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Painless Pyoderma gangrenosum over hand in association with Ca-lung:An unusual association at unusual site with unusual presentation.

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Abstract: *Pyoderma gangrenosum(PG) is a chronic destructive ulcerating skin lesion of unknown etiology. It is associated with some systemic medical illnesses in 50% of cases like inflammatory bowel disease, systemicarthritis, hematological disease and malignancies. Characteristic lesion begins as pustule or vesiculopustule and progresses to an ulcer or deep erosion with violaceous overhanging or undermined borders. It is often a diagnosis of exclusion of other causes of cutaneous ulceration as laboratory and histopathological findings are variable and non-specific. Here we describe a case of PG over hand in an adult male who was suffering from metastatic squamous cell carcinoma lung and subsequently offered palliative chemotherapy as well as systemic and topical therapy for pyoderma gangrenosum.*

Keywords: *Pyoderma gangrenosum, ulcerative, squamous cell carcinoma lung.*

I. Introduction:

Brunsting et al. gave the initial clinical description of Pyoderma gangrenosum (PG) in 1930.[1] PG is a rare, noninfectious, ulcerative, neutrophilic dermatosis commonly associated with underlying systemic diseases. There are four main clinical types:i) Ulcerative (Typical or Classic) ii) Pustulariii) Bullousand iv) Vegetative. In addition peristomal PG is also known.[2]The most common associated diseases are inflammatory bowel disease, arthritis, hematological malignancies and monoclonal gammopathies.Solid organ malignancies of colon, pancreas, breast were reported with extreme rarity of Ca lung.[3] The ulcerative type progresses rapidly(usually> 1 cm/ day) to a painful, necrolytic ulcer with an irregular undermined violaceous border. Lesion may be solitary or multiple coalescing together. If untreated, it may progress to expose muscles, vessels, nerves, fascia and even bones. It heals with cribriform scarring.[4]A characteristic feature of PG is pathergy reaction (only 25% of patients have this feature).Leg is commonest site of involvement (70%), mucosa & genitalia involvement may get involved but dorsum of the hand.[5]Histopathologyshows central necrosis and ulceration of epidermis and dermis surrounded by an intense acute inflammatory cell infiltrate, with a peripheral mixed to chronic inflammatory cell infiltrate. In ulcerative variant, there is a massive dermo-epidermal neutrophilic infiltrate with abscess formation.[2,4,5]Here our case is a 55 year old man in whom PG appeared on dorsum of right hand. Subsequently he was found to be suffering from squamous cell carcinoma of lung which is an unusual association.

II. Case Presentation:

A 58yr oldnormotensive,euglycemic,afebrile male presented with shortness of breath on exertion along with left sided chest pain for last 2 months. He was chronic smoker and alcohol addict. At the same time patient developed a necrotic rapidly spreadingpainless,purulentulcer over the dorsum of right hand including fingers and visited our OPD facility of Dermatology department at CSTM,Calcutta(Figure1).No signs of neurological deficit or history of surgery,trauma,drug intake(warfarin etc),bowel irregularity,joint painor other significant family history could be elicited.

He was average built but pale with no sign of malnutrion.His right axillary lymph nodes and bilateral epitrochlearlymph nodes were palpable.

This tachypneic male had diminished breath sound on left side of chest with audible wheeze bilaterally. Blood investigations like serum urea, creatinine, liver function test, serum electrolytes were within normal limit. Complete haemogram showed leucocytosis with neutrophilia. Routine and microscopic examination of urine and stool showed no abnormality. CRP was within normal range. ESR was raised to 80 mm /hr.HIV-1 and 2 serology were nonreactive. FDP and D-Dimer test were negative.

Punch biopsy taken from the edge of the ulcer showed thick neutrophilic crust in the keratin layer of epidermis. The ulcerated area is covered by inflammatory exudates. The dermis shows an area of necrosis surrounded by inflammatory cells with predominance of polymorphonuclear neutrophils. The epidermis

adjacent to ulcer shows hyperkeratosis, acanthosis and downward elongation of rete ridges. The dermis is also oedematous and shows perivascular inflammatory cell infiltrate(Fig 2).

Chest x-ray was suggestive of left lung collapse with shifting of trachea towards left side with associated left sided pleural effusion(Fig 3).

CECT chest showed left sided pleural effusion with collapsed left lung and left parahillar soft tissue lesion(Fig 4).CT guided FNAC from left lung SOL showed a few discrete and tiny clusters of malignant epithelial cells, degenerated mesothelial cells, alveolar macrophages and inflammatory cells. Some of the malignant cells showed squamous differentiation.

Papanicolaou stain of pleural fluid showed discrete and clusters of large cells with nucleomegaly and hyperchromasia suggestive of malignant epithelial cells in a haemorrhagic background.

III. Discussion:

PG is an inflammatory neutrophilic dermatoses characterized by painful cutaneous ulcerations with mucopurulent exudates and undermined bluish borders with surrounding erythema.

The peak of incidence occurs between the ages of 20 and 50 years with women being more often affected than men. PG in elderly people has occasionally been reported.[6]PG is commonly located on the lower limbs (especially on pretibial area), but may occur anywhere like face, neck, breast, hand, trunk, scrotum or penis.[5] The lesion often begins as a pustulewith rapid growth at the site of minor trauma and subsequently a large ulcer (as a result of pathergy phenomenon).[7]There is associated pain which is often severe and out of proportion to the size of the lesion.Here in our case the lesion occurred over right hand.[8]It began as a pustule but spread rapidly within one to two days into a necrotizing ulcer to involve the whole dorsum of the right hand including fingers. Striking feature was that the ulcer was painless throughout the course.

