Wegener's granulomatosis: Is it rare?

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Summary

A 52 year old lady was admitted with fever, productive cough and breathlessness for one month. She had past history of recurrent sinusitis. Clinically, she had bilateral sensorineural hearing impairment, nasal crusts and peripheral neuropathy apart from signs of consolidation. Vasculitic lesions appeared a day after admission. Investigations revealed multiple nodular like lesions scattered in both lungs on CXR. Blood tests revealed positive cANCA (Cytoplasmic fluorescence Antineutrophil Cytoplasmic Antibodies) and raised inflammatory markers. She was treated with immunosuppressant and antibiotic. Clinical and radiological improvement was noted 2 weeks later.

Background

Wegener's granulomatosis is an uncommon multi-system disease that is characterized by necrotizing granulomatous inflammation affecting predominantly the upper and lower respiratory tract and kidneys. It is rarely seen in Myanmar and its presentations occasionally mimic other diseases such as malignancy.

Case presentation

A 52 year old lady belonging to the Karin ethic race was admitted to the hospital with fever and cough for one month and with progressive breathlessness for 3 weeks. She is a non-smoker. Since one month prior to admission, she developed low grade fever with evening rise in temperature and cough with mucopurulent sputum production. But she did not give any history of haemoptysis, wheezing, chest pain and hoarseness of voice.

At the time of admission, she was conscious, temperature was 100°F, respiration rate 20/min. All other vital signs were stable and there was no cyanosis, clubbing, lymphadenopathy and edema. Reduced chest wall movement was seen. Dull percussion note and crepitations in the right upper and middle zones were noted on chest examination. Normal cardiac and abdomen examinations were reported. Bilateral sensorineural hearing impairment, nasal crust and peripheral neuropathy were also noted. On review of her past history, she had recurrent attacks of sinusitis. On second day of admission, vasculitic lesions appeared over the extremities (Figure 1, 2 & 3).

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Chest X ray was done and it showed multiple masses or nodular lesions of variable sizes scattered in both lungs with consolidation of right upper and middle lobes (Figure 4). Chest CT scan showed 5 × 7 cm radio opaque mass in the right upper lobe with enlarged right hilar and sub-carina lymph nodes along with metastatic lesions in both lungs (Figure 5). She was first diagnosed as having pulmonary metastasis with chest infection. Further investigations were done to find out the source of primary tumor and source of infection (Table 1). While undergoing further investigations she received non-specific antibiotic therapy without significant improvement.

Investigations

Because of the above signs and symptoms, Wegener's granulomatosis was suspected. ANCA were done and c ANCA was found out to be positive. ESR and CPR were also raised significantly.

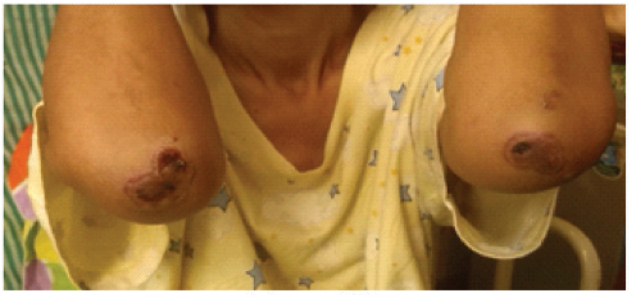
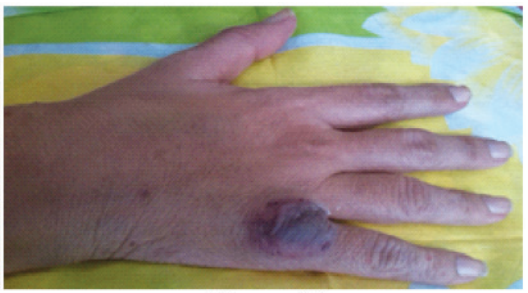


Figure (1)



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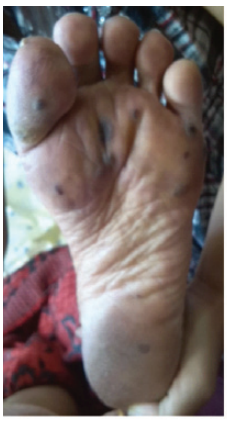

