Atlanto-axial instability in rheumatoid arthritis: a review

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Abstract

Objectives: The purpose of this literature review is to identify common lesions present in the rheumatoid neck with specific emphasis to atlanto-axial instability, review its clinical presentation, imaging findings and management.
Study design: A review of the English medical literature was done with focus on recent studies that covered the presentation, diagnosis, management and clinical outcomes of rheumatoid arthritis of the cervical spine specifically atlanto-axial instability.
Data extraction: A comprehensive literature review of the English medical literature was obtained through PubMed up to 2015 was performed identifying relevant and more recent articles that addressed the presentation, evaluation, surgical management and outcomes of rheumatoid patients with atlanto-axial instability.
Data synthesis: Atlanto-axial instability is a very debilitating disease with high morbidity and mortality if untreated. Onset of myelopathy is a poor prognostic factor with poor long-term survival. High index of suspicion and early intervention results in good outcomes and prevents neurological outcome.
Conclusion: Cervical spine involvement in rheumatoid arthritis is common and debilitating. Atlanto-axial instability is the commonest form. Early diagnosis and treatment is key in management. Early selective choice of patients for surgery results in better outcomes.

Introduction

Rheumatoid Arthritis (RA) is a chronic systemic inflammatory disease that primarily affects the joints. Although inflammatory arthritis of the small joints in the hands and feet is a common clinical manifestation, the cervical spine can also be affected. The first description of RA is found in the dissertation of Augustin Jacob Landré-Beauvais in 1800 in France when he first noticed the symptoms and signs of what we now know to be RA. He examined and treated a handful of patients with severe joint pain that could not be explained by other known maladies at the time (such as "rheumatism" or osteoarthritis)[1]. Rheumatoid arthritis is the most common inflammatory disorder of the cervical spine. Cervical spine involvement is a highly characteristic component in RA and other chronic inflammatory rheumatic diseases[2-5].

In 1890, A. Garrod described 178 patients with cervical spine involvement in a series of 500 patients with RA[6]. The reported prevalence of cervical involvement in rheumatoid arthritis patients varies from 17-86%[7-9]. The atlas-axis—cervical vertebrae 1 and 2 (C1 and C2)—articulation is one of the prime disease targets. The erosive pannus formation at this joint often leads to bony destruction and laxity in the surrounding ligamentous complex, especially the transverse ligament that aligns that atlas and axis. The subsequent loss of anchoring structures results in atlanto-axial subluxation (AAS)[10]. The subluxation can be anterior, posterior, lateral, and rotatory. The Anterior Atlanto-axial Subluxation (aAAS) is the most common subtype; the reported prevalence ranges from 10% to 55% followed by vertical subluxation and sub axial subluxation. The average female representation is 75%. The mean age at the time of outcome assessment was 58 years old (range of 33 to 69 years), and the mean disease duration was 12 years (range of 2 to 30 years)[11].

Clinical presentation

Clinical features are extremely variable. Sub-occipital pain is the commonest accounting for 40-85% of all patients and an early finding[12-14]. Pain that is worse when upright and relieved with recumbency is usually the result of compression of the greater occipital branch of C2, whereas

involvement of the auricular branch of C2 causes ear pain. Pain associated with subluxation is generally aggravated with neck motion, and patients may actually describe a clunking sensation or a feeling that their head is falling forward with flexion. Vague upper extremity clumsiness or weakness is also common, and patients have been described as often having difficulty finding the words to describe their symptoms[15,16]. Vertebrobasilar insufficiency, especially with basilar invagination, may cause tinnitus, vertigo, and loss of equilibrium, visual disturbance and dysphagia[17].

L'Hermitte sign is provoked by flexing the head, and is associated with a palpable subluxation ("clunk test")[18]. Marks and Sharp[19] reported an average delay of 31 days between onset of neurological signs and diagnosis of myelopathy hence high index of suspicion is of great assistance.

Early signs of myelopathy include clumsiness of the hands, gait disturbances and heaviness of the lower limbs. Patients who were initially ambulatory and become wheelchair bound should raise a suspicion of cervical spine involvement. Physical examination demonstrates weakness, spasticity and the presence of pathological reflexes[12]. Less commonly patients presents with features of vertebra-Basilar insufficiency which are variable.

Imaging

Aim of imaging is to help identify patients at risk of neurological injury and define cervical deformity and instability. Although progressive radiological deformities are seen in 43-86% of patients with rheumatoid arthritis, only 7-34% have neurological deficits[12]. Plain radiograph forms the basis of initial evaluation of cervical involvement.

Plain radiograph

Screening X-rays include anterior-posterior, lateral, open mouth odontoid, dynamic flexion and extension lateral views. Overall bony alignment, degree of osteopenia and soft tissue shadows should be assessed. Particular attention should be paid to the anterior and posterior Atlanto-dental intervals (AADI and PADI, respectively), the amount of superior migration of the tip of the odontoid and degree of sub axial subluxation. Routine evaluation of the integrity of the atlanto-axial complex is best evaluated on the lateral flexion-extension radiographs[12]. The classic measurements of AAS are seen on plain radiograph.

