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Case report

A rare case of cavitary lung lesions in an adolescent: Granulomatosis with polyangiitis ☆

Sekhar Iyer, MD, Michael A Simon, MD, Donald Schroeder, MD, Lyle Gesner, MD*

Department of Radiology, RWJBH – Saint Barnabas Medical Center, 94 Old Short Hills Road, Livingston, NJ 07039, USA

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ABSTRACT

Granulomatosis with Polyangiitis (GPA) is a life threatening disease if left untreated which predominantly affects the adult population. As clinical presentation is often non-specific there is a heavy reliance on radiologic, laboratory and biopsy findings in diagnosis. We present a case of a 17-year-old male who presented with a history of tea colored urine and recurrent epistaxis who now complained of cough and congestion. The patient failed multiple courses of outpatient antibiotics and a CT of the chest while in the ED demonstrated multiple cavitary lesions. Subsequent workup and biopsy confirmed the diagnosis of GPA. It is important for the Radiologist and other clinicians to keep GPA in their differential when presented with a cavitary lung lesion as prompt treatment is required for good outcomes.

Introduction

Granulomatosis with Polyangiitis (GPA) is a life threatening disease if left untreated which predominantly affects the adult population. As clinical presentation is often non-specific there is a heavy reliance on radiologic, laboratory and biopsy findings in diagnosis. Often thought of as a disease seen in adults, GPA in the pediatric population is exceedingly rare. A high index of suspicion for the disease is imperative as delay in treatment can lead to renal and respiratory failure.

Case

A seventeen-year-old male with a history of tea colored urine and recurrent epistaxis presented to the Emergency Department with complaints of cough, nasal congestion and fever. Of note, the patient was treated with multiple courses of antibiotics as an outpatient for these symptoms but did not experience any relief.

While in the Emergency Department a non-contrast CT of the chest was ordered which demonstrated multiple areas of

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* Corresponding author. M.A. Simon.
E-mail address: MichaelSimonMD@gmail.com (M.A. Simon).

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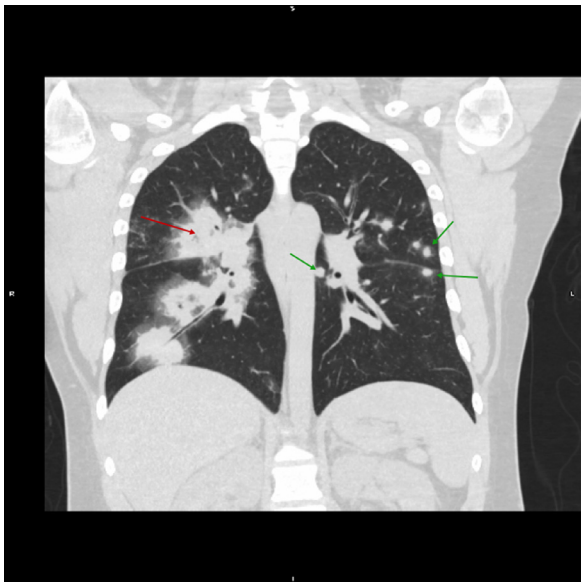

Fig. 1 – Coronal and axial CT of the chest without contrast demonstrates multiple airspace opacities in a central distribution most within the right upper lobe (red arrow). Sporadic nodular densities where also noted (green arrows). (Color version of figure is available online)

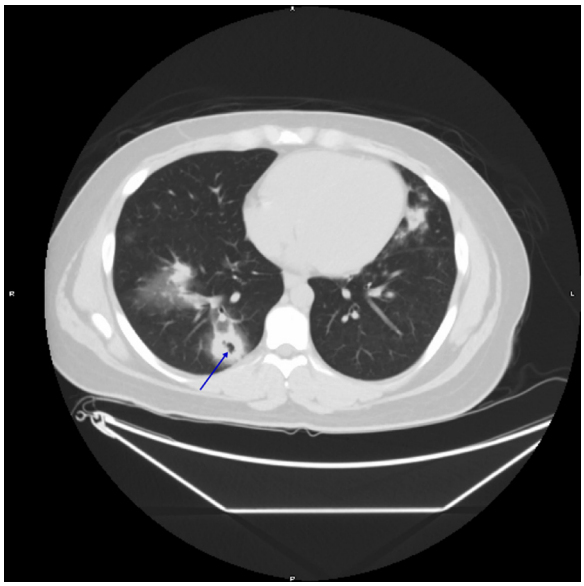

Fig. 2 – Axial image demonstrates bilateral nodular opacities with surrounding ground glass densities. A cavitary lesion is seen within the right lower lobe (blue arrow). (Color version of figure is available online)

consolidation and sporadic nodular densities (Fig. 1), some of which were cavitating (Fig. 2). Laboratory testing at that time was significant for hematuria and elevated inflammatory markers including an erythrocyte sedimentation rate of 93 mm/hr (normal range 0-15 mm/hr), CRP of 23.26mg/dL (normal range 0-0.50 mg/dL) and ferritin of 1206 ng/mL (normal range 30-400 ng/mL).

