Case Report

Wegener's granulomatosis

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Abstract

Granulomatosis with polyangitis is an uncommon disorder that causes inflammation of the blood vessels in nose, sinuses, throat, lungs and kidneys. Granulomatosis with polyangitis formerly called Wegener's granulomatosis.

Wegener's granulomatosis is a rare disease, characterized by necrotizing granulomatous inflammation of important organ of the body. This inflammation damages important organ of the body by limiting blood flow to those organs and destroy normal tissue.

Although the disease can involve any organ system, Wegener's granulomatosis mainly affects the upper and lower respiratory tract, kidneys. Skin, Eye, Musculoskeletal system and nervous system may also be affected.

The prevalence of the disease is 3 persons per 1,00,000/- equally in both sexes. The German pathologist Friedrich Wegener first described the disease in 1936.

In 1954, Godman and Churg more fully described the disease. They established three main clinical criteria of WG (Vasculitis, glomerulonephritis, respiratory tract involvement).The manifestation of this disease and organ involvement may vary widely. We report an unusual case of WG in which the patient presented with multiorgan involvement (Nervous system, Respiratory tract, Skin, Eye, Liver, Kidney)and most acute presentation of them is diffuse alveolar hemorrhage.

Case presentation

A 27 years old male was admitted to the SSMC & Mitford hospital (Dec 2012) with the history of progressive worsening cough, hemoptysis and shortness of breath. One and half month prior to the admission he had developed sudden onset vertigo and red eye, multiple rash in different parts of the body. Over the last one and half months he took consultation from Neurologist, ophthalmologist and chest physician for the above problems. Other past medical history was non contributory and he reported no toxic habit but use of Metamphetamine occasionally.

Due to significant hypoxia and repiratory distress, the patient was admitted in Medical ICU. On admission patient was concious, his body temperature was 37.5°C, pulse 110/min, respiratory rate 30/min, blood pressure 170/90 mmHg. He was anxious, severely pale. His face was edematous, both eyes were congested and multiple purpuric spot in both the upper and lower limbs. On auscultation of the both lungs there were coarse crepitation over whole lung field in the right side. His abdomen was neither tender nor distended, there were no cyanosis or clubbing of the extremeties but edema was significant.

Table-1: Laboratory parameters

Peripheral blood	
WBC	25.0×109/L
RBC	2.1×1012/L
Hemoglobin	5.0 gm/dl
Platelet	686.0×109/L
ESR	120 mm (1st Hour)
Biochemical Parameter	
CRP	25.3 mg/dl
Glucose	11 mmol/L
Urea	84 mg/dl
Creatinine	4.33 mg/dl
SGPT	77 IU/L
SGOT	38 IU/L
Alkaline Phosphatase	129 U/L
S. Albumin	2.4 g/dl
S. Calcium	8.5 mg/dl
S. Magnesium	1.4 mg/dl
S. Uric acid	7.5 mg /dl
S. Sodium	129 m mol/L
S. Potassium	5.3 m mol/L
S. Chloride	91 m mol/L
Urine Analysis	
RBC	8-10 /HPF
Pus Cell	6-8 /HPF
Albumin	(+)
Cast	Granular

Table-2: Immunological analysis

Immunological parameters	
Antinuclear antibodies	Negative
C_3	1.32 g/L
C_4	0.52 g/L
Anti phospholipid IgG	2.26 U/ml
Anti phospholipid IgM	2.65 U/ml
C-ANCA	56.81 U/ml
P-ANCA	3.15 U/ml

Chest X ray showed there was nodular lesion in the right lower zone (figure-1). CT scan of the chest and CT guided FNAC was done later on and the histopathology report suggested that it was granulomatous lesion.

MRI of the brain showed right sided medullary infarct (figure 3). His admission chest radiograph revealed significant interval change consistent with diffuse alveolar hemorrhage in the right side (figure-2).

Sputum for TB PCR, Pneumocystic carinii and fungal smear were negative.

Ultrasonogram of the abdomen showed increased cortical echo with hypoechoic renal pyramids with diffuse fatty change in liver.

Direct immunofluroscence of the skin biopsy showed necrotizing vasculitis (Fig 4&5).

Hematological test showed leucocytosis with severe microcytic hypochromic anemia with thrombocytosis.

On admission we started Inj. Methylprednisolone with other supportive therapy follwed by oral prednisolone.Afetr getting the Skin biopsy report and immunological parameter, we started Inj. Cyclophosphamide pulse therapy. Patient was improved clinically, oxygen requirement was minimum during shifting of the patient from ICU. But radiological improvement of the chest was minimum.

