PLECT or PPLECT? Granulomatous pyoderma gangrenosum in the differential diagnosis of the verrucous syndrome*

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Abstract: Pyoderma gangrenosum (PG) is an uncommon neutrophilic dermatosis, with variable clinical features, nonspecific histopathology and multifactorial pathogenesis, posing a challenging diagnosis for the dermatologist. Pyoderma gangrenosum is a diagnosis of exclusion and should be included in the differential diagnoses of the verrucous syndrome. We report a granulomatous variant affecting the face.

Keywords: Facial dermatoses; Granuloma; Pyoderma gangrenosum

Pyoderma gangrenosum (PG) is an uncommon entity with a diverse clinical spectrum and nonspecific histopathology. Awareness of its various manifestations, as well as its differential diagnoses, is essential for early therapy to reduce morbidity. On initial approach, PG should also be remembered as a clinical hypothesis of the verrucous syndrome.

We report a 72-year-old male patient who sought care for painful facial lesions that had evolved over six months. He reported that the first lesion appeared in the right temporal region and had rapidly and progressively worsened. Subsequently, a smaller lesion, though similar to the first one, appeared in the left temporal region. The patient was a smoker (more than 30 pack-years), previously healthy, and denied the use of medications.

The lesions consisted of verrucous, ulcerated, crusted plaques with erythematoviolaceous and infiltrated borders, initially in the right temporomalar region, with later involvement of the contralateral area (Figure 1). Two KOH preparations yielded blastoconidia and yeast-form hyphae, and oral itraconazole therapy instituted four months before showed no improvement.

On histopathology, a chronic granulomatous inflammatory process (Figure 2), with negative tests for fungi (PAS and Grocott) and mycobacteria (Ziehl-Neelsen), was observed . The samples sent for detection and culturing of fungi, bacteria, and mycobacteria were negative. Direct inspection for *Leishmania sp.* (Giemsa) on the lesion was negative; the Mantoux (PPD) test was nonreactive.

After excluding infectious and neoplastic causes, the hypothesis of granulomatous PG variant was discussed and established as the principal possibility. Prednisone (1mg/kg/day), in combination with trimethoprim/sulfamethoxazole (TMP/SMX; 800mg + 160mg), was initiated, with regression of the lesions in the first month of treatment. Prednisone was gradually tapered and dapsone (50mg/day) substituted for TMP/SMX. Currently, the patient remains asymptomatic on dapsone (100mg/day) alone . The investigation for underlying diseases possibly associated with PG showed no abnormalities.

PG, first described by Brocq[1] and named by Brunsting *et al*.[2] in 1930, is a rare neutrophilic dermatosis of unknown etiology classically characterized by a crusted and painful ulcer with violaceous and undermined borders.[3] The clinical spectrum is varied, and the diagnosis is based on clinicopathological correlation and on the exclusion of other causes.

PG can occur at any age, with predominance in adult women between the second and fifth decades of life. Its pathogenesis is multifactorial and involves neutrophilic disfunction, inflammatory

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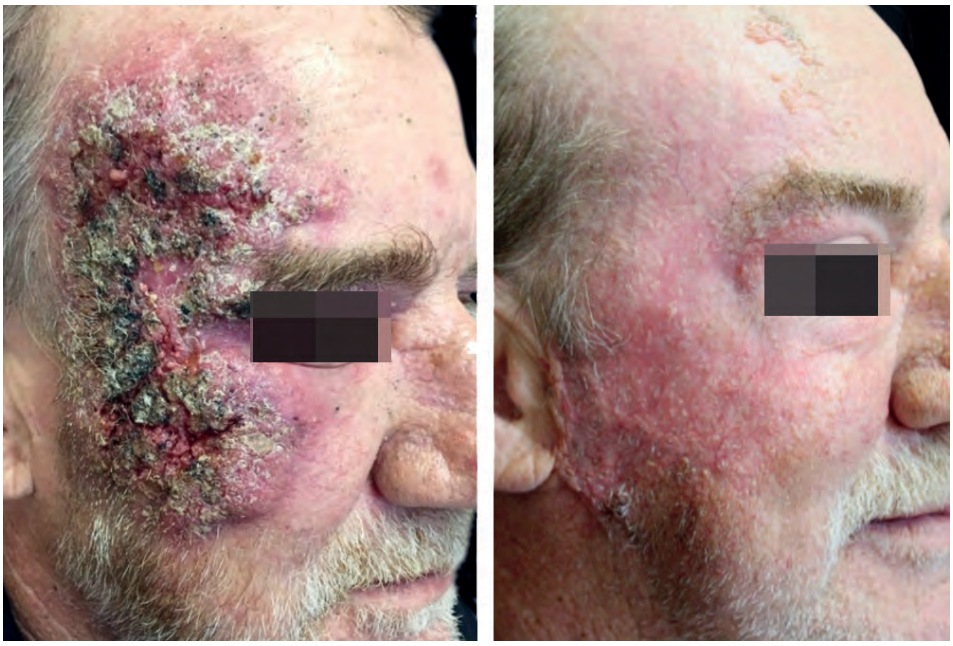

