

# A Rare Case of Wegner's Granulomatosis Misdiagnosed as Pulmonary Koch's in Pregnant Female

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**Abstract:** *Granulomatosis with Polyangiitis (GPA), formerly known as Wegener's Granulomatosis, is a rare form of vasculitis affecting small-sized blood vessels in the upper respiratory tract (nose, sinuses, trachea, ears), lower respiratory tract (bronchus and lungs) and kidneys with estimated prevalence of 3 per 100000. Pregnancy associated with Wegener's granulomatosis is rare. Here we present a case of a 25 years old pregnant female with complains of not perceiving fetal movements, epistaxis, scanty haemoptysis, cough and fever. Initially patient was misdiagnosed as Pulmonary Koch's, but further study revealed it to be a Wegener's Granulomatosis.*

**Keywords:** Wegener's Granulomatosis, Pregnancy, Hemoptysis, Cyclophosphamide

## 1. Introduction

Wegener's Granulomatosis (Granulomatosis with Polyangiitis) is an uncommon systemic vasculitis of unclear pathophysiology characterized by necrotizing granulomatous inflammation of upper respiratory tract (nose, sinuses, trachea, ears) and lower (bronchi and lung) respiratory tracts and general focal necrotizing vasculitis<sup>1</sup>. The incidence of WG peaks in 4<sup>th</sup> and 5<sup>th</sup> decade with slight male predominance. Hence, reports of pregnancy in patients with WG are few. As more effective therapies for inducing and maintaining remission emerge, more patients will fall pregnant and the management of Wegener's in pregnancy will become increasingly important.

Wegener's Granulomatosis can be diagnosed if at least 2 of the 4 criterias are present:

- 1) Nasal or oral inflammation with development of painful or painless or bloody nasal discharge.
- 2) Abnormal chest radiograph showing the presence of cavities, fixed infiltrates or nodules.
- 3) Urinary sediments: proteinuria, hematuria, red cell casts
- 4) Granulomatous inflammation on biopsy<sup>2</sup>. If biopsy is not available, haemoptysis can be substituted as fourth criterion.

## 2. Case Report

We describe a case of 25 years old 34 weeks pregnant female admitted in obstetrics & gynecology department with complain of not perceiving fetal movements. Intra uterine death was confirmed by USG and delivered vaginally. On next day she complained of epistaxis, scanty haemoptysis, cough and fever.

A Chest x-ray was performed which showed multiple cavities in bilateral lung fields. [Fig.1, 2] So she was transferred to respiratory medicine department with the possible diagnosis of Pulmonary Koch's.

Total WBC counts were 31,500 with ESR 110. Sputum AFB and Gene-expert (CBNNAT) for Tuberculosis were done which showed negative results. Sputum culture was also negative. Upper respiratory tract examination was normal.

CECT Chest was performed which showed septic pulmonary emboli more likely over granulomatous lesion with polyangiitis.[Fig 3,4] Due to high possibility of septic emboli, initially patient was given higher antibiotics but no improvement seen in fever and chest x-ray.

USG pelvis and 2D echo were normal.

After 3-4 days, she developed multiple pustular skin lesions over dorsum of right hand & fingers (microscopic polyangiitis) and watering of eyes. So skin biopsy was taken which was also suggestive of Wegener's Granulomatosis. [Fig 5, 6]

Initially urine report was normal but gradually it showed proteinuria. So CECT abdomen was also performed later on to monitor kidney abnormality which showed enlarged bilateral kidney and multiple granulomatous lesions in spleen.

At last laboratory investigation of serum cytoplasmic anti-neutrophilic antibody (C-ANCA) and C- reactive protein (CRP) were also done which showed positive results.

From all laboratory investigations along with radiological findings and skin biopsy results we made the diagnosis of Wegener's Granulomatosis.

So patient was put on combined therapy of Prednisolone and Cyclophosphamide, which yielded positive result and provided symptomatic relief to the patient.