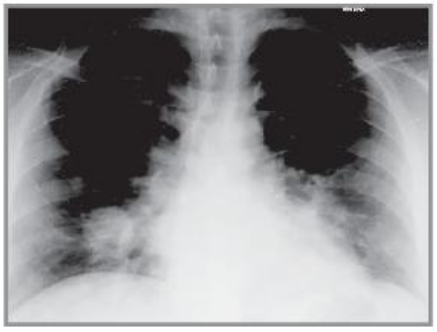

Figure-1: (Nodular lesion in rt lower chest)

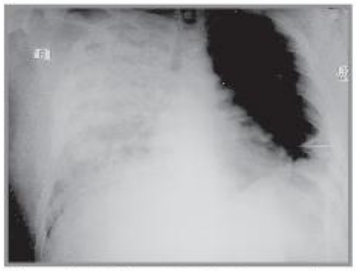

Figure-2: (Diffuse alveolar haemorrahage)

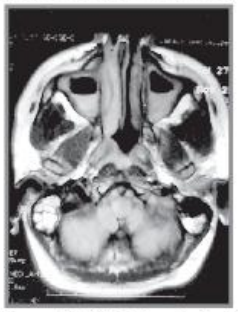

Figure-3: (Medullary infarct)

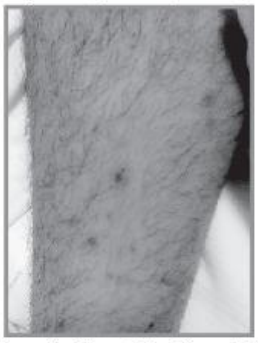

Figure-4: (Necrotising Vasculitis)

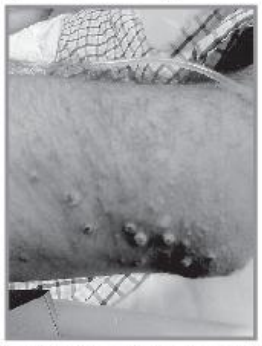

Figure-5: (Necrotising Vasculitis)

Discussion

WG is classified as ANCA positive vasculitis, mostly localized on the small and medium sized blood vessel. It mostly affects the respiratory airways and kidneys.

A characteric feature of the inflammation in Wegener's granulomatosis is the presence of discrete groups of inflammatory cells called granuloma.The disease can affect people at any age (5-90 years) with approximately 15% of cases eginning before 20 years of age.Wegener's granulomatosis strikes both men and women.

It is most common in Caucasian (97%) and is rare in African American (2%).

Although Wegener's granulomatosis may involve other body system, the classic features are vasculitis, glomerulonephritis and granuloma of the respiratory tract.

90% to 95% of patient have lung or sinus disease and involvement of the respiratory tract (sinuses, nose, trachea and lung) is the first sign of the disease in 90% of patients.

85% of patient with Wegener's granulomatosis suffer from kidney disease like glomerulonephritis which causes hematuria and proteinuria.

50%of patients with Wegener's granulomatosis develop skin lesion that appear as small area of rashes, ulcers, nodules. In some patients, the initial symptom of wegener'granulomatosis may include fever, malaise, anorexia, weight loss, joint pain and night sweats.

16% of patients may present first with eye symptom (Excessive lacrimation, retro-orbital pain, diplopia, proptosis). Proptosis and involvement of the respiratory tract disease is highly suggestive of Wegener's granulomatosis.

In 1990, the American College of Rheumatology established the criteria for the classification of Wegener'granulomatosis: Nasal or oral inflammation, Radiologically demonstrated pulmonary infiltrates, abnormal urinary sediment, Granulomatous inflammation on biopsy. Patient shall be said to have Wegener's granulomatosis if at least 2 of these 4 criteria are present. The presence of autoantibodies to proteinase 3/ cANCA is not required for diagnosis of Wegener's granulomatosis.

Clinical presentation can be so diverse that the list of differential diagnosis is vast, ranging from infection, neoplasm, tuberculosis, malignancy, other form of vasculitis (sarcoidosis, Behcet disease, Henoch Scholain purpora.)

In our case, we described an unususal and life threatening clinical syndrome known as diffuse alveolar hemorrhage preceded by nodular lesion in the lung, which is not a typical finding in Wegener's granulomatosis and make a diagnostic problem.

This Patient's progressive multisystem complaints over a period of months, along with elevated ESR, anemia and thrombocytosis, strongly supported the diagnosis of systemic vasculitis.

This patient's clinical course, as well as strongly positive cANCA with negative RF, ANA, pANCA titre were considered diagnostic of Wegener's granulomatosis.

Our patient had manifested many typical Wegener's granulomatosis symptom and sign including Ischemic stroke, hemoptysis, hematuria, scleritis, purpuric and nodular lesion in the skin.

Differential diagnosis of tuberculosis and Wegener's granulomatosis is a great problem.

In both cases, the clinical findings include haemoptysis, sub-febrile temperature, hematuria, Radiological presentation can be the same in both diseases.

Even histopathologic finding can make confusion, since both diseases have granulomatous changes.

In addition, the cases of positive ANCA antibodies have been described in those suffering from tuberculosis. Sometimes, patient with Wegener's granulomatosis were mistakenly treated for pulmonary tuberculosis, and Wegener's granulomatosis was diagnosed when the patients failed to respond to antituberculosis drugs.Toyoshima et al.(12) presented a case of good therapeutic response and restitution of granuloma after the applied antituberculosis therapy.

However, in our case, therapy response was obtained after the applied immunosupressive therapy.

Even though classified in the group of rare pulmonary disease, early diagnosis of disease is of particular significanse for prognosis in the patients with Wegener's granulomatosis.

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