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A Case of Granulomatosis with Polyangiitis Presenting as a Solitary Thick-Walled Pulmonary Cavity

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Abstract

Introduction: Chest radiographic findings in patients with granulomatosis with polyangiitis can be highly variable. Most of them present with multiple unilateral or bilateral nodules which may be sometimes associated with cavities. When the disease causes formation of pulmonary cavities, the cavitary lesions seen on chest imaging are mostly multiple, found in both lung fields and small in size. A solitary cavitary lesion in the chest radiographic image of a patient with granulomatosis with polyangiitis is not common. We present a case of granulomatosis with polyangiitis manifesting as a solitary thick-walled pulmonary cavity.

Case presentation: A 20-year old male presents to the emergency department complaining of a persistent dry cough of ten days and associated intermittent fever of three days. He reports night sweats and weight loss of ten pounds in the past month. The patient denies chills, nausea, vomiting and hemoptysis. He denies travelling outside the United States or exposure to anyone with similar symptoms.

Discussion: On initial evaluation, the patient was febrile and tachycardic. Blood pressure and respiratory rate were within normal limit and oxygen saturation was 100%. His labs revealed leukocytosis and mildly elevated pro-calcitonin. The induced sputum culture came back negative for acid-fast bacilli but grew *Achromobacter denitrificans*. The fungi culture, blood culture, urine antigen for legionella and Quantiferon TB gold came back negative. Other lab values including renal function and urinalysis were within normal limits.

The patient had a chest x-ray that showed a right upper lobe cavitary opacity on PA view which was further investigated with a chest CT that showed a solitary thick walled cavitary lesion measuring 3.5 cm × 3.6 cm in the right upper lobe with adjacent pulmonary consolidation. Treatment for pneumonia with lung abscess was commenced with appropriate antibiotics. After about a week of receiving meropenem, he developed pleuritic chest pain, his fever and cough also did not abate. Pulmonary vasculitis was considered and was confirmed by positive C-ANCA and subsequent respiratory mucosa biopsy which showed necrotizing granulomatous inflammation.

Conclusion: Our patient would have been diagnosed earlier if tests for C-ANCA and Anti-PR3 tests had been done to rule-out vasculitis based on the initial chest image findings. We believe this case shows that pulmonary vasculitis should be considered in the differential diagnosis in a patient presenting with a solitary cavitary pulmonary lesion when there are other constitutional symptoms known to be associated with it.

Keywords Granulomatosis with polyangiitis; Solitary pulmonary cavity; pulmonary vasculitis; Chronic mastoiditis; Otitis media; Bell's palsy; Lung abscess

Abbreviations: C-ANCA: Cytoplasmic Antineutrophil Cytoplasmic Antibody; Anti-PR3: Proteinase 3 Antibody; GPA: Granulomatosis with Polyangiitis

Introduction

Granulomatosis with polyangiitis (GPA) is a multi-systemic disease characterized by inflammation of small- and medium-sized blood vessels and can manifest clinically as constitutional and organ-specific symptoms involving the airway, ear canal, kidneys, cartilages, bones and also the possibility of entrapment of the facial nerve [1,2]. Chest radiographic findings in patients with GPA can be highly variable [3]. Most of them present with multiple bilateral nodules which may be sometimes associated with cavities in about 25% of cases [4]. Other possible radiographic chest findings can include; peripheral wedge shaped consolidation, pulmonary hemorrhage, reticulonodular infiltrates and Pleural effusions which have been reported in about 15-20% of cases [5]. Cavitary lesions seen on chest imaging of GPA patients most of the time are usually small, multiple, and bilateral [6]. Chest radiographic presentation in a GPA patient manifesting with a solitary thick-walled cavitary lesion is not common and we found just one case of it in the search of published case reports [7]. We present a

case of granulomatosis with polyangiitis manifesting as a solitary thick-walled pulmonary cavity.

