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A CASE OF GRANULOMATOSIS WITH POLYANGIITIS MASQUERADING AS LUNG CARCINOMA – A CASE REPORT

Pulmonary Medicine

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ABSTRACT

Granulomatosis with polyangiitis, previously called Wegener's granulomatosis is the most common form of vasculitis to involve the lung. It can lead to diverse clinical manifestations frequently involving the upper airways, lungs and kidneys. We report a 59-year-old male patient, who presented with cough with expectoration and intermittent episodes of blood tinging of sputum for 4 months. Clinically and radiographically a diagnosis of carcinoma of the lung was made. However, biopsy of the mass came negative for malignancy. C-ANCA tested in the patient came positive thus establishing a diagnosis of granulomatosis with polyangiitis. The above case is a rare presentation of Granulomatosis with polyangiitis masquerading as lung cancer.

KEYWORDS

Granulomatosis with polyangiitis (GPA), Wegener's Granulomatosis, Cytoplasmic-Anti neutrophil Cytoplasmic Antibody (C-ANCA)

INTRODUCTION

Granulomatosis with polyangiitis, previously called Wegener's granulomatosis is the most common form of vasculitis to involve the lung. Granulomatous inflammation and necrotizing vasculitis of small sized blood vessels lead to diverse clinical manifestations frequently involving the upper airways, lungs and kidneys[1]. More than 80% of patients with GPA are associated with cytoplasmic immuno-fluorescence pattern (C-ANCA) on ethanol fixed neutrophils that react with proteinase 3(PR3-ANCA). Pulmonary involvement in GPA occurs in 25–80% of cases[2]. The most common radiographic and computed tomography (CT) abnormalities of pulmonary GPA are lung nodules and masses, very often multiple and with cavitation. The varied clinical presentation can make the diagnosis of GPA challenging and makes it difficult to distinguish it from other diseases such as an infection, sarcoidosis, or malignancy. We present a case of a 50, year old male, who presented as lung mass on clinical examination and radiology, later evaluated and diagnosed as GPA.

CASE REPORT

The patient is a 59, year old male, known case of diabetes mellitus for the last 25 years on oral hypoglycaemic agents, presented with chief complaints of cough with scanty mucoid expectoration over last 4 months. Cough was associated with intermittent episodes of blood tinging. History of loss of weight and appetite was present. On presentation, patient was afebrile, heart rate of 90', blood pressure of 120/70mmof hg, respiratory rate of 22 and oxygen saturation of 94%. Physical examination revealed mild pallor and grade III clubbing.

Chest radiograph was performed which showed homogenous opacity with irregular margin in right mid zone silhouetting with right heart border (figure 1). Sputum smear was negative for acid fast bacilli and mycobacterium tuberculosis was not detected on cartridge based nucleic acid amplification test. Based on history, clinical examination and chest radiograph finding a differential diagnosis of carcinoma of lung was made.

Further evaluation with CT of chest showed a large heterogeneously enhancing lesion involving the anterior segment of right upper lobe and medial segment of right middle lobe, measuring 7.2*7.2*6.2cm (AP*TR*SI) in size. There are non-enhancing areas within suggestive of necrosis. Medially lesion is seen to encase the right pulmonary artery, its middle lobar and right upper lobar segmental branches and right superior pulmonary vein. Anterolaterally mass is seen to reach up to pleura. Multiple, enlarged lymph nodes noted in pre-tracheal, right paratracheal, aortopulmonary window, subcarinal and pre-vascular region, largest measuring 3.4*1.5 cm in subcarinal region. Trachea and central bronchi normal. CT findings were suggestive of a likely neoplastic aetiology (Figure 2).

