Summary:

A 74-year-old gentleman presented to the primary care physicians office two years earlier with a few weeks of nasal congestion, cough and shortness of breath. He was found to have bilateral wheezing on physical examination. He was treated for asthmatic bronchitis with antibiotics and inhaled corticosteroids and he improved. Over the next one year he had a few similar episodes and was being seen by pulmonologist and allergist and was treated similarly and he got better. A year later he presented back to the PCPs office. A CT scan of the chest was obtained that showed mediastinal adenopathy and bilateral pulmonary embolism. He was started on DOAC and was referred back to the pulmonologist. Over the next few months he developed peripheral eosinophilia, peripheral neuropathy, and skin rash consistent with vasculitis, new onset atrial fibrillation and acute abdominal pain leading to cholecystectomy. Pathology of the gallbladder confirmed the diagnosis.

Case Presentation:

74-year-old white gentleman with history of hypertension presented to the primary care physicians office in 2018 with nasal congestion, cough and shortness of breath of few weeks duration. He had bilateral wheezing on physical exam. He was treated with antibiotics and steroids which temporarily relieved his symptoms. He had a couple more similar episodes during the following months and was treated similarly and he got better. A chest x-ray and pulmonary function test were normal. In the fall of 2018 he went to Florida and while in Florida he had few episodes of cough and shortness of breath and was diagnosed with allergic asthma with bronchitis and was treated with antibiotics and steroids and got better.

He returned in spring of 2019 and was being seen by the pulmonologist and allergist and was getting treatment for allergic asthma. In late summer of 2019 he presented back to the primary care physician's office with recurrent nasal congestion, cough and shortness of breath. He was found to have inflamed nasal mucosa and had wheezing throughout his lungs. His oxygen saturation was normal. A CT scan of the chest was done which showed mediastinal adenopathy and suspected pulmonary embolism. A CT angiogram confirmed bilateral pulmonary embolism. He was started on DOAC. Hypercoagulable work up was negative. CBC showed elevated white blood cell count of 14.7 and eosinophil count of 42%. He was seen by hematologist and a bone marrow biopsy was done which was normal except for hypereosinophilia. At that point an underlying malignancy was suspected and he was referred back to the pulmonologist. A CT scan of the chest was repeated and the mediastinal lymph nodes were getting smaller. The differential diagnosis at the time was interstitial lung disease, lymphoma, and eosinophilic vasculitis. In the meantime he developed numbness of the legs and back pain. MRI of the lumbosacral spine was unremarkable. He was seen by neurologist and electromyogram was done which was consistent with polyneuropathy. In February 2020 he developed pruritic rash on his legs and has had biopsy done by dermatologist which was consistent with vasculitis. He was given a course of corticosteroids and his rash improved. At the same time his nasal congestion, cough and shortness of breath also improved.

He was referred to the rheumatologist and at that time the rheumatologist was suspecting Eosinophilic granulomatosis with polyangiitis (EGPA). He was given a follow up appointment pending more lab works.

While waiting for the rheumatology follow up patient developed acute abdominal pain in April 2020. CT scan and abdominal ultrasound were consistent with cholecystitis. His troponins was found to be elevated. Therefore he underwent cardiac catheterization which showed insignificant coronary artery disease. Subsequently he underwent cholecystectomy. Biopsy of the gallbladder showed granulomatous vasculitis with necrosis of medium sized blood vessels which was consistent with Eosinophilic granulomatosis with polyangiitis.

Upon return home after cholecystectomy patient developed palpitation and he returned to the emergency department. He was diagnosed with new onset Atrial Fibrillation and was admitted back to the hospital. He continued to have abdominal pain. An EGD done at that time showed Gastritis. Biopsy did not show any significant abnormalities.

The above findings were discussed with the rheumatologist and he was started on prednisone at the dose of 60 mg per day. Once he was started on prednisone his symptoms started improving. At the follow up appointment with the rheumatologist he was confirmed with the diagnosis of EGPA. Later he was started on Rituxan and the prednisone was tapered and slowly discontinued. All his symptoms including nasal congestion, cough, shortness of breath, abdominal pain, loss of appetite, weight loss, and weakness have improved except neuropathy which is slowly getting better. He is now getting physical therapy and his general strength is improving.

Discussion:

EGPA, previously called Churg-Strauss syndrome is a rare multi organ disorder. The incidence in United States is 18 per million population. Median age group is 40 years and there is no gender preference. The most commonly involved organ is lung followed by skin.but it can affect any organ system. Peripheral blood eosinophilia is a cardinal feature. Other features include but not limited to heart failure, cardiac rhythm abnormalities, thromboembolism, peripheral neuropathy, renal insufficiency, hematuria, abdominal pain, diarrhea, myalgia, poly arthritis, lymphadenopathy, loss of appetite, weight loss and weakness

American College of rheumatology has established six criteria for diagnosis of EGP a in a patient with documented vasculitis.

1. Asthma
2. Greater than 10% eosinophilia
3. Neuropath
4. Migratory or transient pulmonary opacities
5. Paranasal sinus abnormality
6. Tissue biopsy showing Eosinophilic accumulation in extra vascular areas.

The patient described had almost all of these clinical features.