Case Report

Granulomatosis with Polyangiitis - A case report

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Abstract

Granulomatosis with Polyangiitis (GPA) is a rare multisystem autoimmune disease. It is characterized histopathologically by necrotizing granulomatous vasculitis. The classical clinical triad consists of upper airway involvement (characterized by sinusitis, otitis, nasal mucosa ulcers, bone deformities, and subglottic stenosis), lower respiratory tract involvement (cough, chest pain, hemoptysis) and glomerulonephritis. We reported here a case of Wegener's granulomatosis presenting as a thick walled cavity.

Key words

Granulomatosis, Polyangiitis, Autoimmune disease, Necrotizing granulomatous vasculitis.

Introduction

Granulomatosis with Polyangiitis (GPA) is a rare multisystem autoimmune disease. It is characterized histopathologically by necrotizing granulomatous vasculitis. The classical clinical triad consists of upper airway involvement (characterized by sinusitis, otitis, nasal mucosa ulcers, bone deformities, and subglottic stenosis), lower respiratory tract involvement (cough, chest pain, hemoptysis) and glomerulonephritis [1].

Although only 40% of patients have renal involvement at presentation, 80%–90% ultimately develop renal disease [2].

It is believed that the disease begins as a localized respiratory tract granulomatosis, which then generalizes into a vasculitis that affects small and medium-sized vessels [3].

Patient presentation varies and depends on the organ system affected. Some patients present with chronic nasal obstruction, which may be

misdiagnosed as chronic sinusitis; others may present with overt acute renal or respiratory failure. Patients with pulmonary involvement often complain of cough with or without Hemoptysis, dyspnea, fever, and chest pain [3].

We reported here a case of Wegener's granulomatosis presenting as a thick walled cavity.

Case report

A 20 year old male presented to the department of Pulmonary Medicine, Meenakshi Medical College Hospital and Research Institute with complaints of cough with expectoration and fever for 1 month. He had cough with minimal whitish expectoration, with no diurnal or postural variation. He had low grade, intermittent fever for 1 month associated with large joint pains. He also complained of exertional breathlessness of Grade 2 MMRC scale. He had history of loss of weight and loss of appetite, with no history of hemoptysis, wheeze or chest pain. He had no prior history of anti-tuberculous treatment and no contact history of tuberculosis.

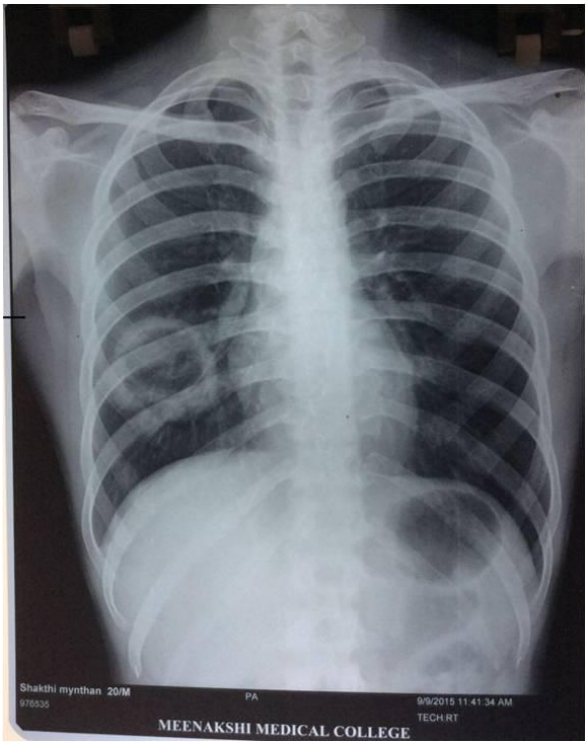
He was diagnosed as a case of rheumatoid arthritis 2 months back elsewhere and was started on methotrexate, hydrochloroquine, sulphasalazine and tapering doses of steroids. On general examination he was found to have skin rashes over back, chest and neck. His respiratory and other systemic examinations were normal.

Investigations

Initial investigation showed hemoglobin 13.7 g/dl, elevated ESR (60/57), and CRP (88.5). His renal and liver function tests were within normal limits. He had elevated RA factors; however Anti CCP titers were low. CXR showed a thick walled cavity in the right lower zone (**Photo – 1**).

Based on clinical and radiological findings, the differential diagnoses of tuberculosis, bacterial pneumonias, lung abscess, and lung cancer, connective tissue disorders like SLE, rheumatoid arthritis with nodules, sarcoidosis, and vasculitis were considered. On further work up, sputum smears were negative for AFB, Mantoux test was negative, sputum gram stain and culture showed no growth.

Photo – 1: CXR showing a thick walled cavity in the right lower zone.



HRCT Chest 1 showed moderate thick walled cavity with surrounding areas of consolidation involving superior and lateral basal segments of right lung (**Photo – 2**). HRCT Chest 2 showed dilated ascending aorta and aortic arch, with thickened aortitis, Thickened left ventricular wall (**Photo – 3A, 3B**).

Photo – 2: HRCT Chest 1 shows: Moderate thick walled cavity with surrounding areas of consolidation involving superior and lateral basal segments of right lung.

