erosions, diameter more than 1 mm
3	Marked erosions
4	Severe erosions, where there is usually no joint space left, and the original bony outlines are partly preserved
5	Mutilating changes, where the original bony outlines have been destroyed

observation, the patient's hands demonstrated severe metacarpophalangeal ulnar drift and proximal interphalangeal joint hyperextension of digits 2–5. It was noted that the patient supported her head with her left hand maintaining a position of significant left lateral flexion of the cervical spine. Furthermore, the patient's left ear appeared to be touching her left shoulder.

With questioning regarding previous treatment for neck or upper quarter pain, the patient described prior physical therapy for the diagnosis of 'torticollis'. The patient noted the curvature in her spine began within the last 18 months and progressed quickly since onset. She had radiographs taken at the time of the initial diagnosis of torticollis and was referred to physical therapy. Her previous physical therapy treatment consisted of soft tissue mobilization and stretching of the left side musculature including the levator scapulae, upper trapezius, and sternocleidomastoid (SCM). The patient elected to discontinue physical therapy after eight visits due to a lack of progress and the cost associated with transportation. The patient was given exercises to perform at home including static stretching of involved cervical musculature and advice for upper cervical positioning. The patient had not sought treatment for her cervical dysfunction since this initial therapy consultation 18 months ago.

The patient denied conventional neurological symptoms of upper cervical instability including a history of dizziness, blurred vision, headache, numbness, tingling, recent loss of coordination, and new onset weakness.¹³ The patient did report a slight dull ache at the left occiput, which had been present for ~18 months. The patient felt that this discomfort had not progressed during that timeframe. Based on this complaint, the patient was asked to complete the neck disability index (NDI). The NDI score was not representative of neck pain, but of overall mobility and activity limitations with the patient scoring an 84/100.

Despite the lack of overt neurological symptoms during the subjective portion of the evaluation, the suspicion of AAI was increased. The patient's past medical history of pericarditis and cardiomyopathy revealed extra-articular organ involvement associated with RA. She also demonstrated significant symmetrical small joint destruction with multiple deformities and reported a history of bilateral total knee arthroplasty. Moreover, the patient demonstrated an abnormal cervical spine posture with the complaint of a dull suboccipital ache. Suboccipital pain, torticollis, and external support of the head have all been noted in the literature as red flags for upper cervical instability.^{20–22} Therefore, the initial physical therapy examination focused on the upper cervical

spine and possible precautions to treatment, rather than the prescribed interventions for a suspected rotator cuff tear.

Palpation revealed significant tenderness and increased muscle tone along the left SCM, levator scapulae, upper trapezius, and scalene musculature. The patient also reported tenderness along bilateral suboccipital muscles upon palpation. The patient demonstrated significantly limited active range of motion in all directions with the cervical spine relatively fixed in left lateral flexion. A rotational postural deformity with limitation of cervical range of motion has been described as the cock-robin position. It is differentiated from spasmodic torticollis by the absence of SCM muscle spasm.²³ Although the patient was tender along the involved SCM on palpation, muscle spasm was not apparent. This presentation suggested that the abnormal cervical spine position was due to a rotational deformity. Based on the results of palpation and active range of motion testing, the physical therapist was hesitant to perform passive range of motion. Therefore, the therapist deferred this portion of the examination.

Following this initial objective testing, the therapist suspected upper cervical instability. The Sharp-Purser test has traditionally been used by physical therapists for the clinical assessment of cervical instability in individuals with RA.²⁴ The Sharp-Purser test has been shown in one study to have a specificity of 96% and a sensitivity of 88% with a predictive value of 85% for AAI specific to individuals with RA with a transverse ligament laxity greater than 4 mm.²⁴ The patient's abnormal posture limited the therapist's level of comfort utilizing the test based on a strong suspicion of instability. Positioning the upper cervical spine in flexion at the initiation of the test to allow for an anterior glide of C1 on C2 could possibly cause additional subluxation and symptom onset if the transverse ligament was not supporting the alignment of the segments. Consequently, special tests for upper cervical instability were not performed.

On the neurological evaluation, the patient presented with hypersensitivity to touch along the left C2 dermatome. All other cervical dermatomes were within normal limits. Long tract testing was also negative. The abnormal cervical position did not allow for traditional myotomal testing of C1–C3, however, the patient was able to actively move the cervical spine independently in the limited available range. Testing the left C5 myotome was limited by left shoulder pain and muscle substitution with shoulder abduction. In addition, strength testing of the wrists and hands was limited by pain and deformity. The C4 (upper trapezius), C6 (biceps), and C7 (triceps) myotomes were within normal limits and equal bilateral. Reflex testing of biceps and

Figure 1 Anterior–posterior view radiograph. This view demonstrates the cock-robin or torticollis position with convexity to the right in the upper cervical spine.

triceps was intact bilaterally at 2+ and Hoffman's sign was absent. The patient continued to lack the neurological signs associated with upper cervical instability including hyper-reflexia, spasticity, paresthesia, and nystagmus.

To further assist with the assessment, the physical therapist did have access to radiology films and reports performed 18 months previously. Radiographs of the cervical spine demonstrated a convexity to the right (Fig. 1). The views of C1 and C2 were obscured on lateral views of the neutral, flexed, and extended cervical spine (Fig. 2). Therefore, assessment of cervical instability based on the EULAR guidelines of lateral radiographs in flexion and neutral was ineffectual. Additionally, an open mouth view was available, however, the distal aspect of the odontoid could not be seen. Furthermore, the right

lateral aspect of the base of C1 and body of C2 could not be visualized (Fig. 3). It has been noted that cervical radiographs obtained in individuals with RA can be difficult to interpret due to the presence of deformity and, in this case, it was documented that the radiologist was unable to visualize the upper cervical spine due to the abnormal cervical spine position. The inability to rule out upper cervical instability could have been overlooked by the physical therapist if the actual radiographs had not been reviewed, in addition to the summary report provided by the radiology department. The primary care physician's note indicated that the patient had been referred to the rheumatologist for follow-up on the cervical radiographs. Additional imaging was not recommended by rheumatology due to a lack of neurological symptoms.

