Radiological Manifestations of Granulomatosis with Polyangiitis

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Abstract

Granulomatosis with polyangiitis (GPA) is a type of small vessel vasculitis that can affect almost all organs of human body but is most commonly found at respiratory system (nose, trachea, and lungs) and kidneys. Most commonly, GPA Presented with upper respiratory tract symptoms, which were found in nearly every patient. Rhinitis, sinusitis, and, in rare cases, nasal ulcers are among the symptoms. Chronic rhinitis is a stage of rhinitis where standard treatment fails, and that is also a common symptoms of this condition. Most people's lungs are affected even if they don't show any symptoms, but if they do, they frequently include cough, hemoptysis, and dyspnea.

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Introduction

Granulomatosis with polyangiitis (GPA) is a type of small vessel vasculitis that can affect almost all organs of human body but is most commonly found at respiratory system (nose, trachea, and lungs) and kidneys. Most prevalent, GPA affected the upper respiratory tract, which were found nearly in every patient. Rhinitis, sinusitis, and, in rare cases, nasal ulcers are among the symptoms.[1] Chronic rhinitis is a stage of rhinitis where standard treatment fails, and that is also a common symptoms of this condition. Most people's lungs are affected even if they don't show any symptoms, but if they do, they frequently include cough, hemoptysis, and dyspnea.[2]

Antineutrophil cytoplasmic antibodies (ANCAs) are such antibodies that target those proteins which are found in cytoplasm of neutrophils. When serum antibodies attach to indicator neutrophils, the cytoplasmic (c) ANCA refers to the diffuse staining pattern seen. Perinuclear (p) ANCA is a staining pattern seen on indicator neutrophils, with the enzyme myeloperoxidase being the principal target of these antibodies; it is seen in >90% of Wegener granulomatosis patients.[3]

GPA usually manifests as inflammation of the respiratory tract that later develops into generalized small vessel vasculitis.[4,5] Increasing age, pulmonary and kidney involvement usually attributes to increased mortality and morbidity.[6] In this case series we randomly selected six patients that presented to clinic and were diagnosed with GPA through combination of clinical features, laboratory findings and radiological features, to help the clinician reach an earlier diagnosis.

Case Report

Case 1: A housewife of 38 years age presented with cough for at-least 6 weeks, fever and weight loss. She didn't have any underlying disease or any family history of asthma, TB or diabetes. On examination she had a few crackles in right lung mid zone with bronchial breathing. Baseline workup was done which showed a raised C reactive protein (CRP) of 22.5mg/dl; it was otherwise unremarkable. For investigation purpose X-ray of chest was advised, results of which showed consolidation in the right lung mid zone with cavitation. CT scan of the chest showed multiple nodules in the right lung, the largest one in the posterior segment showing central cavitation. Bronchoscopy was performed which showed inflamed and friable mucosa on the right side. AFB