Anterior Atlanto-Dental Interval (AADI); is the distance from the posterior border of the anterior tubercle to the dens is less than 3mm in adults. It is measured on lateral radiograph. Greater than 8mm interval suggests rupture of the alar and transverse ligaments and is an indication for surgery[20].

Posterior Atlanto-Dental interval; evaluates the maximum amount of space available for the upper cervical spinal cord. It is a better predictor of spinal cord injury compared to AADI. It represents anterior-posterior diameter of the spinal canal at that level <14mm results in cord compression[20].

Computed tomography

Best modality for assessing bone anatomy is Multiplanar CT with 3D reconstruction. The reformatted sagittal CT scan can precisely document the position of the odontoid with respect to the foramen magnum, the degree of atlanto-axial dislocation, and the relationships among the upper cervical spine joints[21]. CT also allows for accurate visualization of bony erosions, ankylosis, pseudarthrosis, and vertebral collapse, for planning best surgical technique and implant size. CT angiography is significant in evaluating vertebral artery anatomy[21].

Magnetic resonance imaging

Is the modality of choice for early cervical involvement and cord compression in RA because of excellent soft tissue details. It is important in assessing bony anatomy, the periodontoid pannus, brainstem, spinal cord and relationship of the odontoid to the foramen magnum. Pre/post contrast cervical MRI must be done in all patients. MRI can also be used to predict prognosis. T1-weighted spinal cord signal changes are associated with poor clinical status and also poor final postoperative outcome[21]. MRI is also important in assessing cervicomedullary angle. A normal cervicomedullary angle is between 135-175 degrees and an angle below 135 degrees is associated with myelopathy[12].

Modality	Advantages	Disadvantages
Plain radiograph	Lower cost & widely available Screening of asymptomatic patients Low radiation dose Good for evaluation spinal alignment Flexion & extension allow visualization of occult instabilities	Poor anatomical detail, especially at craniocervical & cervicothoracic junction Poor soft tissue visualization Poor visualization of bone erosions
CT Scan	Multiplanar widely available Gold standard for bone evaluation Good for evaluation of ankylosis & pseudarthrosis Useful for surgical planning Flexion/extension for occult injuries	Higher cost compared to plain radiographs Higher dose of radiation (relative contraindication during pregnancy) Risks w/ intravenous injection of iodinated contrast Requires sedation for young or claustrophobic patients Poor evaluation of soft tissues & spinal cord
MRI	Gold standard for soft tissue & spinal cord evaluation Most sensitive & specific for cervical instabilities Flexion & extension allow visualization of occult instabilities Best for evaluation of patients with neurological deficits	Highest cost of all imaging modalities Requires sedation for young or claustrophobic patients Risk w/ intravenous injection of gadolinium, especially in patients w/ kidney diseases (nephrogenic systemic brosis) May be contraindicated in patients w/ implanted pacemakers, stimulators etc.

Management: non operative management

Patients education on the effects of rheumatoid arthritis on the cervical spine and myelopathy during the first encounter with spine surgeon. Disease Modifying Antirheumatic. Drugs (DMARDs) have been shown to reduce the progression of cervical disease hence important to emphasize role of drug compliance[22,23].

Operative management

Indications for surgery: Clear surgical indications for patients with rheumatoid arthritis include persistent intractable pain and/or neurological deficits. Cervical collars do not prevent progression of cervical instability[24].

Spinal cord compression is difficult to assess in patients with joint deformities and distal muscle weakness due to the disease hence progressive neurological compromise is an indication for surgery[25]. Patients with radiological instability with minimal neurological deficit and pain provide a big challenge in management. Radiological evidence of cervical instability is common, but not all patients are at risk of neurological injury[12].

Surgical outcomes: Improved pre and post operative management, newer technologies with well chosen surgical patients has resulted in improved outcomes [26,27]. Mizutani *et al*[28] demonstrated that with appropriate selection, operative intervention can still provide good pain relief, preserve daily activities and improve the quality of life in the elderly patient with rheumatoid arthritis. Clinical success rate for cervical fusion ranges from 60-100% due to variability of the patient population, disease severity at surgical time and surgical technique familiarity[12]. Onset of myelopathy results in increase in long term mortality and reduction in neurological recovery. The perioperative mortality for Ranawat 3B patients is 12.5% with the reported 1-year mortality rate after surgery approaching 61%[12].

Common complications of surgery include infection, wound dehiscence, los of reduction, instrumentation failure, mal-union and late subluxation below a fused segment[29].

Ranawat's classification of neurological deficit[26]

Class	Sign
1	Pain, no neurological deficit
2	Subjective weakness, hyperreflexia, dysesthesias
3	Objective weakness, long-tract signs present
3A	Ambulatory
3B	Non-Ambulatory

Conclusions

Atlanto-axial instability in rheumatoid patients is very common, highly progressive with devastating outcomes. Natural history of the disease results in increased morbidity and mortality. Myelopathy is a poor prognostic factor in above patients with poor survival outcome. Early diagnosis and treatment has been shown to correlate with good outcomes.

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