



Rheumatologic Causes of Neck Pain

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German Valdez

Introduction

Rheumatic diseases are difficult to define and categorize, but generally are viewed as systemic autoimmune and inflammatory conditions which affect organs, bones, joints, and muscles. In the United States, there are approximately 11 million adults who suffer from a rheumatic disease [1] (Rheum.org).

In this chapter we will present various rheumatological diseases which may present with neck pain. Osteoarthritis is often categorized under rheumatic disease; for the purpose of this chapter, osteoarthritis will be excluded, and rather we will focus on autoimmune inflammatory conditions and/or diseases typically treated by rheumatologists.

Rheumatoid Arthritis

Rheumatoid arthritis (RA) is a systemic autoimmune disease characterized by chronic inflammatory synovitis. RA is the most common rheumatologic condition affecting the neck or cervical

G. Valdez (✉)

Department of Rehabilitation and Human Performance, Icahn School of Medicine at Mount Sinai, New York, NY, USA

e-mail: German.Valdez@mountsinai.org

spine [2] (**Krauss**). RA affects 1–2% of the US population and is notably three times more prevalent in women. Among all RA patients, 17–84% of patients have cervical spine disease [3] (**Kim D.H**).

Pathophysiology

Rheumatoid arthritis is believed to cause cervical spine disease when the inflammatory process extends into the neurocentral joints resulting in ligament rupture, apophyseal joint erosions, disc herniation with subsequent instability, and subluxation [4] (**Neo M**). The most commonly affected joint is the occipitotlantoaxial junction. Atlantoaxial subluxation is a common finding, affecting nearly 90% of patients. The more common anterior subluxation is due to transverse ligament destruction, while posterior subluxations are associated with odontoid erosions and fractures.

Late in the disease process, up to 25% of patients develop subaxial subluxation due to the destruction of multiple facet joints, the interspinous ligament, and multiple disco vertebral junctions [3] (**Kim. D.H**).

A feared complication is basilar invagination, a condition in which the skull descends the cervical spine allowing an eroded odontoid to enter the foramen magnum causing compression of the brain stem or cord, or sudden death.

Presentation and Findings

Patients present with neck pain which may radiate to the occipital or temporal region. If there is neurological involvement, symptoms may include motor weakness, sensory impairments, abnormal reflexes, and spasticity.

Symptoms typically develop after 10 years of disease duration. Cervical spine disease correlates with joint erosions, active synovitis, C-reactive protein levels, rheumatoid factor positive, rheumatoid nodules, and age of onset of RA [5] (**Nguyen**).

On physical exam, there may be loss cervical spine lordosis, resistance of passive range of motion. The most common

neurological findings are hyperreflexia, motor weakness, atrophy, spasticity, and gait disorders.

Management

If disease is present, imaging should be obtained every 2–3 years. Early treatment of RA is thought to slow the progression of cervical spinal disease. Prior to procedures requiring anesthesia, patients should also undergo radiographic screening. Anti-inflammatory medications, trigger point injections, and nerve blocks may provide pain relief. Neck strengthening exercises are not helpful. Manipulation of the neck is contraindicated. Surgical indications include intractable pain, instability, myelopathy, or vertebral artery compromise. Most commonly a C1–C2 fusion and occipitocervical fusion is performed.

Ankylosing Spondylitis

Ankylosing spondylitis is a chronic inflammatory disease of the sacroiliac joint leading to ankylosis. Ankylosis refers to severe spinal restriction due to bony or fibrous bridging of the joints. It affects males more than females in a 2:1 ratio [6] (**Feldtkeller**).

Presentation and Findings

Ankylosing spondylitis typically presents before the age of 40 years with the most common symptom being inflammatory back pain. The pain often improves with exercise, is present at night, and improves upon arising. It often does not improve with rest. In the setting of sacroiliac involvement, there is often alternating buttock pain. In the setting of enthesitis of the supraspinatus tendon, Achilles tendon, or intercostal tendon, there may be shoulder pain, heel pain, and costochondritic chest pain, respectively. Non-musculoskeletal symptoms include symptoms from anterior uveitis, psoriasis, inflammatory bowel disease such as

eye pain, visual changes, skin and nail problems, diarrhea, fever, and weight loss [7] (**Elewaut**).

In terms of the neck, similarly to rheumatoid arthritis patients may develop atlantoaxial subluxation leading to neck pain. Patients are thought to be at increased risk due to ligamentous calcification and cervical ossification. Of note, due to secondary osteopenia and osteoporosis, patients are at increased risk of spinal fractures and may develop fractures after minimal trauma.

On exam, patients will commonly have limited range of motion of the spine, SI joint tenderness, peripheral joint synovitis, enthesal tenderness. Laboratory findings include elevated CRP and is also commonly associated with HLA-B27. Imaging will reveal inflammatory sacroiliac joint inflammatory changes such as widening, erosions, sclerosis, or ankylosis [8] (**Huerta-Sil**).

