



## Case Report

# Limited Wegener's Granulomatosis Associated with Multiple Pyoderma Gangrenosum Lesions in a 30 Year Old Indian Woman

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## ABSTRACT

*Pyoderma gangrenosum is a neutrophilic dermatosis characterised by rapidly spreading and painful ulcerative lesions. Wegener's granulomatosis, a small vessel vasculitis can present with cutaneous features in approximately 10% of patients. There can be subcutaneous nodules, papulonecrotic lesions, ulcers which may resemble pyoderma gangrenosum so called 'malignant pyoderma'. We hereby report a rare case of 30 year old female with multiple pyoderma gangrenosum lesions associated with upper respiratory tract, ear, sinus involvement with positive p-ANCA subsequently diagnosed as limited Wegener's granulomatosis with pyoderma gangrenosum. Patient was treated with steroids and immunosuppressives showed significant improvement over time.*

**Keywords:** *Pyoderma Gangrenosum, Wegener's granulomatosis, pANCA.*

## INTRODUCTION

Pyoderma gangrenosum (PG) is a rare, non infectious neutrophilic dermatosis which is commonly associated with autoimmune disorders or any other systemic disease<sup>(1)</sup>. It is most commonly associated with inflammatory bowel diseases, haematological malignancies, arthritis and monoclonal gammopathies<sup>(1-3)</sup>. PG like lesions can be a presenting feature that may lead to an early diagnosis of Wegener's granulomatosis (WG) or can be found in association with it. Institution of appropriate treatment including

corticosteroids and immunosuppressives has been proven to decrease mortality in WG and PG patients<sup>(4)</sup>.

## CASE REPORT

A 30 year old female from Uttarakhand had Congenital Heart Disease- ASD secundum for which minimal invasive surgery was done through a small inframammary incision. After 7 days of discharge from hospital patient developed red raised extremely painful non itchy pus discharging ulcerated lesions over both arms, face and

trunk. The patient developed similar skin lesions at the sites of incision, intravenous cannulations and injection. The lesions started as small papules on the legs which spontaneously ruptured to form intensely painful ulcers which gradually progressed in size.

Patient was readmitted for a month in a private hospital, diagnosis of pyoderma gangrenosum was made; daily antiseptic dressings were done and low dose steroids (prednisolone 20 mg) were given but there was minor improvement. After 2 months of discharge when there was no significant improvement patient was brought to us.

Patient's history was reviewed. Along with it patient also had history of cough with expectoration and recurrent upper respiratory tract infections since last one year. She was also treated for complaints of vomiting dizziness, tinnitus and painless discharge from both ears finally diagnosed as chronic suppurative otitis media and given broad spectrum antibiotics 4 months back. Since then patient reported gradual and progressive loss of hearing from both the ears. No history of similar complaints in the past and there were no other systemic complaints.

On examination, over bilateral upper limbs (at the sites of cannulations), bilateral lower limbs, chest, inframammary (at operative site), inguinal area (at incision site) (Figure 1), buttocks; there were multiple well defined ulcerated papuloplaques of variable sizes ranging from 0.5 x 0.5 cm to 7 x7 cm approximately with irregular borders, violaceous undermined edges and necrotic slough at the floor with evidence of pus discharge (Figure 1). Lesions were tender and indurated on palpation.

On routine investigations, neutrophilia and thrombocytosis were seen along with normochromic normocytic anaemia. Erythrocyte sedimentation rate and C reactive protein were raised. Patient was immunocompetent. Liver function tests, renal function tests and serum electrolytes were within normal limits. Pus, urine and blood culture were sterile. Urine microscopic examination and 24 hour urinary proteins were normal. Chest radiograph showed blunting of right

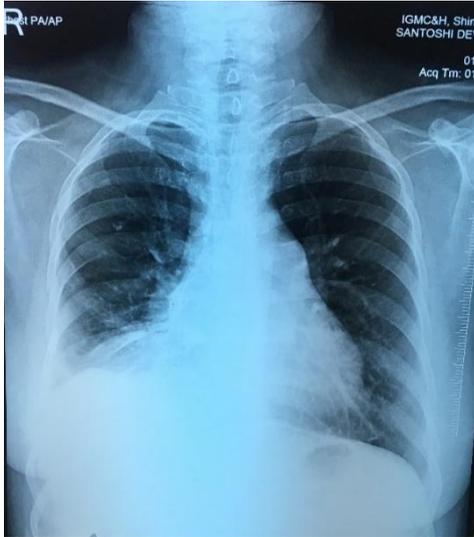
costophrenic (cp) angle (Figure 2). HRCT Thorax showed ground glass opacities in the right lower lobe.

