https://doi.org/10.33805/2694-2216.101
Volume 1 Issue 1 | PDF 101 | Pages 2

EDELWEISS PUBLICATIONS
OPEN ACCESS

Rheumatic Diseases and Treatment Journal

Case Report ISSN: 2694-2216

Delayed Radiographic Presentation of IgG4 Vasculitis

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Citation: Osman N and Morris SJ. Delayed radiographic presentation of IgG4 vasculitis (2019) Rheum dis treatment J 1: 1-2.

Received: Oct 04, 2019
Accepted: Nov 02, 2019
Published: Nov 15, 2019

Key Words: Hepatitis, Immunoglobulin, Joint Pain, Vasculitis, lymphoproliferative disorder.

Abbreviations: Ig- Immunoglobulin, IFE- Immunofixation, C-ANCA- Cytoplasmic Anti-Neutrophil Cytoplasmic Antibodies, p-ANCA- Perinuclear Anti-Neutrophil Cytoplasmic Antibodies, UPEP- The Urine Protein Electrophoresis, SPEP- The Serum Protein Electrophoresis, CT- Computed Tomography, EGD- Esophagogastroduodenoscopy, NSTEMI- Non-ST Segment Elevation Myocardial Infarction, HBV-*Hepatitis B Virus*, PAN- Polyarteritis Nodosa, Positron-Emission Tomography Computed Tomography Angiography- PET-CTA.

Immunoglobulin G4 (IgG4) related disease is a systemic inflammatory process that affects multiple organs and can commonly present with large vessel vasculitis. We present an interesting case of a patient with delayed manifestation of IgG4 related vasculitis on radiographic imaging.

A 76 year old male with medical history notable for Transient Ischemic Attack (TIA), diastolic heart failure, and past resolved hepatitis B infection, was admitted in January 2018 for a three month history of severe left upper and lower quadrant abdominal pain, intermittent nightly fevers, and fifty pound weight loss due to pain with oral food intake. He denied any personal or family history of autoimmune disease. He denied having joint pains, myalgias, muscle weakness, rashes, SICCA symptoms, or skin tightening. Serologies were notable for elevated rheumatoid factor to 216.0 (normal <14 IU/mL), ESR of 85 (age adjusted normal <38 mm/hr), and CRP of 10.6 (normal 0-0.4 mg/dL).

The patient had paraproteinemia with total protein level of 8.4 g/dL on admission, and albumin of 2.1 g/dL. Serum IgG4 was significantly elevated to 1760.0 mg/dL (normal 2.4-121.0 mg/dL). Quantitative IgG was 3440 mg/dL and IgM 290 mg/dL, and Immunofixation (IFE) kappa and lambda were both elevated to 25.30 mg/dL and 20.40 mg/dL, respectively. Antinuclear Antibody (ANA) was 1:80, speckled; Cytoplasmic Anti-Neutrophil Cytoplasmic Antibodies (C-ANCA) and Perinuclear Anti-Neutrophil Cytoplasmic Antibodies (p-ANCA) were negative. Given the patient's paraproteinemia and weight loss, malignancy was considered. The Urine Protein Electrophoresis (UPEP) showed no monoclonal proteins but immunofixation and The Serum Protein Electrophoresis (SPEP) detected an oligoclonal gammopathy.
Our patient underwent a Computed Tomography (CT) abdomen and pelvis which showed questionable mild colitis and no bowel obstruction or other pathology.

Esophagogastroduodenoscopy (EGD) with colonoscopy during the same admission only revealed sigmoid diverticulosis and otherwise normal appearing colon. A bone marrow biopsy performed inpatient showed no pathology. With negative results on malignancy workup, our patient was discharged home with instructions to follow up at three month intervals to rule out development of a lymphoproliferative disorder.

The patient returned to the hospital one year later with worsening dyspnea, found to be in decompensated heart failure and Non-ST Segment Elevation Myocardial Infarction (NSTEMI), with worsening ejection fraction of 10-15%. He underwent a cardiac catheterization which showed diffuse coronary arterial disease. This hospital course was further complicated by persistent abdominal pain for over a month's duration, similar to the pain from the prior hospitalization. At this time, a repeat CT abdomen/pelvis revealed diffuse blood vessel wall thickening of the aorta and its branches-the celiac axis, superior and inferior mesenteric arteries, and bilateral renal and iliac arteries.

The findings were reviewed with radiologists and there was no lymphadenopathy or mass to suggest associated malignancy. The patient was started on IV methylprednisolone 80 mg daily for several days for vasculitis, thought to be IgG4 related vasculitis (likely, given his elevated serum IgG4 levels), polyarteritis nodosa (possibly in relation to patient's prior Hepatitis B Virus (HBV) infection), or a paraneoplastic syndrome. The patient was started on entecavir for HBV prophylaxis while on immunosuppression.

He reported a gradual improvement in his abdominal pain. Steroids were eventually tapered to oral prednisone 80 mg daily, and one rituximab 1 g infusion was initiated for a presumptive diagnosis of IgG4 vasculitis. Due to the patient's symptom improvement he was discharged home on prednisone 60 mg daily, and a plan for outpatient follow up. The patient continued rituximab after discharge, with gradually decreasing IgG4 levels measured at follow up visits.

The differential diagnoses for this patient included not only IgG4 disease but also Polyarteritis Nodosa (PAN), as it is difficult to

distinguish between the two vasculitides. PAN is a common large vessel vasculitis consistent with the CT abdomen findings, and is associated with hepatitis B infection. Unfortunately, the patient expired shortly after this hospitalization and vessel biopsy could not be obtained. Vasculitis can also be associated with solid or hematologic malignancies; however, this patient had more than one normal colonoscopy and bone marrow biopsy result and radiographic imaging did not reveal any tumors or lymphadenopathy.

Our patient demonstrated an interesting delayed manifestation of vasculitis on radiographic imaging. Previous studies have shown the high incidence of vascular involvement in IgG related disease (22.5% in an analysis of 160 patients studied by Perugino et al [1]. The difficult aspect of IgG4 related disease is that it can disturb multiple organ systems, and patients can present in a multitude of ways, from asymptomatic to overt organ involvement. Therefore, in patients with elevated IgG4 and other inflammatory markers, early imaging is recommended. Our patient during his initial hospitalization was evaluated with CT abdomen but a better imaging modality may have been a Positron-Emission Tomography Computed Tomography Angiography (PET-CTA) with and without contrast.

This may have sooner revealed an abnormality of the large and medium vessel walls. This patient was effectively treated with high dose corticosteroids as well as rituximab, an anti-CD20 monoclonal B cell depleting antibody; but earlier treatment may have changed the clinical course had the diagnosis been established sooner. Early diagnosis involves a combination of clinical, serologic, histopathologic, and radiologic findings. As the findings can present in a variable time frame, the mean time to diagnosis for individuals with IgG4 related disease can be five years or more [2].

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