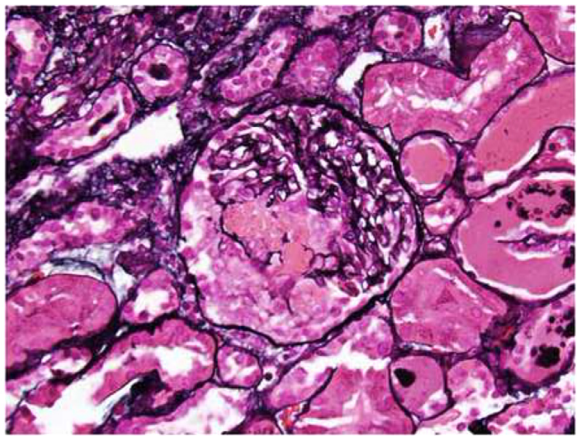

Fig. 3 – Silver stained one micron thick sections of the renal biopsy were examined using an electron microscope and demonstrated cellular crescents and fibrinoid necrosis. Findings were consistent with pauci immune necrotizing and crescentic glomerulonephritis.

The patient was admitted to the PICU for further workup which revealed a positive proteinase 3 antibody (217.6 AI normal Range <1.0 AI). Complement, ANA and single stranded DNA antibody levels were within normal limits.

Given the patient's symptoms and laboratory findings there was a strong suspicion for GPA. A kidney biopsy was subsequently performed by the Interventional Radiology team which revealed the presence of pauci-immune crescentic glomerulonephritis (Fig. 3) consistent with the diagnosis of GPA. The patient was started on a regimen of corticosteroids and Rituximab and was discharged for close follow-up with his rheumatologist.

Discussion

GPA, formerly known as Wegner's Granulomatosis, is an Antineutrophil cytoplasmic antibody associated vasculitis which predominantly affects the small and medium vessels within the older adult population [1]. Organ systems most commonly affected are the upper and lower respiratory tract and kidneys and left untreated the mortality rate is greater than 80% [2].

It is estimated that the incidence of GPA is 12.8 cases per 1 million person years with a slight predilection for Females in the adult population [2,3]. GPA in the pediatric population is estimated to be 1.8 cases per 1 million person years [2]. In both populations the disease is predominantly found in Caucasians [3,4].

Typically, the clinical presentation of GPA is non-specific with complaints including fatigue, fever, weight loss, cough and hematuria. Involvement of the lungs can lead to the development of mass like nodules The clinical course varies with some patients experiencing rapid progression of the disease while others experience months of constitutional symptoms

[5]. The presentation does not differ between the Adult and Pediatric populations.

Diagnosis of GPA can be challenging due to its resemblance to symptoms of many common conditions. Although classification criteria have been developed both by the American College of Rheumatology and European League against Rheumatism/Pediatric Rheumatology European Society to aid in diagnosis laboratory investigation and biopsy are strongly recommended [6]. The combination of symptoms, lab values suggestive of inflammation i.e. PR3- Antineutrophil cytoplasmic antibody which is positive in 92% of patients with GPA, c-reactive protein and erythrocyte sedimentation rate, with imaging findings should prompt a biopsy to confirm diagnosis [7].

The kidney is often the location chosen for biopsy due to its ease of access and common involvement in the disease. The biopsy typically reveals the presence of pauci-immune glomerulonephritis [7].

It is crucial to detect the disease early on for treatment to be effective and successful. Immune suppressants are the mainstay of treatment for GPA. In general, corticosteroids with a combination of immunosuppressive agents such as methotrexate, rituximab and cyclophosphamide are used. Treatment is similar in both the adult and pediatric populations [8].

The prognosis for patients with GPA has greatly improved since its discovery. However, long-term complications exist including chronic kidney failure, deafness and severe respiratory failure. In addition, relapse rates are higher in the pediatric population [8].

Patient consent

Formal consents are not required for the use of entirely anonymized images from which the individual cannot be identified- for example, x-rays, ultrasound images, pathology slides or laparoscopic images, provided that these do not contain any identifying marks and are not accompanied by text that might identify the individual concerned. Therefore, consent was not obtained for our case report.

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