

PYODERMA GANGRENOSUM - A RARE CASE REPORT

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ABSTRACT

Pyoderma gangrenosum presents as a rapidly enlarging, very painful ulcer. It is one of a group of autoinflammatory disorders known as neutrophilic dermatoses. Pyoderma gangrenosum (PG) is a rare ulcerative condition that is diagnostically and therapeutically challenging, as debridement leads to ulcer deterioration (pathergy phenomenon).

KEYWORDS: Autoinflammatory disorders, neutrophilic, Pyoderma gangrenosum.

INTRODUCTION

Pyoderma gangrenosum is a condition that causes tissue to become necrotic, causing deep ulcers that usually occur on the legs. When they occur, they can lead to chronic wounds. Ulcers usually initially look like small bug bites or papules, and they progress to larger ulcers. Though the wounds rarely lead to death, they can cause pain and scarring.

CASE REPORT

35 year old male came with H/O fall from two-wheeler 15 days back, H/O topical application for the wound(+), after which he started to develop plaque over it. No H/O pain, burning sensation. N/K/C/O – DM/HT/TB/BA.

L/E: Verrucous plaque noted over right forearm. Oral mucosa normal. Scalp normal. Nail normal.

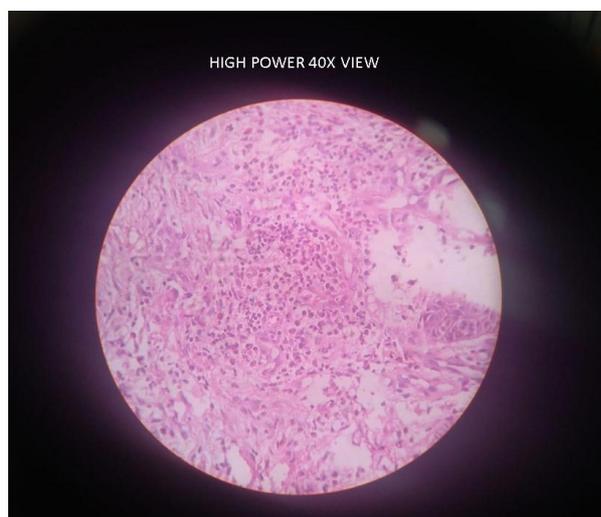
Pathology

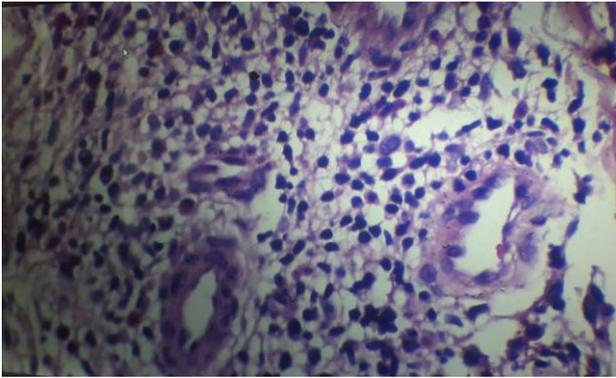
- **Specimen:** 4mm punch biopsy taken from forearm and sent for HPE.
- **Gross:** Received single skin covered soft tissue bit measuring 0.4cm, AE.

MICROSCOPIC FINDINGS

Lowpower(10X) Section shows hyperkeratosis of squamous epithelium continuing numerous pockets of chronic inflammatory cells.

Highpower(40X) Papillary dermis shows diffusely scattered lymphocytes, plasma cells, few eosinophils and foamy macrophages around adnexal ducts and blood vessels.





Mixed lymphocytic and neutrophilic infiltration around the blood vessels.

DISCUSSION

Pyoderma gangrenosum presents as a rapidly enlarging, very painful ulcer. It is one of a group of autoinflammatory disorders known as **neutrophilic dermatoses**.

Causes

- Pyoderma gangrenosum is an autoinflammatory disease (excessive response to an internal antigen) due to some form of neutrophil dysfunction. T lymphocytes and cytokines are involved.
- Drugs are occasionally implicated as triggers of pyoderma gangrenosum, especially cocaine, isotretinoin, propylthiouracil, interferon – alpha.
- Also due to trauma (Koebnerization).