Case Presentation

A 20-year old male presents to the emergency department complaining of a persistent dry cough of ten days and associated intermittent fever of three days. He reports night sweats and weight loss of ten pounds in the past month. The patient denies chills, nausea, vomiting and hemoptysis. He denies travelling outside the United States or exposure to anyone with similar symptoms. Past medical history includes chronic left mastoiditis and left otitis media associated with Bell's palsy which he developed about two months prior. He had a left myringotomy, left mastoidectomy and facial nerve decompression two weeks prior.

On initial evaluation, the patient was febrile with a temperature of 102°F and tachycardic with a heart rate of 120 beats/min. He had normal blood pressure of 128/76mmHg, respiratory rate was 18 breaths/min, and oxygen saturation was 100%. His labs revealed leukocytosis with 18.4 × 103 white cells/mm3 (84.4% neutrophils) and mildly elevated pro-calcitonin of 0.15 ng/ml. The induced sputum culture came back negative for acid-fast bacilli but grew *Achromobacter denitrificans*. The fungi culture, blood culture, urine antigen for legionella and Quantiferon TB gold came back negative. Other lab values including renal function and urinalysis were within normal limits. The patient had a chest x-ray that showed a right upper lobe cavitary opacity on PA view (Figure 1) which was further investigated with a chest CT(Figure 2) that showed a solitary thick walled cavitary lesion measuring 3.5 cm × 3.6 cm in the right upper lobe with adjacent pulmonary consolidation.

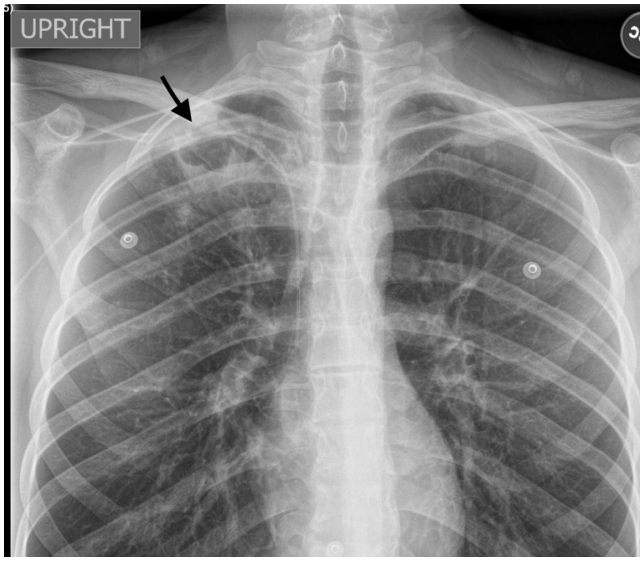

Figure 1: Plain chest x-ray with arrow indicating right upper lobe cavitary opacity.

A diagnosis of pneumonia with lung abscess was made and treatment with appropriate antibiotics was commenced. After about a week of receiving meropenem, he developed pleuritic chest pain, his fever and cough also did not abate. Bronchoscopy was performed which showed stenotic apical segment of the right upper lung lobe (Figure 3) and bronchial lavage of the right upper lobe was sent for culture which came back negative. Transbronchial biopsy of the right upper lobe showed reactive fibrous capsule and benign soft tissue which were non-diagnostic. In view of the right upper lung lobe stenosis and persistent symptoms despite antibiotics, pulmonary vasculitis was considered.

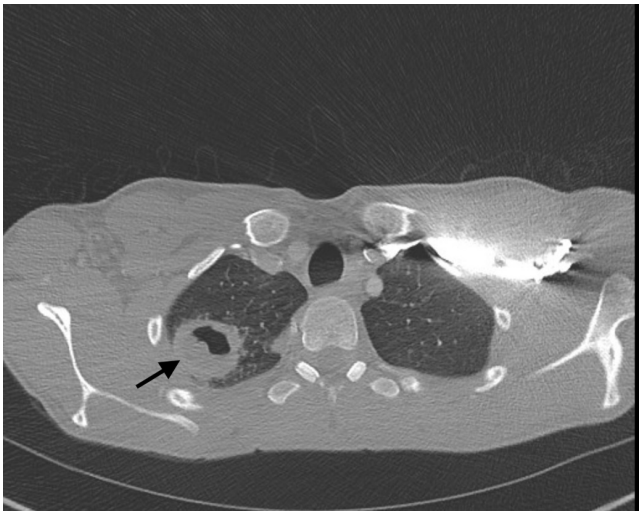

Figure 2: Axial chest CT without contrast with arrow showing a solitary thick walled cavitary lesion in the right upper lobe and adjacent pulmonary consolidation.

