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CASE REPORT

Temporomandibular Joint Disorder and Anterior Scleritis as Initial Presentations of Undiagnosed Rheumatoid Arthritis

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Abstract

Rheumatoid arthritis is an autoimmune inflammatory process that involves multiple organ systems. While symmetric joint swelling of the extremities are the most widely recognized symptoms, the disease can present in a myriad of different ways, of which scleritis and temporomandibular involvement are less recognized manifestations. While scleritis and temporomandibular disorder (TMD) may at times present in isolation, it is critical to consider their strong association with autoimmune disease as it allows for early diagnosis of inflammatory conditions and allow for the formulation of tailored treatment plans to halt their progression.

Keywords: Rheumatoid arthritis, RA, Scleritis, TMJ, Temporomandibular, Temporomandibular joint disorder, Autoimmune

1. Introduction

Ocular involvement of rheumatoid arthritis in particular has a variety of different presentations. Scleritis is inflammation of the white coating of eye, beneath the layers of the conjunctiva and episclera. It can be seen in a smaller subset of patients with rheumatoid arthritis. If left untreated, the late stages of the disease can progress to necrotizing scleritis which may require surgical intervention.

The joints involved in rheumatoid arthritis are more commonly of the hands, wrists, and knees. The temporomandibular joints are less often involved.[1] However, it can lead to irreversible destruction of bone if left untreated in the setting of rheumatoid arthritis.

Both scleritis and temporomandibular joint disorder are initially treated conservatively, with non steroidal anti-inflammatory agents (NSAIDs) being the first line pharmacotherapy. However, it is important to consider autoimmune drivers such as Rheumatoid Arthritis in these patients[2] as it confers a worse prognosis due to resistance to NSAIDs and often require systemic glucocorticoids.

2. Case presentation

A 50 year old Hispanic female with no chronic conditions or family history of autoimmune disease presents with 8 weeks of left eye pain and redness in addition to jaw pain worse on the left side with associated ear pain and fullness. She also endorsed diffuse joint pain most prominent in her bilateral hands and wrists that began a few weeks after her ocular and jaw symptoms. Her history was negative for any preceding viral, diarrheal, or genitourinary infections.

On exam, she had scleral injection with lid edema of the left eye (Fig. 1). She also had symmetric synovitis of her wrists and hands with associated erosions on x-ray (Fig. 2). No saddle nose deformity, rashes, or neurologic deficits were present.

Labs revealed elevated inflammatory markers of ESR 115 mm/h (reference range 0–30 mm/h) and

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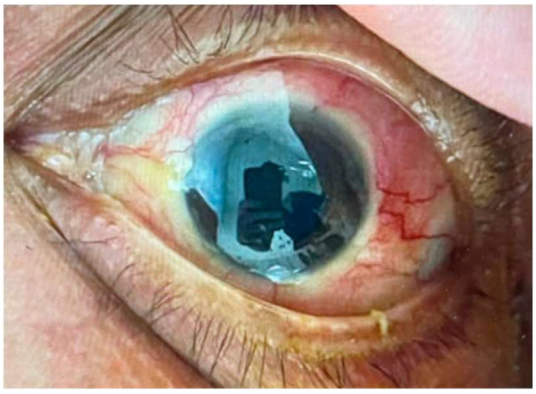

Fig. 1. Scleral injection with lid edema.

high sensitivity CRP 20 mg/dl (reference range 0–1 mg/dl). She also had significant elevations of her rheumatoid factor at 520 IU/ml (reference range 0–14 IU/ml) and anti cyclic citrullinated peptide (CCP) antibody at 2197 CU (reference range <20 CU). Her antinuclear antibodies (ANA), Anti-neutrophilic cytoplasmic antibody (ANCA), double strand DNA antibodies (anti ds-DNA), and human leukocyte antigen B27 (HLA B27) were negative.

She was evaluated by ophthalmology and was diagnosed with anterior scleritis. Her limited jaw movement in association with pain and ear fullness suggested temporomandibular involvement of undiagnosed rheumatoid arthritis.

Her differential initially included Giant Cell Arteritis (GCA) in the setting of ocular and jaw involvement with elevated inflammatory markers.

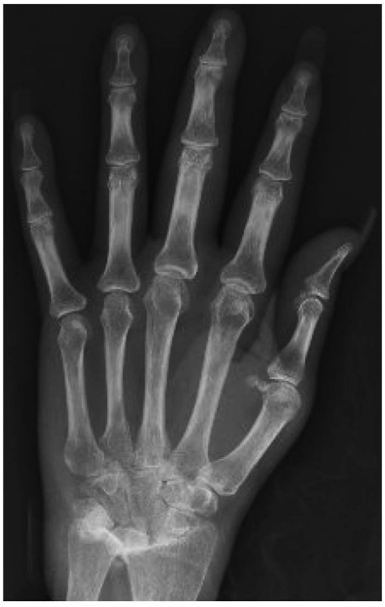

Fig. 2. Bony erosions with periarticular osteopenia.

