An Unusual Presentation of Granulomatosis with Polyangiitis (GPA): A Case Report

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Introduction: Granulomatosis with polyangiitis (GPA) is a small vessel necrotizing vasculitis that involves the upper and lower respiratory tracts and the kidneys. Patients commonly present with symptoms such as sinusitis, otitis media, hemoptysis, and features of glomerulonephritis, such as microscopic hematuria and renal dysfunction1. In this report, we describe an unusual case of GPA, with the patient initially presenting with fever and jaw claudication, which are more commonly associated with other vasculitides like giant cell arteritis (GCA), Takayasu arteritis, and polyarteritis nodosa.

Case Presentation: A 66-year-old Middle Eastern male was admitted to the hospital with a 2-week history of bilateral severe headache, jaw claudication, blurry vision, sinus congestion and feverish sensation. Medical history was significant for essential hypertension and type 2 diabetes. Sedimentation rate (ESR) was 83. The patient's symptoms were initially suspicious of giant cell arteritis (GCA); however, temporal artery biopsy was normal. CT scan of the sinuses showed complete opacification of the left frontal and left maxillary sinuses (image 1). The patient's urinalysis revealed microscopic hematuria with proteinuria and protein creatinine ratio was elevated at 1.5. Labs further revealed elevated ESR, positive C-ANCA, and proteinase-3 antibodies. A kidney biopsy showed evidence of focal segmental pauci-immune glomerulonephritis. Overall, kidney biopsy findings along with CT scan results align most consistently with GPA. The patient was started on Rituximab infusions and prednisone. On follow up visits, the patient has had complete resolution of his sinus congestion, jaw claudication, and blurry vision with decreasing ANCA and PR3 titers, and resolution of proteinuria.

Discussion: This case demonstrates an unusual presentation of GPA with the patient initially presenting with features of headache, blurry vision, and jaw claudication that mimics presenting symptoms of other vasculitides. It is important to highlight unique cases such as this one to increase awareness to all primary care physicians, including rheumatologists, of how GPA may present to prevent delays that could affect patient outcomes.

