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## Pyoderma Gangrenosum Vegetans

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### Abstract

Pyoderma Vegetans (PV) is a chronic inflammatory disease that usually presents as verrucose plaques. It belongs to the group of neutrophilic dermatoses and it is diagnosed right after other inflammatory and infectious diseases are excluded. It shows no association to systemic disorders and has great response to oral corticosteroids. We hereby report a patient presenting PV of lower limbs.

**Keywords:** Pyoderma gangrenosum; Pyoderma vegetans; Neutrophilic dermatoses; Ulcerated plaque

#### Introduction

Within the pyoderma gangrenosum (PG), the variety vegetans is a rare form. It was first described by Hallopeau in 1898 [1-3]. Its etiology remains unknown. It starts as a superficial ulcer or ulcerated plaque of vegetative aspect, typically in the trunk [1]. It has a slow progression and resolves with hypo and hyperpigmented atrophic scars. It is rarely associated with systemic diseases and responds to systemic corticosteroids.

#### Clinical Case

A 37 year old man with a history of diabetes mellitus and dyslipidemia presented with painful lesions on both lower limbs of 2 months of duration. Physical examination revealed multiple verrucose plaques of 5 cm diameter with central ulceration and well-defined purple borders (Figures 1 and 2).

They were painful and exudative. Inguinal lymphadenopathy was found. No evidence of mucosal or appendages involvement was noted. He remained afebrile and in good general condition. The rest of the physical examination was unremarkable.

Complete blood count, kidney, liver function tests and serum electrolyte levels were within normal limits. Serum glucose level and lipids were elevated.

Wound culture taken from lesion to detect fungi, bacteria, leishmania and common germs were negative. Polymerase chain reaction (PCR)



Figure 1: Verrucous plaque with central ulceration, elevated borders and satellites lesions.



Figure 2: The same plaque one month later.

for atypical mycobacterias was negative. The histopathological study of the skin lesion revealed hyperkeratosis and acanthosis. The dermis showed inflammatory infiltrated comprising plasma cells, neutrophils and eosinophils mostly and areas of fibrinoid necrosis (Figures 3 and 4).

Periodic Acid-Schiff (PAS) and Ziehl Neelsen's stain were negative. According to the clinical and histopathological findings, after ruling out infectious origin of the cutaneous lesions the patient was diagnosed as a vegetans variant of PG. Treatment was initiated with meprednisone at doses of 1 mg/kg/day with remission of the lesions after 4 months of therapy. Currently, he is undergoing systemic corticosteroids tapering without signs of recurrence of his disease.

The prognosis of PV is very good, and heals with cribriform scar appearance. In our patient atrophic scar was observed (Figure 5).

## Discussion

Pyoderma gangrenosum variety vegetans is a rare entity, which has

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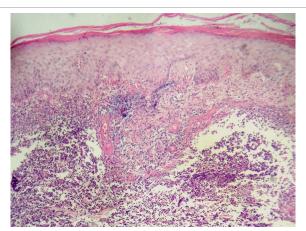


Figure 3: Epidermis: hyperkeratosis and acanthosis. Dermis: inflammatory infiltrated and areas of fibrinoid necrosis.

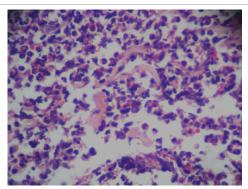


Figure 4: Dermic inflammatory infiltrated comprising plasma cells, neutrophils and eosinophils.



Figure 5: Atrophic scar with cribriform aspect.

been described mostly in middle-aged adults. Its prevalence is unknown but it is estimated that there are around three cases per million people/year. The incidence is slightly superior in women [2,4,5].

Pyoderma vegetans etiology is unknown, but it has been postulated an immune mechanism due to its favorable response to immunosuppressive therapy. Defects in chemotaxis of neutrophils and overexpression of IL-8, a neutrophil chemotactic agent, could be some of the mechanisms involved in the development of this entity [1].

Clinically, PV starts as a nodule that evolves into a superficial ulcer or a verrucous plaque. It has a slow progression and the most frequent location is the trunk. It has also been observed in face, scrotum and limbs [1]. Pathergy phenomenon is not seen as in the classic PG.

When vegetans variety of PG is clinically suspected, complete blood count, erythrocyte sedimentation rate, kidney, liver and bone profile, investigation of autoantibodies, thyroid function studies and chest radiography must be requested. Swabbing of injuries and tissue samples to be sent for culture are needed to exclude bacterial, fungal and mycobacterial infections [6].

Histopathology is nonspecific. Papillomatous hyperplasia of the epidermis, dermal granulomas and focal neutrophilic abscesses are the main histological features [6]. Anyway, the absence of granulomas does not invalidate the diagnosis. Therefore, clinical correlation is mandatory. The diagnosis is also supported by the good response to the therapy with steroids.

Currently there is not a guide to treatment, but systemic corticosteroids are usually the first choice [1]. Doses of 1 mg/kg/day orally were reported to be successful to control the inflammation and relieve pain. This dose should be continued until the lesions show evidence of healing. At this point, steroids dose can be gradually decreased. A steroid-sparing agent, such as minocycline, dapsone, rifampicin, vancomycin or clofazimine could be added [1].

The diagnosis is oriented with the clinical and histopathology, and confirmed only once other causes of verrucous plaques are excluded [2]. Finally, the good response to treatment confirms the diagnosis.

#### Conclusion

In conclusion, the variety vegetans of PG is a rare condition of unknown etiology. Its evolution is slow and heals with cribriform pattern. Its diagnosis is a challenge and should exclude other inflammatory and infectious causes of verrucous plate. Unlike other varieties, systemic corticosteroid therapy is highly effective. It has minimal association with systemic diseases and complete healing is the rule.

## **Ethical Agreement**

Written consent of the patient was requested to publish the photos.

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