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Original Research Paper

Radio-Diagnosis

WEGENER'S GRANULOMATOSIS: A CASE REPORT

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ABSTRACT Wegener's granulomatosis is a rare necrotizing vasculitis with multiple manifestations involving multiple systems. Therefore, pulmonary manifestations are best described on high resolution CT. Imaging findings may include nodules or masses associated with cavity formation; focal or diffuse consolidation - ground-glass opacity secondary to pulmonary hemorrhage. Active Wegener's granulomatosis can mimic pneumonia, septic embolism and metastasis. This case report will illustrate the high resolution CT appearance of Wegener's Granulomatosis.

KEYWORDS : high resolution CT, Wegener's granulomatosis, vasculitis

INTRODUCTION

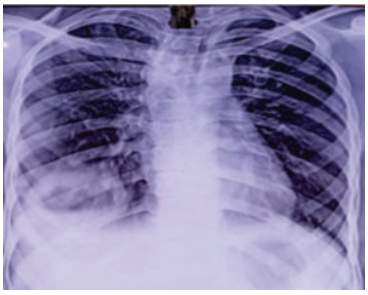
Wegener's granulomatosis is a rare multisystem necrotizing vasculitis. The disorder can occur at any age, but most often affects people between 40 and 60 years of age. It presents with the clinical triad of upper and lower airway involvement and glomerulonephritis. The upper respiratory tract is involved in most of patients and the lungs and kidneys are involved in 90% and 80% of patients, respectively [2]. Other less affected organs include the central and peripheral nervous systems, spleen, and orbit. Without treatment, granulomatous polyangiitis progresses rapidly and is fatal within a year of diagnosis. Appropriate medical treatment improves long-term survival [3].

Case Report

An 18-year-old male presented with a 15 days' history of dry cough, intermittent fever, chest pain associated with loss of weight and appetite. On the basis of these findings and an abnormal chest radiograph showing consolidation and cavity formation, he had been admitted and treated course of different antibiotics, but he had deteriorated further and was not maintaining normal oxygen saturation at room air.Patient had history of haematuria and oliguria in childhood. Now, ultrasound finding shows raised echotexture in bilateral kidneys and hepatosplenomegaly with right lower lung consolidation.

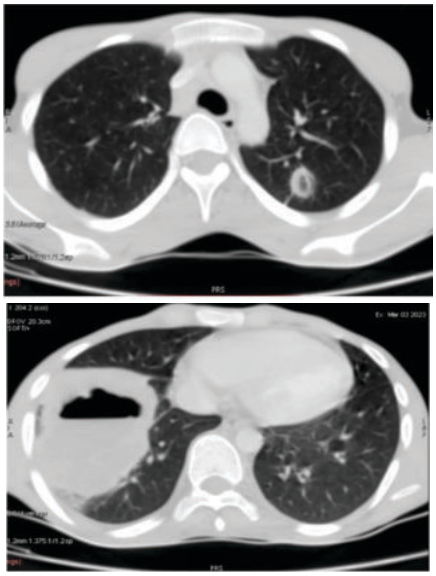
Routine laboratory tests revealed WBC: 8,800 x 109 cumm, haemoglobin: 10.8 gm/dl. The erythrocyte sedimentation rate was 90 mm/hour. C-reactive protein was 48 microgram/ml. Sputum and blood cultures were negative for common pathogens. Urine routine chemical examination shows blood in urine (+++) and microscopic examination shows plenty of red blood cells with presence of epithelial cells. Other biochemical investigations like serum creatinine, serum urea, serum bilirubin, serum electrolytes were in normal range.

Imaging Findings

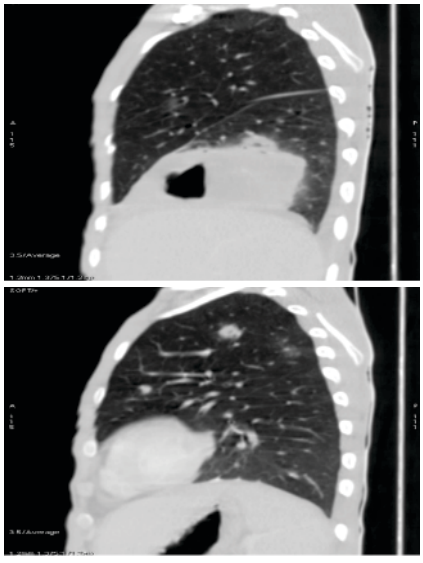


(Figure 1)