Sites

Lower extremities and trunk.

AGE GROUP: 30 to 50

Variants

- Superficial granulomatous variant – Florid pseudoepitheliomatous hyperplasia may be seen along with granulomatous inflammation with plasma cells and eosinophils.

- Vesiculopustular variant – associated with IBD. Necrotizing pustular follicular reaction seen.

Clinical features

Begins as a folliculocentric pustules/nodules, lesions ulcerate and have sharply circumscribed violaceous, raised edges in which necrotic pustules may be seen.

Associated conditions

IBD, Acute lymphoid and myeloid leukemias and myeloma, rheumatoid arthritis and SLE, Monoclonal gammopathy (usually IgA), hepatopathies, chronic active hepatitis, primary biliary cirrhosis.

Diagnosis

1. Biopsy

- Shows central necrosis, suppurative inflammation, ulceration, lymphocytic vascular reaction with lymphocyte infiltrate and epithelial undermining.
 - *Sweet's-like vascular reaction* (mixed lymphocytic and neutrophilic vascular reaction).
2. The pathergy test is usually positive (a skin prick test causing a papule, pustule or ulcer).
 3. Some patients may have a positive ANCA (antineutrophil cytoplasmic antibody), elevated levels of IL-8.
 4. Direct Immunofluorescence test shows deposits of IgM, C3.
 5. PCR shows clonal expansions of T cells in peripheral blood and skin.
- DIFFERENTIAL DIAGNOSIS:**
- Sweet's syndrome (rarely folliculocentric and absence of dermolysis)
 - Mixed cryoglobulinemia, behcet's disease, rheumatoid vasculitis, pustular drug reaction. (It also shows necrotizing pustular follicular reactions).
 - Bowel arthritis dermatosis syndrome, pustular vasculitis (shows Sweet's like vascular reaction).

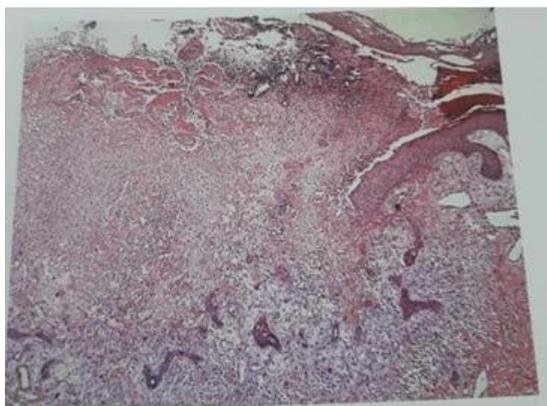


FIGURE 16-3. Pyoderma gangrenosum. The center of the lesion shows a neutrophilic infiltrate with leukocytoclasia and dermolysis. This biopsy is from a patient with Crohn's disease, as evidenced by the presence of multinucleated histiocytes within the infiltrate.

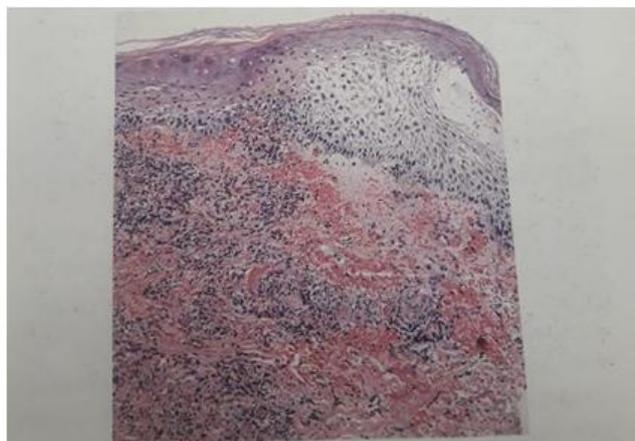


FIGURE 16-4. Pyoderma gangrenosum. The undermined epidermis often shows spongiosis or pustulation.

CONCLUSION

Although PG is rare, clinicians should suspect it in the presence of purplish wound edges with a necrotic center. A close coordination between dermatologist and pathologist is crucial to the timely and accurate diagnosis.

REFERENCES

1. Lever's Histopathology of the skin, 10th edition.