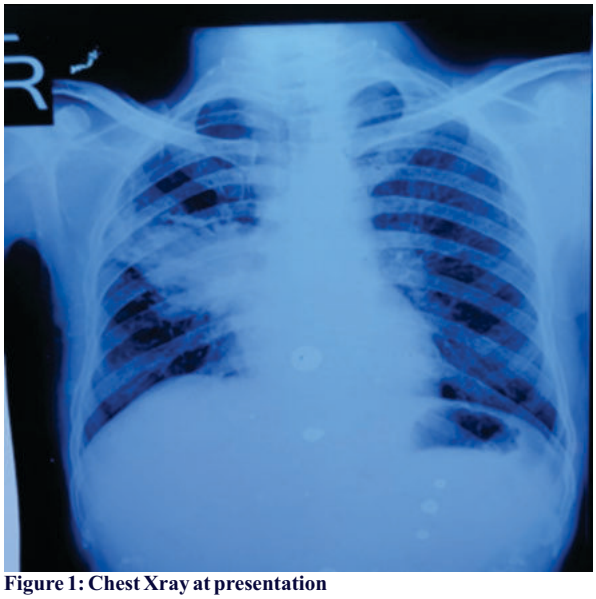

Figure 1: Chest Xray at presentation

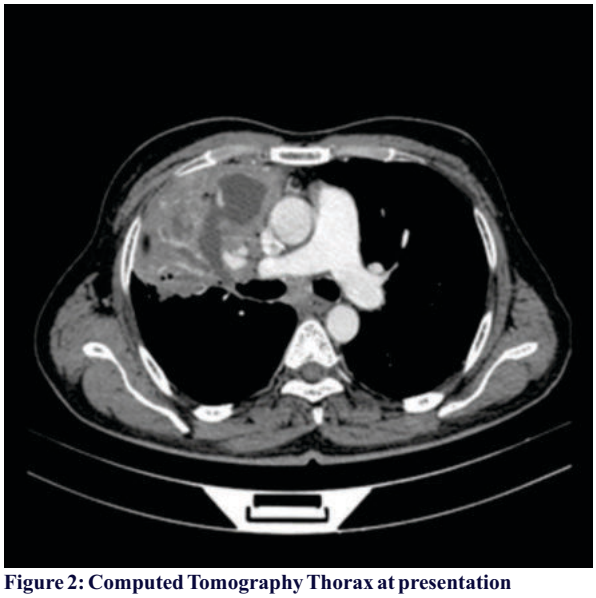

Figure 2: Computed Tomography Thorax at presentation

Video bronchoscopy done also opined the nodular lesion present in anterior segment of right upper lobe as? lung carcinoma.

PET CT done a month later showed hypermetabolic mass with spiculated mass in right anterior lobe extending in to right middle lobe suggestive of primary tumour and metastasis to mediastinal lymph nodes.

However, biopsy of the mass revealed only blood clots with dense acute on chronic inflammatory infiltrate. Occasional blood vessels showed evidence of vasculitis with neutrophilic infiltrate in the wall. Immunohistochemistry also showed unremarkable glandular tissue showing lymphoplasmacytic infiltration and fibrosis. Aggregates of lymphoid tissue and perivascular collagen are noted along with vascular proliferation, favouring non-specific interstitial pneumonitis and underlying vascular and inflammatory pathology.

C-ANCA tested in the patient came positive thus establishing a diagnosis of granulomatosis with polyangiitis, aka Wegener's granulomatosis.

The patient was started on remission induction therapy consisting of oral prednisolone at dose of 1 mg/kg per day in combination with oral cyclophosphamide at a dose of 1 mg /kg for a duration of 6 months. Repeat chest radiograph done at end of remission induction therapy showed good improvement and patient was shifted to remission maintenance therapy with oral azathioprine 50 mg od for one year. Repeat chest radiograph (figure 3) and CT thorax (figure 4) done at end of treatment showed good clearance.