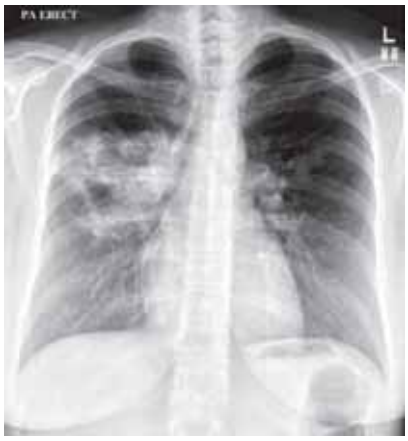

Figure 1: X-Ray showing consolidation on right mid-zone

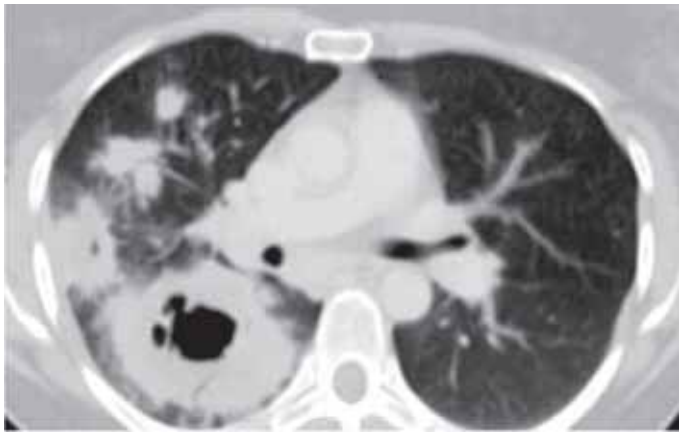

Figure 2: CT Scan showing posteriorly cavitary lesion on the right side

smear, Gene Xpert, Fungal smear and culture, and Nocardia were all negative. C-ANCA and P-ANCA were sent, which showed a raised C-ANCA level of 13.71 U/ml, and patient was diagnosed as Granulomatosis with polyangiitis.

Case 2: 22 years old, engineering student studying in Australia presented with 2 months history of fever, cough, post nasal drip, exertional dyspnea, joint pain and documented weight loss of 5 kg (in 2 months). On examination he had conjunctival hyperaemia suggestive of conjunctivitis. His total leucocyte count was 7.4 x 109/L with a normal differential. His chest X-ray reveled cavitary nodule present in the left lung mid zone. Urine detailed report showed RBCs >20 and hemoglobin +3. Sputum for AFB smear, gram stain and culture showed no growth. P-ANCA and c-ANCA were sent; his c-ANCA level was 39.35 U/mL, hence a diagnosis of Granulomatosis with polyangiitis (GPA/Wegener's Granulomatosis) was made.

Case 3: Another case is a 30 years old farmer from Multan which came with presentation of fever, cough, hemoptysis, hematuria and skin rash for the past six months. He also said he had lost 10 kg weight. His chest x-ray revealed consolidation on right side involving mid and lower zones. P-ANCA and c-ANCA were done which were raised and thus a diagnosis of

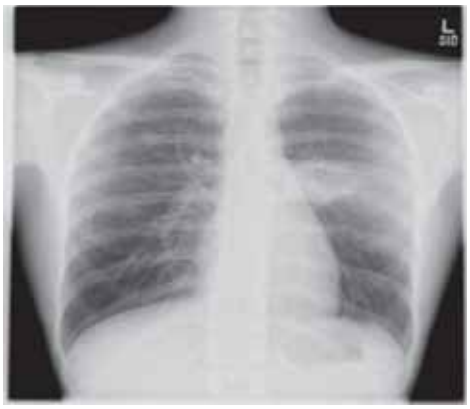

Figure 3: Chest X-ray showing a left mid zone cavitary nodule

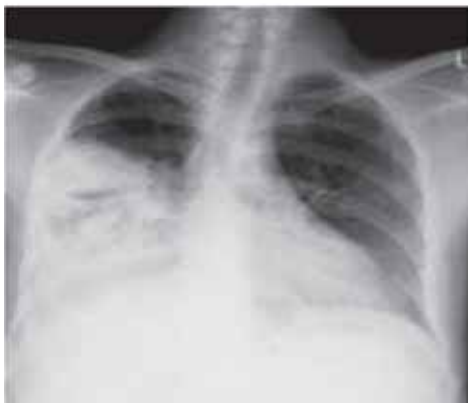

Figure 4: Consolidation on right side

Granulomatosis with polyangiitis (GPA/Wegener's Granulomatosis) was confirmed.

Case 4: 45 years old man presented with 6 months history of gangrene in his foot. He came to the emergency department, where baseline workup showed deranged liver function tests, and chest x-ray was suggestive of mild effusion, more on the right side, which was confirmed on ultrasound. U/S guided pleural tap was done which showed an exudative pleural effusion. Two weeks later his CT scan chest showed bilateral mild to moderate pleural effusion with few loculations on the left side and associated basal atelectasis. No evidence of pulmonary consolidation or focal mass in bilateral lungs. He was