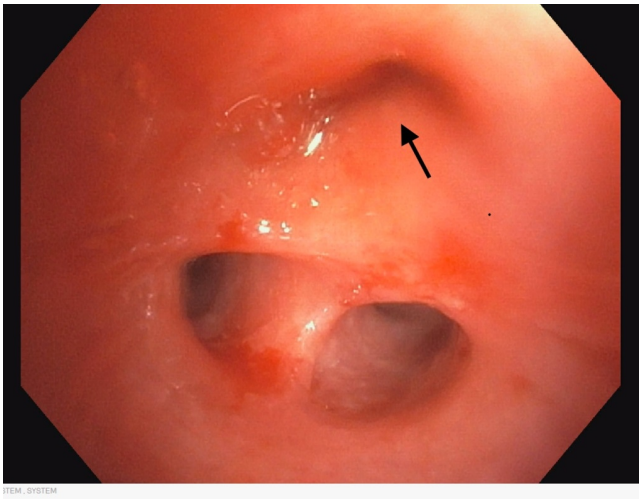

Figure 3: Transbronchial image with arrow showing stenotic apical segment of the right upper lung lobe.

When we reviewed the petrous CT scan without contrast done prior to his mastoidectomy, it showed complete opacification of the right maxillary sinus and asymmetric opacification of the right nasal cavity in keeping with sinusitis along with features of the left-sided mastoiditis (Figure 4). The left mastoid bone that was excised during his surgery was sent for culture but grew no organism. Histological evaluation of the excised left mastoid bone was apparently not done.

We therefore ordered cytoplasmic antineutrophil cytoplasmic antibody test (C-ANCA) which came back positive. Confirmatory proteinase 3 antibody assay (Anti-PR3) also came back positive. Biopsy of the upper respiratory mucosa was done which showed extensive necrotizing granulomatous inflammation and this further confirmed our diagnosis of GPA (Figure 5).

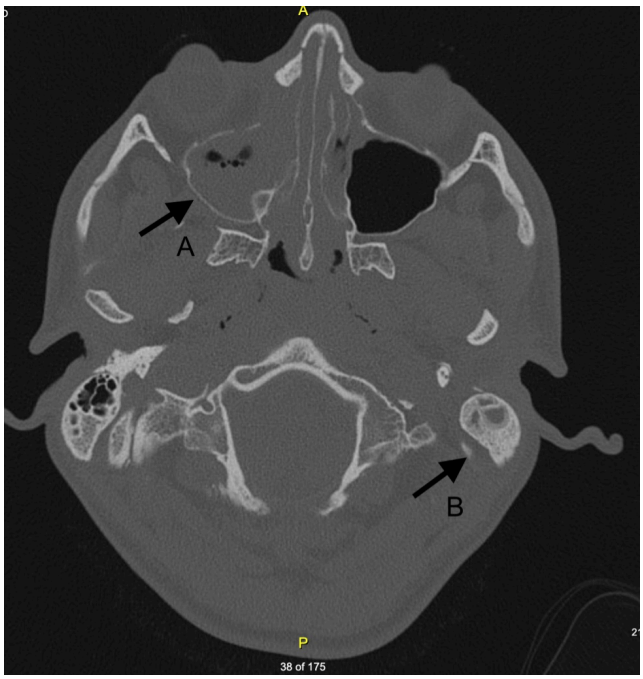

Figure 4: Petrous CT scan pre-mastoidectomy showing with arrow A showing opacification of the right maxillary sinus and asymmetric opacification of the right nasal cavity. Arrow B shows left-sided mastoiditis.