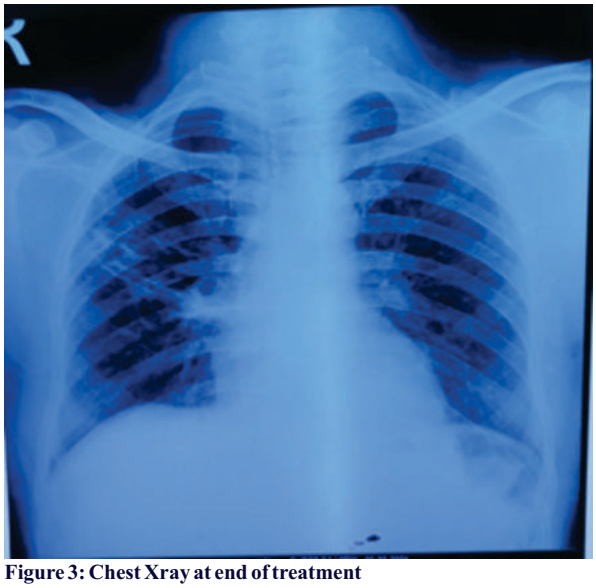

Figure 3: Chest Xray at end of treatment

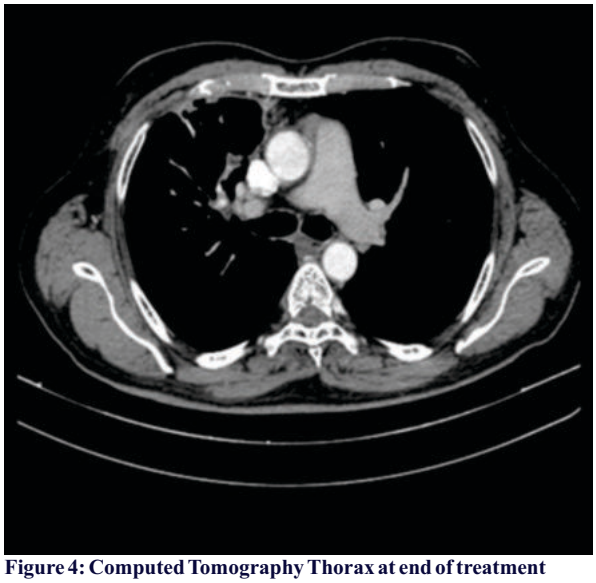

Figure 4: Computed Tomography Thorax at end of treatment

DISCUSSION

GPA is currently characterized as one of the ANCA-associated small vessel vasculitis. It is distinguished clinically by its predilection for affecting the upper and lower respiratory tracts and kidneys and by the histologic presence of necrosis, granulomatous inflammation, and vasculitis[3]. The etiology of ANCA associated vasculitis remains unknown. Several different pathways and mechanisms have been proposed for the pathogenesis. ANCA are predominantly immunoglobulin G (IgG) autoantibodies directed against constituents of neutrophil's primary granules and lysosomes of monocytes. Two different kinds of ANCA can be distinguished according to their fluorescence pattern: cytoplasmic (c)-ANCA and perinuclear (p)-ANCA[4]. In more than 80% of patients with GPA, ANCA occur and are associated with a cytoplasmic immunofluorescence pattern (C-ANCA) whereas perinuclear pattern (P-ANCA) occurs in fewer than 10% of patients with GPA.

Sex distribution of the disease is equal, most of the patients present in the fifth decade, although the disease can occur at any age. Clinical presentation can be so diverse that the list of differential diagnoses is vast, ranging from infection, neoplasm, tuberculosis, malignancy, other forms of vasculitis like sarcoidosis, Behcet's disease etc. Pulmonary involvement in GPA occurs in 25–80% of cases. Pulmonary involvement in GPA usually presents radiographically as nodules or mass lesions, which may cavitate. Inflammation and stenosis of the tracheobronchial tree occur in at least 15% patients with lung involvement. Pleural effusion may occur, but are usually small, asymptomatic and incidental finding. Atypical presentations are interstitial lung disease, hilar mass or pneumothorax[5]. In this case based on history, clinical findings and radiological features a differential diagnosis of lung carcinoma was made, however biopsy showed no evidence of malignant cells. The presence of C-ANCA positivity helped in coming towards the diagnosis of GPA.

Traditionally, initial therapy of GPA consists of daily oral cyclophosphamide-corticosteroid combination therapy. This treatment has been quite effective in inducing remission in more than 90% of patients who adhere to this regimen; approximately 75% experience complete remission[6]. Other therapies for GPA include methotrexate and prednisone, which is another alternative for patients with active but not immediately life-threatening disease and normal or near-normal renal function[7]. Prednisone alone is not a recommended therapy for GPA. Rituximab is another agent that can be used in place of cyclophosphamide in severe ANCA -associated vasculitis. Once remission induction is achieved remission maintenance therapy is continued for at least 12 months. Azathioprine is usually the agent of choice. Mycophenolate mofetil is an alternative for patients who do not tolerate azathioprine.

CONCLUSION

The above case is a rare presentation of GPA masquerading as lung cancer. Hence it is a learning lesson in clinical practice to always consider the possibility of alternate diagnosis in each case.

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