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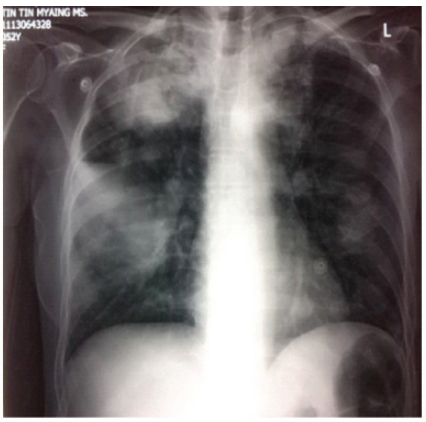

Figure (4)

Figure (5)

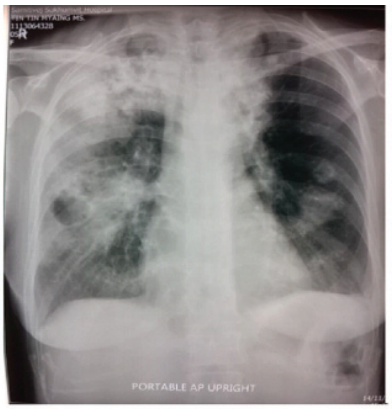

Figure (6)
CXR at time of admission

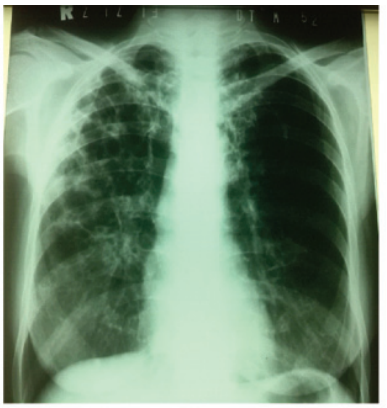

Figure (7)

Table 1. Tests performed on the patient and corresponding results

Tests	Result
AFP	0.68 ng/ml (normal)
CA 19 - 9	9.92 U/ml (normal)
CA 153	14.9 U/ml (normal)
CEA	1.78 ng/ml (normal)
USG (abdomen and pelvis)	No abnormality
LDH	Normal
CT guided FNAB	Soft tissue with mixed inflammatory cells infiltrate including lymphocytes, plasma cells, histocytes with few multinucleated giant cells and patchy foci of necrosis. No malignancy seen
Bronchoscopy	No intra-luminal growth
Bronchial washing cytology	Mixed neutrophil, mild alveolar macrophage and tiny cluster of benign bronchial epithelial cell. No malignant cell
Bronchial washing AFB & G stain	No organism
Sputum Culture	Growth of normal flora, no fungus, no AFB
TB PCR	Negative

Treatment

IV Coamoxiclav and oral azithromycin were given along with oral cyclophosphamide and steroid therapy.

Outcome

After two weeks of therapy, the patient improved clinically. Radiological improvement was also noted (Figure 7). She was discharged taking oral steroid and cyclophosphamide only and was asked to come back for follow-up two weeks later.

Discussion

Wegener's granulomatosis was first described by Klinger in 1933 followed by Wegener in

1936 and 1939 and Ringertz in 1947. It is a systemic disorder that involves both granulomatosis and polyangiitis. It is a form of vasculitis that affects small- and medium-size vessels in many organs. Damage to the lungs and kidneys can be fatal. According to reports in the literature, lungs are affected in 90% of these patients. In 2011, ACR (American College of Rheumatism) and EULAR (European League Against Rheumatism) regarded Wegener's granulomatosis as a category of "**Granulomatosis with polyangiitis**". Although the presence of autoantibody to cANCA is not required for diagnosis according to ACR, it is a valuable tool in the absence of histopathology for Wegener's granulomatosis. According to ACR criteria, two or more positive criteria are needed for diagnosis. The ACR criteria are as follows:

1. Nasal or oral inflammation:
   - painful or painless oral ulcers or purulent or bloody nasal discharge
2. Lungs: abnormal chest X-ray with:
   - nodules, infiltrates or cavities
3. Kidneys: urinary sediment with:
   - microhematuria or red cell casts
4. Biopsy: granulomatous inflammation
   - within the arterial wall or in the perivascular area

In this case, the patient was first diagnosed as suffering from pulmonary metastasis due to its radiological lesions mimicking cancerious signs. Since the patient was a female having multisystem involvements with vasculitis lesions, the diagnosis of autoimmune disease was considered. Finally, the diagnosis was confirmed by the patient's nasal symptoms, radiological findings and supportive investigations.

Take Home Messages

- Although Wegener granulomatosis is a rare disease especially in Myanmar, a high level of suspicion must be kept in mind when a patient presents with vasculitis, upper airway symptoms and necrotizing pneumonia.
- High index of suspicion is important.

References

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