Case Report

Wegener’s Granulomatosis : An Isolated Lung Mass Responding to Antituberculosis Therapy and Atypical Course

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Abstract
We describe an unusual case of Wegener's granulomatosis (WG), in a middle aged, non-smoking female who presented with a lung mass with constitutional symptoms. FNABC from mass revealed a single ill-defined granuloma without necrosis. There was a definite clinical and radiological response to antituberculosis treatment. She was later found to have another mass lesion in nasopharynx. ANCA was negative initially but became positive once disease flare up occurred. Multisystem involvement with clinical features of vasculitis were seen during the flare up and resulted in a fatal outcome. Unusual features and literature on this entity is discussed.

INTRODUCTION

Wegener's granulomatosis is an uncommon disease with a wide spectrum of organ involvement and severity. Diagnosis is usually considered in a patient with classic triad of organ involvement that is upper and lower respiratory tract and kidneys. Outcome is good when treatment is started early using corticosteroids and cytotoxic agents. We came across a patient with this disease who had an atypical presentation and showed clinical as well as radiological response to anti-tuberculosis therapy. This case is reported with the aim of highlighting the importance of keeping a high index of suspicion in patients with atypical profile so that timely diagnosis of this potentially treatable disease could be made.

CASE REPORT

A 50 year old female presented with low grade continuous fever which was associated with cough and scant expectoration of one month duration. She also complained of pleuritic pain in left lower chest, anorexia and significant weight loss. There was no history of haemoptysis, purulent expectoration, joint pains or swelling of feet. On examination there was mild pallor and chest examination revealed decreased breath sounds in left infra-axillary and infra-scapular areas. On investigation, haemoglobin was 95 gm/L, TLC was 15,900/cmm with 81% neutrophils and ESR was 91 mm/1st hour (Westergren). Renal functions, liver functions and urine examination were normal. Chest radiograph showed a sharply demarcated homogenous opacity in left lower zone suggestive of a mass lesion (Fig. 1A). Three sputum samples each for acid-fast bacilli (AFB) and malignant cytology were negative. Mantoux’s test was negative. CT chest showed mass lesion in left lower lobe with minimal pleural effusion (Fig. 1C). There was no mediastinal lymphadenopathy or bone destruction. FNABC from lung mass showed no malignant cells but few inflammatory cells with an ill-defined granuloma. Stains for AFB and fungi were negative. Bronchoscopy was normal. Bronchial washings revealed acute inflammatory cells and were negative for AFB, fungi and malignant cells. A thoracoscopic or open lung biopsy was refused by the patient.

A trial of anti-tubercular treatment (ATT) was planned and patient was started on four drug regimen (rifampicin, isoniazid, ethambutol and pyrazinamide). One and a half months later, patient felt better and a repeat chest radiograph showed significant improvement (Figs. 1A and B). ATT was continued.

One month later patient returned with decreased hearing in left ear. On ear examination the tympanic membrane was retracted and lusterless. Pure tone audiogram showed moderate conductive hearing loss (40 dB). Impedance audiometry showed type B curve signifying middle ear effusion. A high resolution CT of temporal bone and nasopharynx revealed a bulge in the nasopharynx on left side caused by mass lesion in left nasopharynx extending to left middle ear through the eustachian tube (Fig. 1D). X-ray

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Over the next two days patient developed right 3rd cranial nerve paralysis. A CECT head was normal. One-day later gangrene of left hand appeared with mononeuritis involving left common peroneal nerve and patient developed multiple vasculitic ulcers and palpable purpura on the limbs. She had bouts of haemoptysis and went into respiratory failure. Haemoglobin fell from 91 gm/L to 67 gm/L over one day. Chest radiograph was consistent with diffuse pulmonary haemorrhage. Urine examination for the first time showed proteinuria (24-hrs albumin - 2.8 gm) with active sediment. A repeat ANCA was done and this time it was strongly positive with cytoplasmic pattern.

A diagnosis of Wegener’s granulomatosis with CNS, renal, cutaneous involvement and diffuse pulmonary haemorrhage was made. Patient was initiated on methylprednisolone (1 gm/day for 3 days) and cyclophosphamide (900 mg/day). Four days later patient developed right lower lobe pneumonia. Adequate antibiotics were started but patient went into sepsis with ARDS. She required positive pressure ventilation but kept deteriorating and succumbed three days later.

**DISCUSSION**

Our patient with WG had an atypical presentation and very unusual course which led to delayed diagnosis and treatment. The unusual features included presentation with a lung mass with pleural effusion. Also, ANCA was negative till she had a fulminant flare up. This kind of flare up with multisystem involvement is not described earlier. Absence of vasculitis on repeated biopsies was also unusual.

The usual lung involvement in WG is with fleeting shadows, which are commonly nodular with cavitation occurring occasionally. WG presenting as mass lesion is rare. On the other hand various forms of other systemic vasculitides have also been reported to be associated with vasculitis (Fig. 2).
tumour-like lesions. Kariv et al1 analysed 79 cases of systemic vasculitides presenting as tumour-like lesions of which 28 cases had WG. Lung is not a favoured site for mass lesion in WG as there were only three cases with lung involvement in Kariv’s series.2 In fact most common site for mass lesion was found to be breast (7 cases), which is not a usual site for involvement in WG. This may signify a different pathogenetic mechanism in the formation of mass lesion. Our case is unique as there was mass lesions at two classical sites of involvement in WG which is very rare.

Our patient was given ATT in the beginning. The decision to treat with empirical ATT was taken in view of presentation with low grade fever, weight loss and pleural effusion, finding of an ill-defined granuloma, high prevalence of tuberculosis in our area and unwillingness on the part of patient to undergo thoracoscopic biopsy. She showed a definite clinical as well as radiological response to ATT. Response to ATT has been earlier described by Toyoshima et al.2 This was thought to be due to antimicrobial effect of one of its constituent, possibly rifampicin.3 It has been shown that nasal carriage of Staphylococcus aureus in patients with WG is associated with higher relapse rate.3 It was postulated that treatment with antimicrobial agents could lead to eradication of these microbes and may reduce relapses. In a double blind, placebo controlled trial, Stegeman and co-workers, showed that relapses of WG could be significantly reduced with trimethoprim/sulfamethoxazole.4 We feel that in our case also response to ATT was possibly mediated through antimicrobial effect of one of its constituents. Although it is suggested that rifampicin may have immunosuppressive effects which may be responsible for inducing remission,4 this appears very unlikely. The fact that case reported by Toyoshima et al5 eventually responded to another antimicrobial agent (trimethoprim/sulfamethoxazole) is also an evidence of efficacy of ATT through its antimicrobial effect.

In our patient ANCA (ELISA technique) done at the time of detection of nasopharyngeal mass, when lung mass was already present, was negative. At this point of time our patient would be classified as having limited form of WG. The sensitivity of ANCA in limited form of WG (67-86%) is far less than that in the classic form (> 90%) when kidney involvement occurs.6 However, ANCA became positive when disease flared and kidney involvement occurred.

Finally, the fulminant flare up with appearance of florid, widespread features of WG is rather unusual and is not described in the literature. The aim of this communication is to highlight the extremely atypical features, which might be associated with this disease. With the availability of highly effective treatment of WG, every effort must be made to diagnose it at an early stage and this kind of clinical presentation should be kept in mind.

**REFERENCES**