X-ray paranasal sinuses showed haziness in right maxillary sinus (Figure 3) and on HRCT mucosal thickening of bilateral maxillary sinuses was seen. On pure tone audiometry patient had bilateral profound sensorineural hearing loss. Skin biopsy showed features of neutrophilic dermatosis; with scattered infiltration of neutrophils and lymphocytes in upper, mid and lower dermis. The subcutaneous fat showed thickened fibrous septae with a scattered inflammatory infiltrate. The biopsy was consistent with pyoderma gangrenosum. p-ANCA was positive. Patient was not willing for biopsy of lung and sinus.

A diagnosis of pyoderma gangrenosum was made with limited Wegener's granulomatosis. Patient was started on high dose steroids (tab. Methylprednisolone 40mg) for 3 weeks and broad spectrum antibiotics. Antiseptic dressings were done. Patient showed some improvement with occasional new lesions on forearm. Injection methylprednisolone 1 g intravenous pulse for 3 days was given and was started on weekly methotrexate 15 mg and steroids were tapered slowly. Patient showed continuous improvement since the institution of therapy within 6 weeks (figure 4,5,6).



**Figure 1** patient at the time of presentation showing ulcerative lesions and pathology positive at incision sites.



**Figure 2** Obliteration of right costophrenic angle with opacity right lower zone.



**Figure 3** haziness of bilateral maxillary sinuses.



**Figure 4** – pre and post treatment(at 6 weeks) pictures of lesions on right forearm



**Figure 5** – pre and post treatment(at 6 weeks) lesion on right shin



**Figure 6** – pre and post treatment(at 6 weeks) lesion on ankle

**DISCUSSION**

Pyoderma gangrenosum is a extremely painful ulcerative neutrophilic dermatosis who fulminant course. It is associated with systemic diseases in 50-70 % cases more commonly with inflammatory bowel disease and rheumatoid arthritis but has rarely associated with wegener’s granulomaosis (WG). WG now known as granulomatosis with polyangitis (GPA) is characterised by systemic granulomatous small vessel vasculitis, necrotising granulomas of upper and lower respiratory tract (80%) & glomerulonephritis. Cutaneous features are present in about 40% of the patients. Approximately 73% patients have nasal, tracheal, sinus and ear involvement. Renal involvement is initially seen in only 18 % patients but eventually 77% develop glomerulonephritis <sup>(5)</sup>. Our patient had no renal involvement at the time of presentation.

WG affects males and females equally, incidence is 5–10 per million per year, more common in Caucasians<sup>[6]</sup>. The average age at onset is 40 years<sup>[7]</sup>. Pulmonary involvement can be in the form of infiltrative lesions or haemorrhage, recurrent pneumonias which on healing gives ground glass appearance on chest X-ray. Cavities may give rise to haemoptysis; pleural effusions may occur<sup>(8)</sup>. The early changes in classical WG involve the upper respiratory tract, with destruction and crusting of the nose, otitis media and involvement of paranasal sinuses.

A positive C-ANCA has 66% sensitivity (91% in active disease) and 98% specificity for classical WG<sup>[9]</sup> but p-ANCA can be positive in patients with WG who are negative for c-ANCA/anti-PR3. 10–20% of patients with WG have negative tests for ANCA (or may develop a positive test later in the disease). p-ANCA is found in 5% of patients of WG. In localised variant of Wegener's c-ANCA positivity is found only in 39% of patients as compared to the generalised WG it is seen in 86%. Also patients with p-ANCA positivity have less organ involvements such as respiratory tract and renal<sup>(10)</sup>.

Our patient had a concordance with the limited WG by testing negative for c-ANCA but positively for p-ANCA, respiratory involvement, otitis media, hearing impairment and, maxillary sinus involvement with sparing of renal system. The characteristic morphology of lesions with rapidly growing painful ulcers with violaceous margins, with positive pathology and the histopathology report concluded the diagnosis of pyoderma gangrenosum. So final diagnosis of pyoderma gangrenosum with associated limited Wegener's granulomatosis was made. Also there was good response to the high doses of corticosteroids and immunosuppressives.

Although we could not document it on lung or sinus biopsy because of patient's disapproval.

Standard treatment for WG includes extended systemic glucocorticoid and concomitant cytotoxic drug therapy. Treatment options are cyclophosphamide, methotrexate with folic acid

and leucovorin, trimethoprim-sulfamethoxazole, and cyclosporine. Generalized WG may smolder for years before clinical signs appear, and if untreated has a greater than 90% fatality rate. Therefore early detection by strong suspicion and immediate treatment is necessary to improve the survival of such patients<sup>(11,12)</sup>.

The association of WG with PG is rare and encountered in very few patients till date<sup>(13)</sup>. Our case presented as PG subsequently found to be associated with localised variant of WG. Patient showed marked improvement on combination of oral steroids and weekly methotrexate therapy and still on follow up.

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