

CLINICAL VIGNETTE

Polymyalgia Rheumatica and Giant Cell Arteritis in a Patient with Lower Extremity Weakness

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Case Report

A 74-year-old woman with hyperlipidemia presented to the emergency room (ER) with bilateral lower extremity pain, stiffness, and weakness. Symptoms were present for two months and acutely worsened three weeks prior to presentation. Her pain and weakness were present in her bilateral proximal quadriceps muscles with burning pain and without radiation. She also reported her muscles in this region had atrophied. She was previously physically active and exercised daily. Unfortunately, ambulating was now difficult secondary to ongoing pain, weakness, and leg stiffness. She was concerned about Lyme Disease as prior to symptom onset she would hike through wooded areas. Additional questioning revealed subjective fevers, a dry cough, night sweats and occasional jaw pain which she attributed to temporomandibular joint (TMJ) dysfunction. She denied unintentional weight loss.

Upon presentation to the ER, her vitals showed a temperature of 36.5°C, a heart rate of 63 beats per minute, respiratory rate of 18 breaths per minute, blood pressure of 95/47 mmHg and oxygen saturation of 94% on room air. Her physical exam was notable for appearing fatigued, right lower extremity 1+ pitting edema and left lower extremity trace edema with 5/5 motor strength throughout. She had a slow, steady gait. Laboratory results were notable for an elevated d-dimer, C-reactive protein (CRP) in the 200 mg/dL range (N<0.3 mg/dl) with a normal lactate level. Bilateral venous doppler ultrasound of her lower extremities was negative for deep venous thrombosis (DVT). Her case was discussed with Neurology who recommended admission for magnetic resonance imaging (MRI) of her lumbar spine and pelvis.

MRI Lumbar spine showed degenerative changes at L5-S1 with moderate to severe right foraminal stenosis impinging the right L5 nerve root. MRI Pelvis was unremarkable. Additional laboratory testing was unremarkable for creatine kinase (CK), angiotensin-converting enzyme (ACE) level, ANCA levels, cytoplasmic neutrophil antibody, thyroid stimulating hormone (TSH), procalcitonin, ammonia, vitamin B12, folate, anti-neutrophil antibody (ANA), Rheumatoid Factor (RF) and a Lyme disease panel. A repeat CRP remained in the 200s and her ESR (Erythrocyte sedimentation rate) (N<30 mm/hr) was in the 70s.

The Rheumatology service was consulted as her evaluation far was non-revealing and concern for an underlying rheumatologic

disorder. Additional history included prior left macular pucker (wrinkles or bulges in the macula) with worsening vision in her left eye over the prior three weeks. She also reported a headache with tenderness in her bilateral temporal regions with occasional episodes of trismus. Vascular surgery was consulted for a temporal artery biopsy and she was empirically started on intravenous methylprednisolone. Bilateral temporal artery biopsies were done the following day. Pathology confirmed evidence of healed temporal arteritis. Her symptoms and inflammatory markers improved one day after starting methylprednisolone and she was discharged on a slow, prolonged prednisone taper.

Discussion

GCA is an inflammatory vasculitis that varies in presentation, typically affecting medium to large sized arteries in patients over the age of 50. Common predisposing risk factors include advancing age and Northern European ancestry with the highest incidence observed in those between 70 and 79 years of age with a mean age of onset of 75.^{1,2} GCA is thought to be an immune-mediated disease involving an improper response to vascular endothelial damage activating vascular dendritic cells.¹ Dendritic cells recruit T-cells in the arterial wall which activate macrophages resulting in propagation of vascular smooth muscle cells and formation of giant cells leading to an inflammatory response.¹

Presentations include nonspecific constitutional symptoms, and suspicious headaches, vision changes and jaw claudication. Tongue pain, though rare, significantly increases the possibility of GCA.³ Headaches are usually described as temporal headache, however, can be frontal, occipital or generalized that can worsen or subside.¹ Constitutional symptoms can include depression, anorexia, weight loss, general malaise and fevers up to 39° Celsius in 15% of patients, although more commonly are low grade.^{1,3} Classic jaw claudication includes masseter pain and fatigue with mastication with improvement with cessation.