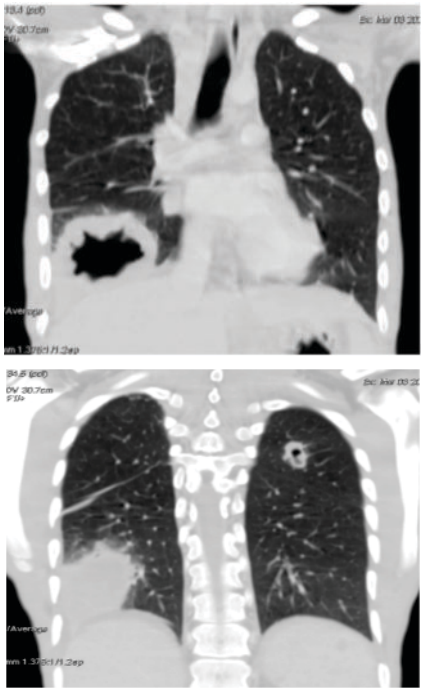
A chest radiograph showed consolidation with thick-walled cavity formation in the right lower lung zone (Figure 1).



(Figure 2) Axial Images



(Figure 3) Sagittal Images



(Figure 3) Coronal Images

Pre and post-contrast conventional chest CT and HRCT were also performed and these (Figure 2, 3 & 4) revealed multiple areas of consolidation with internal thick walled cavitation and surrounding areas of ground-glass opacity involving right lower lobe. Multiple randomly distributed areas of consolidation with surrounding ground glass opacity, randomly distributed predominantly peribronchovascular and subpleural in location involving bilateral lung parenchyma, with mediastinal lymphadenopathy and right minimal pleural effusion.

Abnormal chest radiograph with chest CT findings and patient not improving after appropriate management, based on these findings, Wagener's granulomatosis was suspected and patient was further advised for C-ANCA correlation. C-ANCA correlation was done and a serum test for antineutrophil cytoplasmic antibodies came positive by immunofluorescence (69 IU/ml), thus diagnosis of Wagener's granulomatosis was confirmed.

DISCUSSION

Granulomatous polyangiitis (GPA) also known as Wegener's granulomatosis, is a rare autoimmune disease of unknown etiology involving multiple systems. GPA is antineutrophil cytoplasmic antibody (ANCA) associated vasculitis disorders. It is characterized by necrotizing granulomatous vasculitis of the upper and lower respiratory tract, glomerulonephritis, and small-vessel vasculitis [1].

Ground glass opacities may result from alveolar hemorrhage, mosaic perfusion secondary to small vessel inflammation or necrotic cell infiltration into the alveoli. Interlobular septal thickening may be due to lymphatic congestion or an accumulation of hemosiderin-rich macrophages. Consolidation of Wegener's granulomatosis may be secondary to organizing pneumonia or pulmonary infarction [2].

Patients may have symptoms of more than one system, like pulmonary, renal, CNS and orbital manifestation. In pulmonary presentation of Wagener's granulomatosis, patients may present with respiratory signs and symptoms, including cough, haemoptysis and dyspnoea. History of nasal obstruction, rhinitis, and epistaxis. Renal manifestations include proteinuria and hematuria.

Patient may be treated with multiple course of antibiotics but may not improve, in these case it becomes necessary to diagnose. By clinical features, patient not improving with antibiotics, based on imaging findings, we can help physician for further evaluation of disease to diagnose wegeners and improve patients' management and better survival.

Chest CT findings include nodule, cavity, consolidation and ground glass opacity. Lung consolidation and ground-glass opacity results from haemorrhage [5]. Lung consolidation is often seen in pneumonia and wegener's granulomatosis may be diagnosed when symptoms and consolidation persists despite appropriate management.

High serum titers of c-anti-neutrophil anti-cytoplasmic anti proteinase-3 (c-ANCA) can be used to determine disease activity [3]. Biopsy is gold standard for the diagnosis of WG. Kidney biopsy is the most common method and most often reveals nonspecific glomerulonephritis. Lung biopsy may reveal necrotizing granulomatous vasculitis of small vessels [4].

Treatment consists of immunosuppressive therapy, most often systemic steroids and cyclophosphamide. Remission rates are around 90%, but relapses can occur. Resolution of radiographic signs of Wegener's granulomatosis usually occurs later with clinical improvement [2]. Generally death occur within a year of diagnosis but early diagnosis can help in imrpoving patient's outcome and better survival.

CONCLUSIONS

The findings of Wegener's granulomatosis in high-resolution CT varies according to presentation, ranging from nodules and masses to ground-glass opacity and lung consolidation with cavities. Airway involvement may be focal or extensive. The high-resolution CT findings of Wegener's granulomatosis can mimic other conditions like pneumonia, neoplasm and inflammatory diseases. Although Wegener's granulomatosis is an uncommon disease, recognition of its pulmonary and airway manifestations may help suggest the diagnosis and treatment may helpful in long term survival of patient.

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