However epidemiology did not fit her profile as GCA has a peak incidence at age 70–80 and more common in Caucasians. Furthermore her ocular pain and ophthalmologic exam suggested scleritis, as opposed to the painless vision loss secondary to ischemia and inflammation seen in GCA. Systemic Lupus Erythematosus (SLE) was also considered but less likely given her negative ANA, anti ds-DNA, and the symmetric non migratory distribution of her joint pain, which differs from the asymmetric migratory joint pain seen in SLE. Finally, Reactive Arthritis was also less likely with no signs of current or preceding genitourinary or diarrheal infection, and with her negative HLA B27.

She was started on NSAIDs as needed and prednisone 30 mg daily for her arthritic pain, constitutional symptoms, and anterior scleritis and experienced rapid improvement supporting an inflammatory etiology. She was discharged with outpatient Rheumatology follow up to taper her steroids and transition to disease-modifying anti-rheumatic drug (DMARD) therapy.

3. Discussion

Peripheral joint arthritis is the most common presentation of rheumatoid arthritis. It is uncommon for TMD or scleritis to be the initial presentation, with less than 10% of patients having these symptoms on disease onset. The most prevalent ocular manifestation of rheumatoid arthritis involves the cornea as keratoconjunctivitis, which can occur in about 18% of patients with rheumatoid arthritis, primarily in the setting of concomitant Sjogren Syndrome.[3] Scleritis, on the other hand, is inflammation of the white tissue layer which protects the rest of the eye. It has a rarer occurrence of 0.7–6.3%.[4] What helps distinguish scleritis from other pathology in the anterior chamber is the constant and severe discomfort, commonly described as a “boring” pain. This is in contrast to conjunctivitis which causes sharp and superficial irritation.[5] It is important to recognize scleritis early, which when untreated, leads to severe scleral thinning and necrosis that can cause ocular perforation.[6]

Scleritis secondary to rheumatoid arthritis has shown clinical response to oral NSAIDs as first line pharmacotherapy. However, recurrence and treatment failure to NSAIDs is common, after which oral steroids, and DMARDs can be used.[7] The prolonged use of these systemic agents however is challenging as steroids and agents such as hydroxychloroquine themselves have adverse ocular effects with cataracts and retinal pigment deposition, respectively.

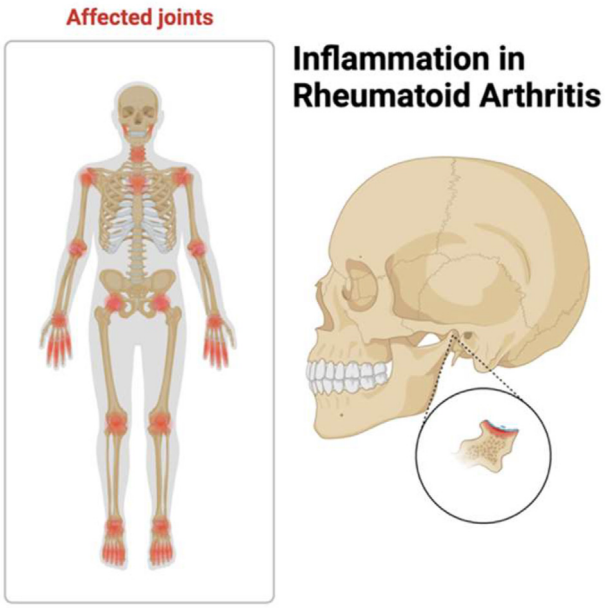

Fig. 3. Joint involvement in Rheumatoid Arthritis.[10]

There should be a low threshold to pursue expert evaluation with close follow up in these patients with ophthalmological symptomology prior to aggressive therapy.

Central joint involvement, including the cervical atlantoaxial and temporomandibular jaw joints, is less common than peripheral arthritis, affecting 20–50% of patients (Fig. 3). Treatment of TMD begins with conservative approaches of bite splints and physical therapy.[8] However, unlike primary TMD, rheumatoid arthritis associated TMD more often persists, requiring systemic NSAIDs as it is an inflammation mediated process. Similar to scleritis, treatment often requires escalation to corticosteroids, DMARDs, and biologic medications, including tumor necrosis factor-alpha inhibitors.[9] In severe cases, maxillofacial surgery is also considered. If untreated the inflammation can damage surrounding cartilaginous tissue of the jaw, leading to chronic pain, airway sleep disorders, and tinnitus.

4. Conclusion

Scleritis and TMD are less recognized but important presentations of rheumatoid arthritis. Scleritis in particular has a significant association with systemic disease, with 50% of cases having an association with inflammatory disease with closest link being with rheumatoid arthritis followed by granulomatosis with polyangiitis. All patients with TMD and scleritis warrant evaluation for systemic autoimmune with specialized serological testing guided by thorough physical exams, particularly in populations at risk for autoimmune conditions. Early diagnosis of underlying autoimmune conditions not only facilitates a more targeted treatment for the destructive conditions of TMD and scleritis, but also prevents the overall progression of multi-organ damage that often accompanies rheumatoid arthritis.

Conflict of interest

No conflicts of interest exist for the authors.

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