A Rare Presentation of GCA in the Absence of Classic Symptoms: A Case Report

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Introduction

Giant Cell Arteritis (GCA) is a medium and large-vessel vasculitis affecting individuals over 50 years old. It classically presents with headache, scalp tenderness, jaw claudication, and visual symptoms. However, 20–25% of patients may present atypically, including with systemic symptoms or isolated polymyalgia rheumatica (PMR) like complaints. Delay in diagnosis can result in irreversible complications, including permanent vision loss. Prompt recognition and initiation of glucocorticoids are essential.

Case Presentation

A 60-year-old male with hypertension presented with 4 weeks of burning symmetric proximal bilateral upper and lower extremity pain after being discharged from an outside hospital where he underwent significant workup for suspicion for acute infection given his leukocytosis and thrombocytosis, including negative spinal tap, spinal, and brain MRI. All infectious workup including blood cultures and a spinal tap were negative. At this time, thrombocytosis and leukocytosis were thought to be reactive in the setting of negative peripheral smear. The patient was started on 20mg of prednisone due to concern for underlying rheumatologic process with no improvement and was discharged. Due to persistence of symptoms, the patient presented again to the hospital for further workup. The patient had no recent illnesses, fever, weight loss, rashes, headache, scalp tenderness, and jaw claudication; however did state that 3 hours before arrival, he had a 60-second transient "film" go over his right eye which was a new finding. Lab work was significant for WBC 30.1 with neutrophilic predominance, platelets 877, ESR > 120, CRP 26.5. Due to suspicion for GCA, the patient was initiated on 500 mg IV methylprednisolone. The patient subsequently underwent temporal artery biopsy, which showed transmural inflammation compatible with temporal arteritis.

Patient was started on prednisone 60mg/day with improvement in myalgia however did have recurrent episode of transient vision loss prompting initiation of Actemra with resolution of symptoms.

Discussion

- At Polymyalgia Rheumatica diagnosis, Giant Cell Arteritis is found in about 22% of patients; with a similar percentage of PMR patients going on to develop GCA. GCA classically presents with cranial symptoms; although 25% of patients with confirmed GCA can present solely with transient vision loss with approximately 48% developing permanent blindness if untreated.
- Although cranial symptoms such as headache, scalp tenderness, and jaw claudication are the classic hallmarks of GCA, up to 1/4th of patient may lack these symptoms and present with nonspecific constitutional or musculoskeletal symptoms complicating diagnosis and leading to initial misclassification as infection, malignancy, etc.
- Temporal artery biopsy remains the gold standard for diagnosis although color coded duplex sonography, MRI, and PET-CT are being utilized more frequently for initial evaluation.
- High dose corticosteroids remain first line treatment for patients with suspicion for GCA to rapidly reduce arterial inflammation. Emerging therapies, including tocilizumab (Actemra) have shown promise in reducing relapse rates and sparing steroid exposure

Figure

	At time of Diagnosis	4 weeks Prednisone 60mg/day	4 weeks Prednisone 40mg/day + Actemra
ESR	>120	20 mm/hr	4 mm/hr
CRP	26.5 mg/dl	0.3 mg/dl	0.2 mg/dl
WBC	30.1 K/uL	17.4 K/uL	15.6 K/uL
Platelets	877 K/uL	287 K/uL	217 K/uL

Figure 1: Trend of ESR, CRP, WBC, and Platelet count after initiation of steroids and Actemra

Conclusion

- Clinicians should have a high index of suspicion for GCA in patients over 50 with elevated inflammatory markers presenting with symmetrical polyarthralgia in the absence of classic cranial symptoms of Giant Cell Arteritis.
- Early initiation of high-dose glucocorticoids is crucial to prevent permanent vision loss
- Tocilizumab (Actemra) is recommended for patients with relapsing or refractory GCA or in those who are unable to tolerate high dose steroids for a prolonged period of time.

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