Figure 1: Chest X-ray suggestive of multiple cavities

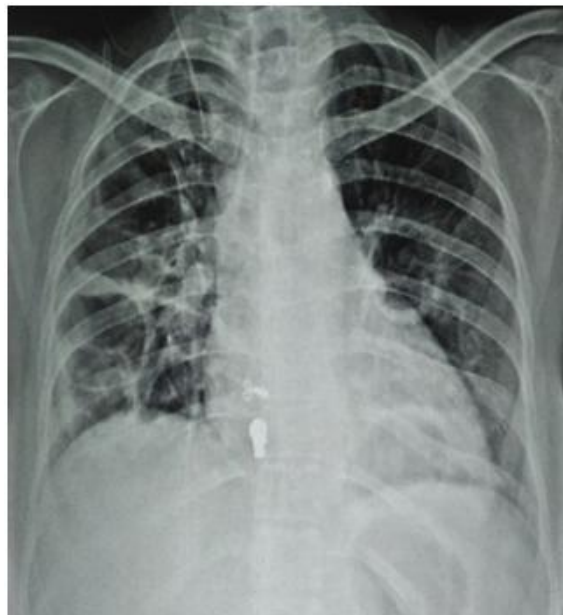


Figure 2: After treatment of 2.5 months

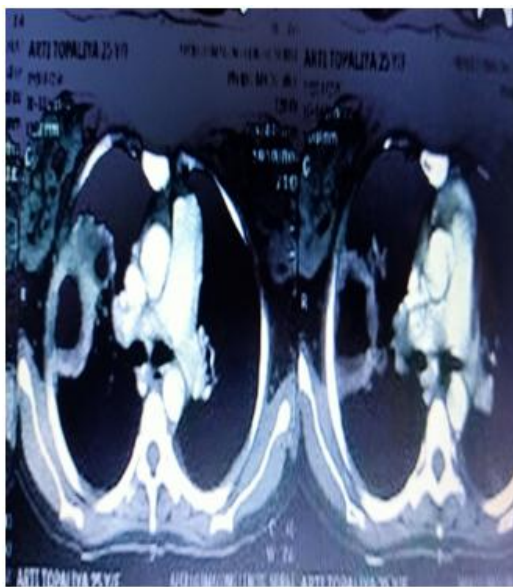


Figure 3 & 4: CECT Chest suggestive of Cavitory lesions



Figure 5 & 6: Pustular Skin Lesions over dorsum of finger and hand

### 3. Discussion

Classified as necrotizing granulomatous vasculitis of small and medium vessels, granulomatosis with polyangiitis (GP) is a rare multisystem disease.<sup>3</sup>

Since 2013, due to the recommendations of the American College of Rheumatology (ACR), the American Society of Nephrology (ASN) and the European League Against Rheumatism (EULAR), a new Chapel Hill Consensus Conference (CCHC2012) updated the classification of Wegener's granulomatosis to granulomatosis with polyangiitis.

The involvement of the upper respiratory tract occurs in 50-80% of cases, and mainly in the form of chronic sinusitis.<sup>5</sup> Studies show that skin manifestations may appear in 16% to 77% of cases and with varied presentations<sup>4,5,6,7</sup> Papulonecrotic lesions represent the most common injuries and they occur mainly in the lower limbs.<sup>5,7</sup> Involvement of skin and mucosa, alone, feature a rare variant called localized granulomatosis.<sup>8</sup>

Renal vasculitis, necrotizing and granulomatous, happens in less than 50% of cases of GP. These vasculitis are late and don't occur in the limited form of the disease. The most frequently observed renal damage (75% to 80% of patients), though not decisive in GP, is the focal and segmental necrotizing glomerulonephritis and it may, in some cases, evolve into generalized glomerulonephritis. Vasculitis, in its granulomatous form in the renal tissue, is very rare.<sup>4,9,10</sup>

An important aspect in the diagnosis of GP is its antineutrophil cytoplasmic antibody profile. Originally these antibodies are associated with necrotizing vasculitis and they can be subdivided according to its stimulating antigen.<sup>3,9</sup> There are currently 2 main classifications for this class of antibodies: specific pattern for protein myeloperoxidase (MPO-ANCA) of perinuclear presentation (p-ANCA) and the specific pattern for proteinase 3 (PR3-ANCA) of cytoplasmic display (c-ANCA).<sup>9</sup>

Early treatment of remission induction is crucial to reverse the renal damage, and it is performed in 40% of cases within the first 3 months of symptoms, relying on the use of corticosteroids and cyclophosphamide.<sup>3,4,11</sup> Currently, hopes are directed towards immunobiological agents. Among them, the most studied is Rituximab, which found a place in the current EULAR recommendations as an alternative drug for refractory disease, being used at a dose of 375 mg/week for 4 weeks.<sup>12</sup> Another alternative in the treatment of remission is methotrexate 15-25 mg/week combined with folic acid. Maintenance can be done with azathioprine 2 mg/kg/day; Leflunomide 20-30 mg/day or Rituximab 1g IV every 6 months for 2 years.<sup>12</sup>

The management of WG during pregnancy has to be on an individual basis. With the potential life threatening consequences of WG, an aggressive approach to the treatment of the disease in pregnancy should be adopted. The choice of drugs will depend on the stage of pregnancy and the severity of the disease flare. The teratogenic effects of Cyclophosphamide have been reported previously<sup>13</sup> and it

should be used with caution particularly early in pregnancy. However, there is evidence that it can be safely used with steroids in the second and third trimesters of pregnancy.<sup>14</sup>

In our patient the diagnosis of WG was made on the basis of characteristic clinical, immunological, and radiographic findings in the absence of biopsy findings and she responded well to treatment.

### 4. Learning Points

- Wegener's granulomatosis is rare in pregnancy but needs to be considered as a differential diagnosis in patients presenting with otolaryngological and respiratory symptoms and raised inflammatory markers.
- Steroids and immunosuppressive agents can be administered safely depending on the disease severity and gestation period.
- Aggressive treatment with a multidisciplinary approach can improve the outcome for both mother and fetus.

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