Figure 2 Lateral view – neutral, flexion, and extension positions. Note the inability to view entire cervical spine including the anterior arch of C1 and the odontoid process.

Figure 3 Radiograph – open mouth view. Although this projection should reveal C1 and C2 positioning, there is an inability to visualize the relationship between C1 and dens on the left, therefore limiting the assessment of upper cervical stability.

Clinical Impression

Although Larsen's grades of the peripheral joint degeneration and the large joint index were not available for review, the patient's multiple joint deformities and previous history of total joint replacement suggested that she was at increased risk of AAI. On examination, the patient's abnormal neck position with need for external support and complaints of a dull ache at the left occiput further increased the therapist's suspicion of instability. Limitations in the interpretation of the radiographs previously taken of the upper cervical spine supported concerns regarding precautions and contraindications related to continuing the physical therapy evaluation including joint mobility testing and passive range of motion of the upper cervical spine. The therapist believed that there were sufficient findings of concern to warrant physician referral prior to continuing the examination or initiating conservative treatment.

Referral

The patient was referred back to her internal medicine physician with a request for additional diagnostic imaging to address possible contraindications related to upper cervical instability prior to proceeding with physical therapy. The patient's previous radial fracture with rod placement excluded the use of magnetic resonance imaging (MRI). Computed tomography (CT) was performed without contrast in order to examine the upper cervical region. Severe degenerative changes were noted at

Figure 4 Computed tomography (CT) sagittal view. The CT shows a loss of disk height throughout cervical spine and reversal of the cervical curve. This view also reveals a relatively superior position of the anterior arch of C1 on Dens.

the C1 level with a pseudoarticulation between the odontoid and the occipital condyle (Figs. 4 and 5) indicating lateral displacement of C1.

The patient was referred by her internal medicine physician to neurosurgery, and surgical stabilization of the upper cervical spine was recommended. The patient declined upper cervical fusion due to risks associated with surgical intervention based on her past medical history of cardiomyopathy.

She returned to physical therapy and was examined for a power wheelchair with tilting and lower extremity elevation options. A custom head support was fabricated to assist with head positioning and to decrease cervical muscle tone associated with attempts to maintain an upright head position. The

Figure 5 Computed tomography (CT) axial view. The CT section demonstrates the displacement of lateral masses of C1 relative to dens.

tilting capability of the chair gave the patient multiple options for head support and weight shifting. Moreover, the wheelchair allowed the patient independence with mobility in her home and the community. Physical therapy did not include direct treatment of the cervical spine due to the patient's contraindication related to upper cervical instability.

Discussion

The patient did not present to physical therapy with the primary complaint of cervical pain. Nevertheless, the therapist not only deferred testing for upper cervical instability, but also requested that additional diagnostic imaging be performed prior to completing the examination and initiation of treatment. The patient's presentation with multiple small joint deformities, a history of previous total joint arthroplasty, an abnormal cervical spine position, and complaint of a dull ache at the left occiput increased the therapist's suspicion of AAI. Her presentation did not represent the overt neurological signs and symptoms typically associated with upper cervical instability. However, conventional myotomal testing was limited by the patient's joint deformities and left shoulder injury resulting in inconclusive neurological testing and the possible concealment of a new onset of weakness. In addition, it is important to note that neurological findings may not be positive with upper cervical instability.

Multiple precautions and contraindications are present in the literature regarding conservative treatment of AAI, which would limit physical therapy intervention. It has been noted that mobilizations should not be performed due to the risk of increasing C1 and C2 subluxation and the possible resulting catastrophic consequences.^{25,26} In regards to exercise, cervical protraction and retraction have also been associated with exacerbating upper cervical subluxation.²⁷ Furthermore, prolonged positioning of the cervical spine in flexion has been shown to result in the death of an individual with anterior atlantoaxial subluxation.⁶ Despite the inconclusive symptoms of AAI and multiple limitations in physical therapy intervention when instability is present, a survey of Australian therapists noted that only 64.4% of respondents would screen for upper cervical instability in patients with known RA prior to treating the cervical spine.¹² The possibility of devastating results with conservative physical therapy treatment of the cervical spine should guide therapists to use caution in the management of patients with multiple risk factors for instability and to include an assessment of upper cervical diagnostic imaging.

Conclusion

Due to the high incidence of AAI in individuals with RA, screening for instability is essential to ensure a

safe assessment. In this case, the patient presented without significant neurological symptoms yet demonstrated clinical signs of instability requiring advanced imaging and a neurosurgical consultation. In these cases, a thorough history and assessment of risk factors may assist the therapist in determining precautions for the examination and highlight the need for outside referral prior to completing standard tests and measures of the cervical spine. In addition to standard red flags for upper cervical instability (including suboccipital pain and abnormal cervical spine position), degeneration of multiple peripheral joints and a history of previous large joint arthroplasty are considered to be risk factors associated with AAI. It is imperative to note that procedures typically considered benign, such as cervical retraction exercises, can have a detrimental result when AAI is present and therapists should seek to rule out instability through diagnostic imaging prior to commencing treatment when certain clinical signs or risk factors are present.

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