CLINICAL VIGNETTE

Acronym Battle – RS3PE vs PMR vs RA?

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A 79-year-old male with hypertension, chronic kidney disease and diet controlled type 2 Diabetes Mellitus presented to the rheumatology with several weeks of pain and swelling in upper and lower extremities. The pain had acute onset and he was seen in the past two weeks by two orthopedic surgeons. He was started on gabapentin by the hand surgeon for carpal tunnel symptoms. Patient denied any recent travel, new medications, or sick contacts. Due to constant body pain, he lost appetite with a fourteen pound weight loss over the past month. He reported history of polymyalgia rheumatica diagnosed seven years ago. He recalled several months of steroid taper treatment which was tapered and transitioned to hydroxychloroquine. However, he stopped taking hydroxychloroquine after 1 year, but does not recall why this was stopped. Other significant past medical history includes colon surgery in 1985 for "cancerous polyps", and further colon resection in 2008. He retired from the fire department. He quit smoking 27 years ago. He had no family history of rheumatic disease.

On physical exam, patient had significant swelling with pitting edema of his upper and lower distal extremities. There was limited range of motion in wrists, bilateral metacarpophalangeal (MCP) joints, proximal interphalangeal (PIP) joints, ankles, and feet. He had no lymphadenopathy, tenderness over the lower back and sacroiliac joints, and features of enthesitis. The rest of the clinical examinations including cardiovascular, respiratory, abdominal, and neurological examinations were unremarkable. Laboratory evaluations revealed significantly elevated sedimentation rate of 103 mm/hr and elevated C-reactive protein of 8.0 mg/dL (normal range of less than 0.8). Serological markers for rheumatoid arthritis, lupus, and other autoimmune and rheumatic conditions were all normal. Recent thyroid stimulating hormone was normal, and hemoglobin A1C remained elevated at 7.5.

The patient was diagnosed with **R**emitting Seronegative **S**ymmetrical **S**ynovitis with **P**itting **E**dema ("RS3PE" Syndrome), and started on prednisone 20mg daily. At one week follow up, he reported significant improvement in swelling and range of motion of the distal extremities. An MRI of the hand did not show evidence of joint or tendon changes. After one month of prednisone 20mg daily, tapering started along with addition of steroid sparing therapy with hydroxychloroquine. Currently the patient is off all steroids with complete resolution of joint swelling. His nerve conduction study prior to rheumatology consultation was consistent with bilateral carpal tunnel syndrome and he underwent right carpal tunnel release four months after diagnosis of RS3PE.

Remitting Seronegative **S**ymmetrical **S**ynovitis with **P**itting **E**dema (RS3PE) is a rare rheumatological condition that is also known as puffy edematous hand syndrome or boxing glove syndrome. It is characterized by symmetric tenosynovitis of the upper and lower extremities, especially most obvious in dorsa of the hands.[1] Range of motion is limited in hands and wrists, but can extend up to the shoulders. Laboratory evaluation reveals extremely high inflammatory markers, with negative assays for rheumatoid arthritis and other rheumatic subserologies. Imaging can be done, with ultrasound and MRI being the best modalities. Some of the findings on imaging include extensor tenosynovitis of the forearms and hands, usually sparing flexor tendons and joints of the hands. RS3PE is very responsive to low dose steroids, non-steroidal anti-inflammatories, and hydroxychloroquine. The condition was first described in 1985, with a two to one predominance for men over the age of 60 years old.[2] There are associated co-morbidities such as type 2 diabetes mellitus or impaired glucose tolerance, and rare cases of association of RS3PE with certain diabetes medications. There has been discussion of RS3PE as a paraneoplastic syndrome, in particular for certain solid tumor malignancies and hematologic malignancies. These patients usually do not respond well to the conventional therapies for RS3PE.

RS3PE was initially thought to be a subset of rheumatoid arthritis, but now characterized separately.[3] A few factors differentiate from rheumatoid arthritis, including pitting edema (not usually present in RA), negative serologic antibodies (rheumatoid factor and anti-CCP Ab), and only partial response to low dose prednisone. Another common differential diagnosis is another rheumatological acronym: "PMR" (Polymyalgia rheumatica). This patient had a prior diagnosis of PMR several years ago, while living outside of California. Records were not attainable. In PMR, swollen joints are rare but inflammatory markers are exquisitely elevated as in RS3PE. Both conditions respond well to low dose prednisone, but it is rare to use steroid sparing agents such as hydroxychloroquine in PMR. The patient had been given hydroxychloroquine 7 years ago, so this raises the possibility of seronegative rheumatoid arthritis. RS3PE, PMR, and RA, have significant overlap in the symptoms, objective findings, and treatment. However, R33PE has different associations and treatments.

REFERENCES

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