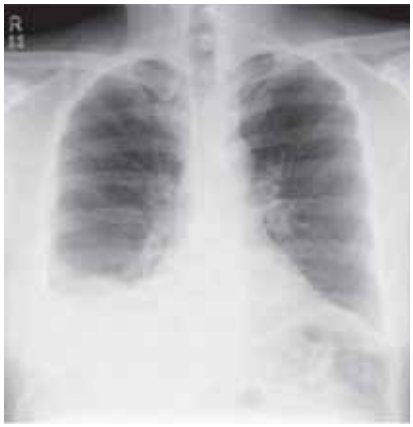

Figure 5: Chest X-ray suggesting a pleural effusion on the right side

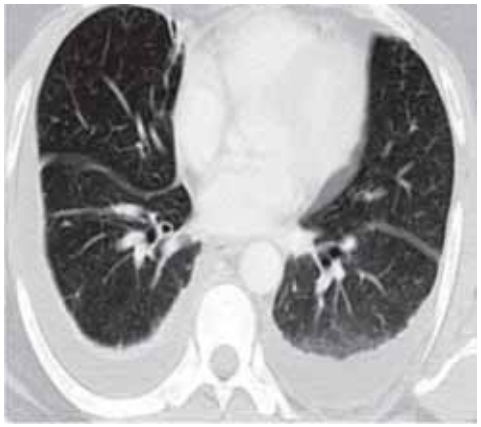

Figure 6: CT scan showing bilateral mild to moderate pleural effusion

followed up in clinic where more workup was done including ANA profile, Anti-DsDNA, C3 and C4 levels and ANCA levels. His C-ANCA level was >100 U/mL and a diagnosis of Granulomatosis with polyangiitis (GPA/Wegener's Granulomatosis) was made.

Case 5: 22 years old female presented to clinic with history of productive cough and fever for one week. Baseline workup done was unremarkable. Her chest x-ray showed multiple cavitary lesions in lungs bilaterally for which relevant workup was done. CT-scan chest showed multiple cavitary lesions and patchy areas of ground-glass opacity identified in bilateral lung fields. P-ANCA and c-ANCA were raised

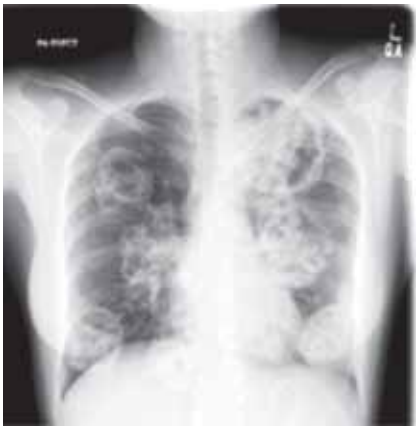

Figure 7: Chest X-ray shows multiple cavitary lesions bilaterally.

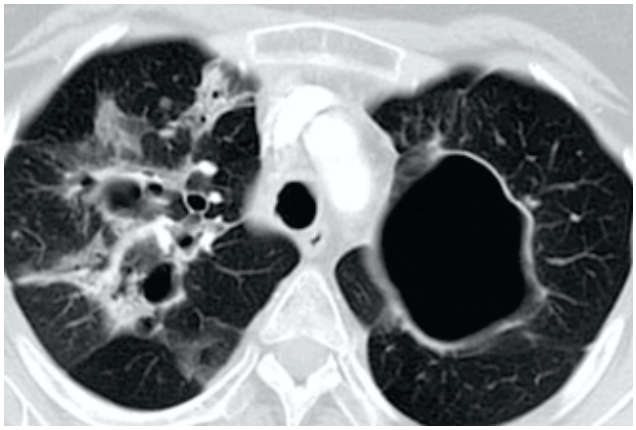

Figure 8: CT scan chest shows multiple cavitary lesions in bilateral lung fields with some patchy areas of ground glass. The largest one in the left upper lobe measures approximately is 65 x 58 mm in size.

which confirmed the diagnosis of Granulomatosis with polyangiitis (GPA/Wagner's Granulomatosis).