Figure 1: Before and after treatment. Left: extensive verrucous plaque in the right temporomalar region, with erythematous and infiltrated borders and scaly, crusted central ulcerations. Right: appearance after the established therapy

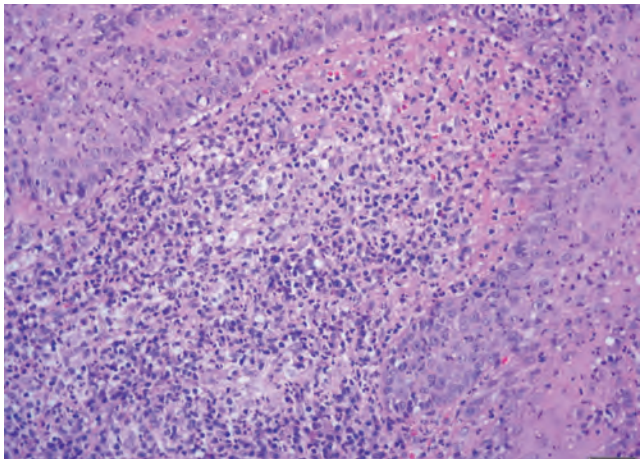

Figure 2: Histopathology: chronic granulomatous inflammatory process (Hematoxylin & eosin, x200)

mediators and genetic predisposition. It has been described in association with underlying systemic diseases in 50% of cases, most commonly inflammatory bowel disease, rheumatological or hematological conditions (monoclonal gammopathy, myelodysplasia, acute myeloid leukemia).[4]

Although the classic morphology comprises nodules or papules that evolve into rapidly progressive, painful ulcers with violaceous borders and a purulent base, there are many clinical variants such as ulcerative, bullous, pustular and granulomatous subtypes.[5] Granulomatous PG can exhibit verrucous characteristics, as in the present case.

The histopathology is nonspecific and depends on the subtype of the lesion and on the duration of evolution, but it contributes to the exclusion of other diseases. There is neutrophilic infiltration in recent lesions, and necrosis associated with fibrosis in the chronic lesions, as well as a granulomatous process; however, these findings are not pathognomonic.[5]

The diagnosis is often challenging and delayed, requiring clinical and pathological correlation; hence the importance of familiarity with the differential diagnoses and morphological aspects. It should be noted that PG may be included as a differential diagnosis of the verrucous syndrome, classically recognized by the mnemonic term "PLECT" (paracoccidioidomycosis, leishmaniasis, sporotrichosis, chromomycosis, tuberculosis). Histoplasmosis should also be considered.

Negative screening investigation for systemic disease prompted us to consider the idiopathic nature of PG in our patient. Facial involvement was striking and the excellent response to both steroid and sulfonamide should be highlighted. Given the distinct verrucous aspect that granulomatous PG can present (which has long been described in the literature), could a second "P" be added to the term PLECT (pyoderma-PLECT, or PPLECT)? The reminder of this clinical form of PG by means of a well-known mnemonic tool probably results in greater diagnostic accuracy in difficult cases of the verrucous syndrome.

The most effective treatment for PG is systemic corticotherapy, which can be administered through pulse therapy for more severe cases. Other medications with antineutrophilic potential, such as dapsone and other sulfonamides, and clofazimine can also be used, as in the present case. Cyclosporin, azathioprine, and infliximab have also been employed as alternative drugs in refractory cases.[6] ❑

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