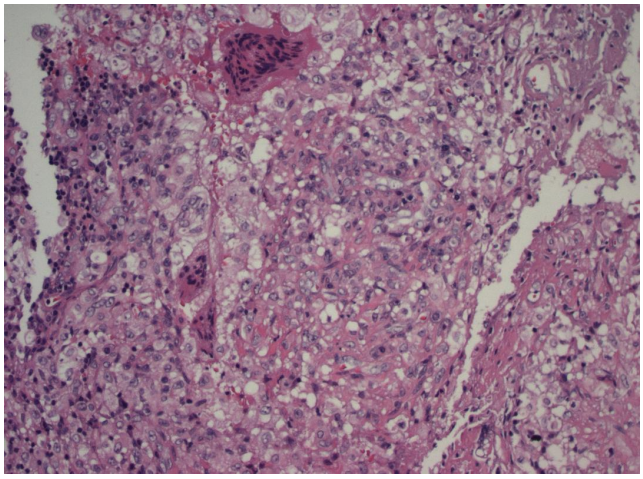

Figure 5: Microslide of upper respiratory mucosa showing extensive necrotizing granulomatous inflammation.

Our patient was managed with IV methylprednisone at 1g daily for three days. He received the first of the 4 doses of weekly rituximab infusion at 375 mg per square meter of body surface area while still in the hospital. His IV steroid medication was transitioned to oral prednisone tablets at 60 mg daily and he got oral Bactrim three times weekly for prophylaxis against *Pneumocystis jirovecci* pneumonia. He responded well to the treatment as the intractable cough, chest pain and intermittent fever ceased. He was discharged afterwards to receive the remaining rituximab infusions as outpatient. His dosage of oral prednisone was reduced to 40 mg daily. He was also prescribed alendronate at 70 mg weekly to prevent steroid-induced bone loss.

Follow up chest x-ray showed regression in the size of the right upper lobe cavitary lesion. CT scan of the chest has been scheduled for 2 months after completing the rituximab infusions. A petrous CT to follow up on his rhinosinusitis has been scheduled but pending at the time of this report. He remains symptom-free and he is tolerating the treatment well without any adverse effects.

Discussion

The most common symptom in patients with GPA is respiratory symptoms which may affect either the upper airway, lower airway or both. Kubaisi et al. postulated that about 85–90% of GPA patients develop lung disease eventually [8].

There have been reported cases of GPA misdiagnosed as pulmonary infections or even as tumors and is due to high variability in the pulmonary manifestations [11]. The initial diagnosis of pneumonia with lung abscess in our patient was on account of the cavitary lesion and surrounding consolidation in the setting of other symptoms of infection. A diagnosis of GPA would have been made earlier in this patient if C-ANCA and Anti-PR3 tests were ordered immediately after review of his chest images to rule out vasculitis. The fact that the petrous CT scan without contrast done prior to his mastoidectomy was not initially available at the time of the initial assessment also delayed bringing up pulmonary vasculitis to the top of the differential diagnosis.

With ANCA being positive in about 90% of GPA, it has been recognized to be sensitive for GPA [12]. ANCA is however not specific for GPA because high titers are also seen in inflammatory bowel disease, in autoimmune-mediated liver diseases, in rheumatoid arthritis and in systemic lupus erythematosus [13,14]. Most ANCA-negative GPA is seen in patients whose GPA manifestation is limited to the upper respiratory tract sparing the lungs and kidneys [15]. Anti-PR3 is usually positive in about 90% of GPA with active and generalized GPA. The Anti-PR3 titer has been shown to correlate with the disease activity and can also predict the possibility of relapse in some patients thought to otherwise be in complete remission [16-18].

Conclusion

Our patient would have been diagnosed earlier if tests for C-ANCA and Anti-PR3 tests had been done to rule-out vasculitis based on the initial chest image findings. We believe this case shows that GPA should be suspected in patients who have a solitary cavitary pulmonary lesion and a background of other constitutional symptoms associated with it.

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