Case 6: 20 years old female presented with complaints of fever, cough and shortness of breath. Baseline investigations were done which showed raised total leucocyte count of 32 x 10E9/L. Chest x-ray showed bilateral alveolar infiltrates. Sputum culture did not show any growth, blood culture was also negative. Bronchoscopy was done and AFB smear, Gene Xpert, Fungal smear and culture, and Nocardia were all negative. P-ANCA was significantly raised (62.90 U/mL) and a diagnosis of granulomatosis with polyangitis was made (P-ANCA positive vasculitis).

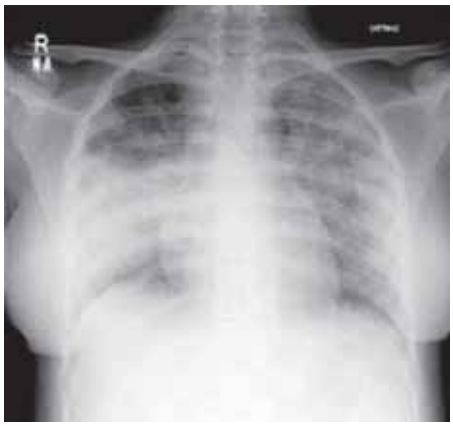

Figure 9: Patchy infiltrates noted in the right mid and lower lung zone. Patchy infiltrates are also noted involving the entire left lung

Discussion

Wegener's granulomatosis (GPA) is an uncommon autoimmune disease characterised by small-vessel vasculitis of the respiratory system, kidneys, and other organs (typical Wegener's granulomatosis). The diagnosis of GPA is based on clinical symptoms, ANCA serology, and histologic evidence of vasculitis from a relevant organ biopsy, such as the skin, lungs, or kidneys.[7] Most patients have respiratory symptoms, therefore chest radiographs are taken initially, followed by chest computed tomography (CT) scans to further describe the abnormalities. Pulmonary nodules, fixed pulmonary infiltrates, and pulmonary cavities are all possible radiographic findings.[8]

Many most studies have been conducted on radiological findings in GPA patients. In a 1990 investigation, Cordier et colleagues found that lung nodules were the most common manifestation. In a case series,[9] pulmonary symptoms in 14 Wegener's granulom-atosis patients were described, each with its own set of characteristics.[10] The American College of Rheumatology established a diagnostic criterion for GPA in 1990 that includes the presence of abnormal radiological abnormalities on a chest radiograph.[11] In the diagnosis of small vascular vasculitis, a CT chest scan is regarded more trustworthy than a chest X-ray.[12] In 2016 another study published revealed that 33 patients out of 44 had abnormal CT and the most common finding was nodules and masses.[13] In another case series, chest CT scans of 10 patients with diagnosed GPA were reviewed which revealed multiple pulmonary nodules in seven of the patients.[14]

There are also a few studies done in the subcontinent. A case series conducted in India showed that 9 out of 11 patients had abnormal chest radiographs in which nodular infiltrates were the most common finding.[15] Another study conducted in Karachi, Pakistan found that 16 out of 51 patients had pulmonary infiltrates in the form of consolidations on radiological examination, making it the most common presentation in our setting.[16]

The studies conducted so far show nodules to be the most common radiological presentation in patients with GPA. The studies conducted in sub-continent also revealed nodular masses to be the most common finding. In this case series patients are of various ages, gender and ethnic groups; we can see that every patient is different from the other in terms of symptoms and radiological manifestations. Two of the cases had almost similar symptoms but their radiological presentations were different. One of our cases had pleural effusion. While two of the cases showed cavitation in lungs on x-ray and they had completely different clinical manifestations. Yet another case showed bilateral infiltrates on chest x-ray.

To conclude, this case series of six patients shows that Wegener's granulomatosis can present with multiple radiological features. Whenever there is a mass, consolidation, a nodule, an effusion, a cavity or bilateral diffuse infiltrates, Granulomatosis with polyangiitis (GPA) needs to be kept in the differential diagnosis, especially if history and clinical features are suggestive. It also indicates that radiological features can be so diverse in GPA that they are not enough to diagnose it. A combination of clinical features, radiological findings, ANCA levels and biopsy from a relevant organ are needed to diagnose it.

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