Management

There is typically a long delay between 5 and 10 years between the arrival of symptoms and diagnosis. Management includes exercise, physical therapy, NSAIDS, and tumor necrosis factor (TNF) antagonists.

Diffuse Idiopathic Skeletal Hyperostosis

Diffuse idiopathic skeletal hyperostosis (DISH) is a condition characterized by calcification and ossification of spinal ligaments and entheses. The incidence of DISH increases with age and is more commonly seen in men [9] (**Belanger**). The pathogenesis of DISH is currently not well understood.

Presentation and Findings

Patients with DISH most commonly present with thoracic back pain, but may also present with neck pain, low back pain, or extremity pain. Roughly 80% of patients will present with

morning back stiffness. Involvement of the cervical spine often presents with dysphagia, but may also present with hoarseness, stridor, aspiration pneumonia, sleep apnea, atlantoaxial subluxation, or thoracic outlet syndrome [10] (**Mader**). More serious complications such as spinal cord compression may arise when there is involvement of the posterior longitudinal ligament.

On physical exam, patients commonly have decreased range of motion of thoracic lateral flexion, along with tenderness and/or palpable nodules over entheses. The palpable nodules are typically found over the calcaneus, olecranon, and patella.

Diagnosis of DISH is made via radiographic imaging. Hallmark findings include ossification of the paravertebral ligament and peripheral entheses. In addition, imaging will often reveal linear calcification and ossification along the anterolateral aspect of vertebral bodies [11] (**Forestier**).

Management

Treatment is aimed at symptomatic relief and maintaining function. Pain is often addressed with Acetaminophen or NSAIDs while function is addressed with range of motion and stretching exercises [12] (**Al-Herz**). Surgery may be warranted to remove bony spurs leading to the more severe complications such as dysphagia and myelopathy.

Myositis

Dermatomyositis and polymyositis are both inflammatory myopathies with a prevalence of 1 per 100,000 and is more commonly seen in woman in a 2:1 ratio.

Presentation and Findings

Patients typically present with gradually worsening proximal muscle weakness. Commonly including muscles of the trunk,

shoulders and upper arms, thighs, and neck extensors. Due to the weakness patients will often report difficulty brushing their hair, rising from chair, and walking uphill [13] (**Harris Love**). Patients may also present with myalgias, which may include the neck. In dermatomyositis patients also present with distinct skin rashes. Laboratory findings include an elevated creatine kinase.

In both the conditions, physical exam may reveal mechanic's hand; i.e., thickened cracked skin at the tips and lateral aspect of the fingers and palm [14] (**Sunkureddi**).

In dermatomyositis, physical exam may reveal as a violaceous or erythematous scaly rash over extensor surfaces of the elbows, knees, and the MCP and IP joints, also known as Gottron papules. Skin findings also include a heliotrope rash with periorbital edema, and the shawl sign, a diffuse erythematous rash over the chest and shoulder in a shawl-like distribution.

Laboratory findings include elevated muscle enzymes such as creatine kinase, lactate dehydrogenase, aldolase, aspartate aminotransferase, alanine aminotransferase. Other abnormal labs which may be found are anti-Sm, SS-A, SS-B, antiribonucleoprotein, and anti-Jo-1.

Management

Treatment includes corticosteroids and other immunosuppressive agents such as methotrexate, azathioprine, cyclosporine, cyclophosphamide, rituximab, mycophenolate mofetil, rituximab, TNF antagonist, and high-dose IVIG.

Fibromyalgia

Fibromyalgia is described as a nonprogressive diffuse pain syndrome of unknown cause associated with multiple tender points. Fibromyalgia affects 1–4% of the population, with roughly 75% of patients being female [15] (**Weir**).

Presentation and Findings

Patients often present with diffuse body pain including the neck and the bilateral upper and lower extremities. Pain is often migratory and waxes and wanes in intensity. Along with pain, patients commonly present with fatigue and sleep disturbances [16] (Bellato).

On physical exam, patients will have 11 of 18 tender points as described by the American College of Rheumatology, otherwise the musculoskeletal and neurological physical exams are unremarkable.

Management

Treatment of fibromyalgia includes education of the condition, aerobic exercise, cognitive behavioral therapy, stress management, optimizing sleep hygiene, and pharmacologic therapy. Psychotropic agents such as tricyclic antidepressants, serotonin reuptake inhibitors, and norepinephrine serotonin reuptake inhibitors have all shown to be efficacious. Other efficacious agents include anticonvulsants, gabapentin, and pre-gabalin.

Conclusion

Although not as common as conditions previously discussed in this book, systemic rheumatological conditions may also be responsible for neck pain. Clinical suspicion for these illnesses is critical as the correct diagnosis will dictate proper management.

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