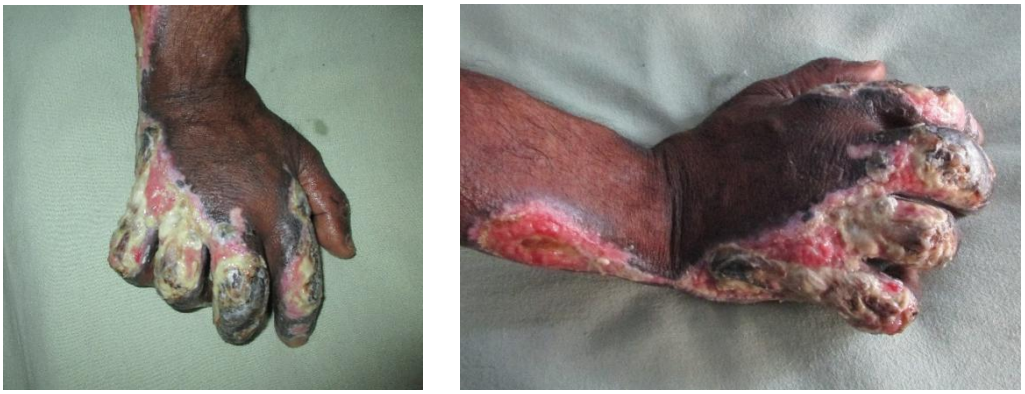
Ulcerative variant of PG may be due to pathergic trigger like surgery or associated with IBD, arthritis, monoclonalgammopathy or internal haematological malignancies . Diagnosis relies on clinical signs first and is supported by histopathology.No laboratory parameter is diagnostic. Histopathology is non-specific.Diagnosis is often made by exclusion of other possible disorders.Massive neutrophilic infiltration in the absence of vasculitis and granuloma formation is typical of PG.

PG is included in the class of neutrophilic dermatoses.[7]Approximately 50% of patients with PG have an underlying systemic disease.Depending upon the associated conditions PG is classified as follows:[1]

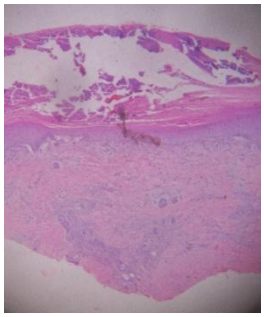
- Parainflammatory (associated with IBD, collagen vascular diseases, arthritis, etc)
- Paraneoplastic (associated with malignancy)
- Hemotologic (leukemias, polycythemia)
- Drug induced
- Idiopathic

Many patients with PG used to be treated by antibiotics before the diagnosis, but they usually have no role. The mainstay of therapy is systemic corticosteroids, but in our case we did not give it due to underlying malignancy and LRTI. We have treated the patient with antimicrobials (for LRTI), dapsone 100mg daily and clofazimine 100 mg daily along with local dressings and absorbent bandages (changed daily) for the ulcer. The lesion responded very well to these drugs due to their anti –inflammatory effects and effect on alteration of neutrophil function.[9] The patient also receivedPaclitaxel and Carboplatin based palliative chemotherapy for squamous cell carcinoma of lung.Though there was extensive necrosis of the skin with vital tissues such as tendons and ligaments were exposed at the ulcer bed, we did not go for surgical interventions because of the risk of pathergy. It has been reported that pathergy is more common in PG associated with systemic disease. Subsequently skin grafting may be necessary for this case.

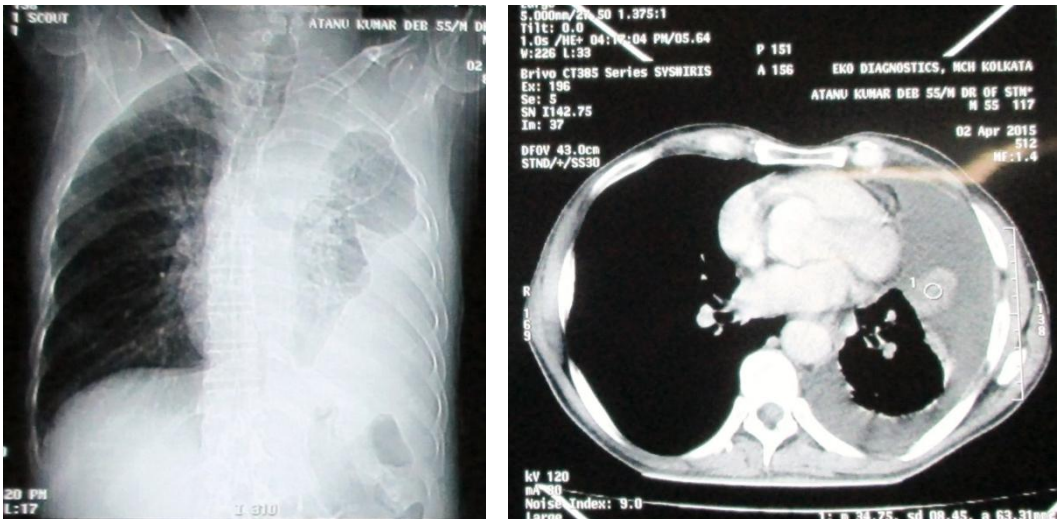
To the best of my knowledge ulcerative variant of PG occurring at dorsum of the hand in 58 yrs old male presented as a painless lesion in association with Ca-lung is a medley of uniqueness and therefore reported.



Ulcerative Pyoderma gangrenosum over dorsum of the hand with bluish undermined edge &overhanging violaceous border(Fig 1)



Histopathology (10 X view) shows hyperkeratosis with predominant neutrophilic infiltrate of dermis (Fig 2)



CXR: Lt sided opacity with collapseCECT:Prahilar SOL with collapse & effusion on lt lung (Fig 3)

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