Visual disturbances can be transient to abrupt partial field defects, which have been described as a curtain descending over one eye to sudden, painless, permanent vision loss in one or both eyes which can be partial or complete.^{1,4} Diplopia can occur prior to vision loss and rarely visual hallucinations may occur.¹

Large vessel involvement includes the aorta and proximal branches. Symptoms include chest pain, back pain, and upper extremity claudication. Less common symptoms include transient ischemic attacks or strokes in the vertebrobasilar system, upper respiratory tract symptoms, pericarditis, dysarthria, and sensorineural hearing loss.¹ Concomitant GCA should be considered when investigating PMR. Forty to sixty percent of patients diagnosed with GCA have documented PMR and 16-21% of patients diagnosed with PMR were reported with GCA.¹ PMR is characterized by pain and stiffness around the neck, shoulders, and hip area more commonly seen in adults over age 50. Similar to GCA, PMR is associated with elevated inflammatory markers including CRP and ESR levels. The etiology is unknown however several studies have reported an association between pneumonias, diverticulitis, and vaccinations.⁵ PMR is found predominately in Caucasian populations and uncommon in Asian, black and Hispanic populations. The pain and stiffness in PMR is symmetric in the shoulders, neck, hips, thighs, upper and lower back. Stiffness is common and worse in the morning and after prolonged inactivity. Similar to GCA, patients may note constitutional symptoms including fatigue, malaise, weakness, anorexia, weight loss and low-grade fevers.⁵ A small percent also report arthritis, carpal tunnel syndrome, distal extremity pitting edema (as seen in our patient) and distal tenosynovitis.⁵ Physical examination may show muscle tenderness and reduced active range of motion secondary to pain in the shoulders, arms and thighs with muscle strength usually intact. Laboratory values are similar in both GCA and PMR, including elevated ESR and CRP. Additional laboratory findings may include anemia, thrombocytosis, elevated liver enzymes and unremarkable ANA, RF and CK levels.

The treatment for GCA and PMR involves glucocorticoids. High dose oral glucocorticoids are used in GCA if there is no evidence of cranial ischemia at the time of presentation. Typical daily dosing is 40 to 60mg of prednisone.² For patients presenting with vision loss or diplopia, 500 to 1000mg of intravenous (IV) methylprednisolone is given for three days followed by high dose oral glucocorticoids.¹ High dose glucocorticoids are continued until symptoms resolve and inflammatory markers normalize, generally from two to four weeks.² After improvement, a prolonged taper is initiated with the goal of getting patients off glucocorticoids over 12 to 18 months.^{1,2} Tocilizumab should be considered in patients who do not tolerate glucocorticoids or those who relapse.¹ For PMR, the doses of glucocorticoids are lower, starting at a range of 12.5 to 25mg of daily prednisone. A slow, gradual taper is then begun for a minimum of 12 months.⁶ Glucocorticoid sparing agents include methotrexate, leflunomide and tocilizumab.⁵

Our patient demonstrated findings consistent with both PMR and GCA. These included constitutional symptoms of fevers, dry cough, night sweats and bilateral thigh pain with stiffness in the setting of elevated ESR and CRP levels. Further questioning revealed symptoms concerning for GCA with visual disturbances, headaches, jaw claudication and temporal tenderness. Her physical exam demonstrated pitting edema,

more consistent with PMR. On day two she was started on prednisone and quickly responded with improved energy, and decreased stiffness, pain and inflammatory markers.

GCA and PMR can be seen in predominately Caucasians over age 50. Patients with PMR should be evaluated for GCA given the increased frequency of these conditions coexisting. This patient presented with symptoms concerning for PMR and after further questioning she was also diagnosed with GCA. Treatment for both involves high dose glucocorticoids with excellent prognosis with early diagnosis and treatment.

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