CLINICAL VIGNETTE

Let Not the Curtain Fall

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Case

A female in her mid-70s presented with a several months of rest predominant pain and stiffness in the proximal shoulder and hip girdles. Moderate relief was achieved with over-the-counter non-steroidal anti-inflammatory drugs and stretching. There were no fevers/chills, unexplained weight loss, headache, scalp tenderness, jaw claudication, or visual changes. She was free of new rashes, hot/swollen joints, hair/nail changes, or pertinent bowel symptoms. Examination showed no proximal muscle weakness, periorbital/extremity rashes, or joint synovitis. Autoimmune serological testing included normal anti-nuclear antibody and rheumatoid factor, as well as a persistent low positive anti-cyclic citrullinated peptide. Creatine kinase, aldolase, and thyroid-stimulating hormone were also within normal limits. Sedimentation rate and C-reactive protein were elevated at 39 mm/hr and 1.4 mg/dL.

The patient was diagnosed with polymyalgia rheumatica (PMR). Symptomatic remission was achieved with initiation of prednisone 15mg daily and her inflammatory markers normalized. Prednisone was successfully gradually tapered over 8 months until reaching a daily dose of 1mg. While on this dose, the patient noted onset of new symptoms including left-sided headaches and temporal scalp tenderness. Her prednisone was increased to 60mg daily and she was referred for a diagnostic temporal artery biopsy. While awaiting her scheduled biopsy, she developed an episode of visual disruption described as several minutes of flashing lights in the left eye. She was referred to the hospital for intravenous corticosteroids as empiric treatment of vision-threatening Giant Cell Arteritis. Symptoms resolved and she was transitioned to a high dose oral prednisone upon discharge. Subsequent left temporal artery biopsy demonstrated findings consistent with Giant Cell Arteritis. Due to osteopenia, steroid-sparing strategies were discussed and she was switched to tocilizumab therapy.

Discussion

Giant cell arteritis (GCA) – also known as temporal arteritis – is the most common primary systemic vasculitis. It is typically found in adults over the age 50 with incidence peaking in the 70s.[1] Unlike the other systemic vasculitides, GCA has a unique association with polymyalgia rheumatica (PMR). 16-21% of patients with PMR develop GCA over time and 40-60% of patients with GCA have PMR symptoms at time of diagnosis.[2] As illustrated in our case, patients who have been treated or who are undergoing appropriate treatment for PMR are still at risk for developing GCA. Therefore, all members of the clinical team should remain vigilant in screening patients with PMR for symptoms of this potentially life changing diagnosis.

Clinical manifestations of GCA can be protean as with all systemic vasculitides. Constitutional symptoms include fatigue, malaise, night sweats, fevers, and unexplained weight loss.[1] The most defining features of GCA, however, include a constellation of primarily intracranial symptoms, although extra-cranial vessel involvement has been well-described. These include headaches, scalp tenderness, and jaw claudication.[3] The frequency of headache approaches 90%, highlighting the importance of physician awareness when caring for patients with increased risk for GCA.[1] Scalp tenderness frequency is 3-50% and jaw claudication is 40-50%.[1] Up to 20-30% of patients with GCA experience visual disturbances. Some may progress to permanent blindness without early recognition and prompt initiation of high dose steroids.[2] Visual disturbances manifest differently between patients. Some experience visual field deficits described as a falling curtain while others present with blurred vision or amaurosis fugax.[2,4] Less common manifestations include audiovestibular symptoms (hearing loss, tinnitus, vertigo, etc.), hoarseness, and neurological deficits.[1] Extra-cranial vessel involvement can lead to loss of pulses and limb claudication.[3]

The mainstay of treatment remains immunosuppression with initial use of high dose corticosteroids. More recent guidelines support early utilization of a biologic medication, tocilizumab.[3] Tocilizumab is given once weekly as a subcutaneous injection and continued after successful tapering of oral corticosteroids. Collaboration with all providers of the multidisciplinary care team is key to monitoring symptoms of disease activity and treatment related adverse effects.

In conclusion, regardless of treatment status all patients with active or former PMR should have regular screenings for symptoms of GCA. Awareness and a high degree of clinical suspicion amongst all healthcare providers, including non-rheumatologists, may help prevent complications and disability associated with delayed recognition of GCA.

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