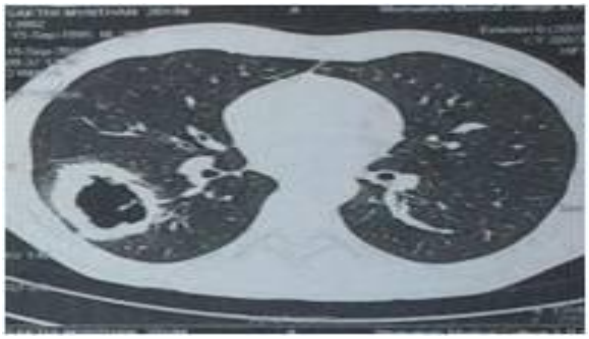
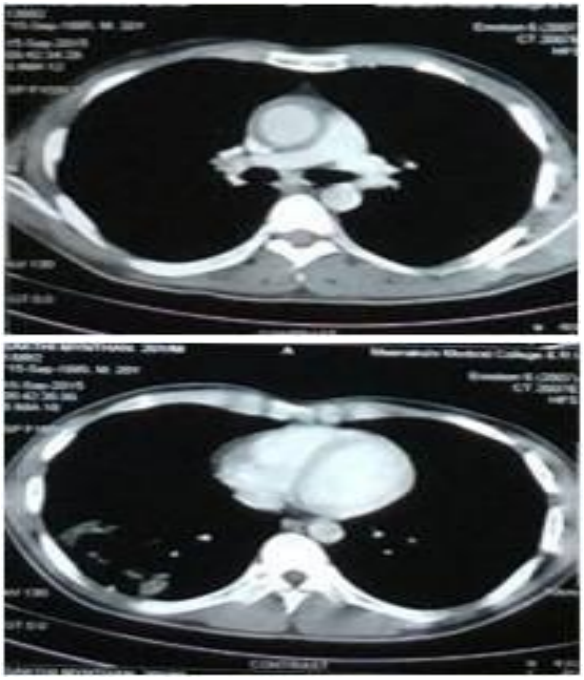


Photo – 3A, 3B: HRCT Chest 2 shows: Dilated ascending aorta and aortic arch, with thickened aortitis, Thickened left ventricular wall.



Bronchoscopy done was normal. BAL samples sent for Gene Xpert PCR was negative. BAL gram stain and culture showed no growth. BAL AFB smear was negative. Cytology report showed inflammatory cells. On further detailed history he complained of frequent painful oral ulcers with repeated sinusitis and an episode of epistaxis. TBLB showed granuloma seen in cavity wall, with epithelioid and Langhans type multinucleate giant cells and foamy histiocytes.

ENT examination with nasal endoscopy showed purulent secretions of sinuses, suggestive of pan sinusitis. Serological tests for ANA were negative. There was significantly elevated titre of C-ANCA of 73.2 units. Urine routine analysis showed red cell casts. Nephrologist suggested renal biopsy; however patient refused to undergo the test.

Our patient had classical clinical features and radiological findings of vasculitis with involvement of ENT, lungs and kidneys, which was confirmed by immunological tests of elevated CRP and C- ANCA and hence a final diagnosis of Granulomatosis Polyangiitis (Wegener's) was made. He was started on high dose steroids and cyclophosphamide with an advice on early follow up.

He presented to our department two months later with respiratory distress and saturation of 75% on room air. He was found to be non compliant with medications. Repeat investigations showed hemoglobin of 5g/dl, urine sediment showed red cell casts, C ANCA of 300 units. His chest X-ray showed progressive pulmonary infiltrates suggestive of alveolar hemorrhage. Our patient developed diffuse alveolar hemorrhage and succumbed to the disease.

Discussion

GPA features include necrotizing granulomatous inflammation and pauci- immune vasculitis of medium sized vessels. Circulating antibodies directed against cytoplasmic components of neutrophils (C-ANCA) has been detected in >90% of patients [4]. Anti-neutrophil cytoplasmic autoantibodies specific for proteinase 3 (PR3-ANCA) are implicated in the cause of granulomatosis with polyangiitis and thus also should be specified in the diagnosis [5].

Wegener's granulomatosis is an uncommon necrotizing vasculitis having variable presentations in the chest that are best depicted on high resolution CT. Imaging findings may include nodules, which may cavitate, thin or thick walled cavities; ground-glass opacity secondary to pulmonary hemorrhage; and airway stenosis and ulcerations, ground – glass halo ('CT halo 'sign) or reverse halo ('atoll' sign) can be seen. Our patient had a thick walled cavity in the right lower lobe. Active Wegener's granulomatosis can mimic pneumonia, septic emboli and metastasis [6].

CT imaging can detect the disease even when the radiograph appears normal. In cases where opacities have already been detected in bilateral lung fields on the plain radiograph of the chest, their distribution and characterization is possible

with CT scanning. Moreover, CT has the advantage over biopsy of being noninvasive [7].

Although Wegener granulomatosis is a small vessel vasculitis, aortitis and periaortitis are also reported as in our patient. Periaortic inflammation occurs due to the extension of granulomatous tissue through the vessel wall, whereas granulomatous inflammation in takayasu's arteritis is limited to the vessel wall and it is the vasa vasorum vasculitis in polyarteritis nodosa. Directed extensions of granulomatous tissue into the pulmonary artery can also occur [3]. American College of Rheumatology Criteria for Wegners granulomatoses is mentioned as below [8].

Criterion	Definition
Nasal or oral inflammation	Development of painful or painless oral ulcers or purulent or bloody nasal discharge
Abnormal chest radiograph	Chest radiograph showing the presence nodules, fixed infiltrates, or cavities
Urinary sediment	Microhematuria (>5 red blood cells per high power field) or red cell casts in urine sediment
Granulomatous inflammation on biopsy	Histologic changes showing granulomatous inflammation within the wall of an artery or in the perivascular or extravascular area (artery or arteriole)

Presence of 2 or more of any criteria – positive
Our patient satisfied 3 features of the criteria to consider Wegener's granulomatosis.

Conclusion

A high index of clinical suspicion is important for the diagnosis of vasculitis. Delay in diagnosis can lead to rapid progression of the disease leading to grave prognosis as seen in our patient.

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