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Case Report

Vulvar Pyoderma Gangrenosum and Inflammatory Disease

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A B S T R A C T

Pyoderma Gangrenosum is an ulcerative skin lesion commonly associated with systemic diseases or malignancies. Genital pyoderma gangrenosum is very rare and may be misdiagnosed. We report a case of vulvar pyoderma gangrenosum in a 25-year-old patient with a challenging differential diagnosis to discuss between Crohn's disease and Behçet's disease.

Keywords: Pyoderma Gangrenosum, IBD, Behçet's Disease, Genital ulcerative skin lesions

Abbreviations : IBD: Inflammatory Bowel Disease ; CD: Crohn's Disease; BD: Behçet's Disease; PG: Pyoderma Gangrenosum; EDM: Extra-Digestive Manifestations.

Introduction

Pyoderma gangrenosum is a deep ulcerated skin lesion affecting the lower limbs in 70% of cases. In over 80% of cases, it is associated with a systemic disease. Genital involvement is extremely rare, and it represents a real problem of differential diagnosis, particularly when it is associated with digestive and general lesions, common to various conditions. Thereby, leading to discussing the association between PG and CD and/or BD.

Case Report

LZ is a 25-year-old woman, with no history of unprotected sexual intercourse and no family history of IBD, tuberculosis or neoplasia. However, she reports recurrent mouth ulcers since 5 years. Four months ago, she presented with a left vulvar ulceration considered as a Bartholinitis by her gynaecologist. The disease rapidly worsened after antibiotic treatment becoming painful and preventing her from walking. The patient's general condition was preserved. Abdominal, joints and skin examinations were unremarkable. At the perineal site, there is a terebrating lesion on the left vulvar lip, with no anal lesion (**Figure 1**).

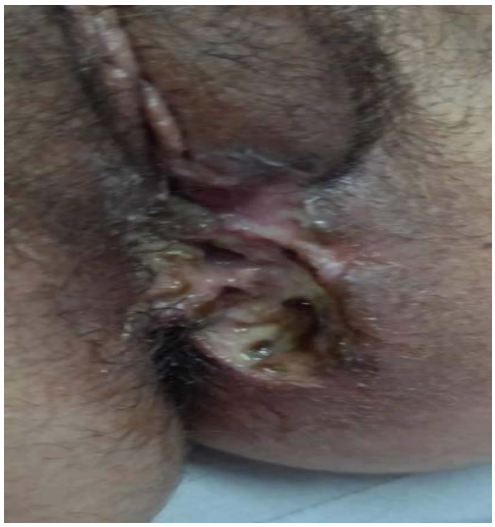

Figure 1. At the admission, the vulvar lesion of the major left lip was terebrating, ulcerated, irregular, covered by a thick deposition of purulent aspect, with raised nodular and purplish margins, extending nearly to the anal orifice.

The biological tests revealed anemia, hyperleukocytosis and an elevated CRP to 135 mg/l. The histolopathology described a conjunctive-adipose tissue heavily infiltrated with inflammatory cells and numerous neutrophils, with two arterial sections showing thrombosis and multiple abscessed structures. The perineal MRI showed no anoperineal or rectovaginal fistula, but a large ulceration of the soft tissues of the left external genitalia. The colonoscopy showed aphthoid ileocecal erosions with an histological infiltrate of mononuclear inflammatory cells and neutrophils. The entero-MRI showed circumferential thickening of the last ileal loop, moderately reducing the lumen and extending to the ileocecal valve. Serological tests for sexually transmitted infections were negative. The HLA B51 typing was positive and the pathergic test was negative. The ophthalmological and the neurological examinations were unremarkable. After the dermatologist consultation, the diagnosis of left vulvar PG, associated with CD or BD-like overlap was proposed. The patient was treated with antibiotics and steroids, leading to rapid resolution of all symptoms (**Figure 2 and Figure 3**).

Figure 2. At day 15 after antibiotics and steroids treatment, the vulvar lesion presented with healing granulation tissue in the excavated lesion which was clean.

Figure 3. At 4 weeks after traitement, a sequelae of the vulvar lesion presented with a retracted scar.

Discussion

PG is a neutrophilic, destructive, autoimmune-mediated dermatosis of unknown etiology. In more than 80% of cases, this entity is associated with an inflammatory or a neoplastic systemic pathology, in half of the cases, it is an IBD. In our young patient, the typical clinical aspect rapidly evolving with the histological data (Daniel Su criteria) may lead to the diagnosis of vulvar PG with a high likelihood. The extremely rare genital localization, reported in 23 cases (1965-2013) of the literature, asks about etiological differential diagnosis between infections, the main cause of diagnostic error, as shown in our patient who was initially treated for Bartholinitis; others possibilities include neoplasms or PG integrating a CD or BD.

CD can be associated in 40% of cases with arthritis, uveitis, skin lesion, mouth ulcers, which are also frequent during BD representing the major differential diagnosis. The diagnosis of CD is likely in our patient despite the lack of histopathologic granuloma. This diagnosis was proposed on the young age of the patient, the ileocecal localisation of the illness confirmed by the ileocolonoscopy and the entero-MRI and the association of Extra-Digestive Manifestation as mouth ulcers associated with PG, revealing an IBD in 1/4 of cases. The severity and the activity of the two clinical forms are not parallel; as reported in our patient, a severe extra-digestive form may be accompanied by a minimal digestive form. The PG during the CD is very rare, sitting in most of cases in the legs or near intestinal stomas. Even more, the vulvar localization of PG is exceptionally reported[1]. The favorable response to the steroids is also compatible with the diagnosis of CD with cutaneous EDM.

BD borrows a wide clinical spectrum involving the digestive tract in 40% of cases. In the absence of pathognomonic markers, the diagnosis of BD is based on various clinical criteria : recurrent oral aphthous, ileocoecal aphthoid lesions present in more than 14% of cases, an eosinophilic infiltrate with vascular thrombus at the histology, as well as a positive HLA B51 typing (odd's ratio MB: 1.1–22.2), although present in 15% of patients with IBD and in 20% of healthy subjects living along the «Route of Silk» extending from Asia to the Mediterranean Area, from where our patient comes ; despite a negative pathergic test and the absence of ocular, neurological, vascular involvement. In addition, an incomplete probability score (Criteria of the International Behçet Study Group) supports this diagnosis in our patient. Although it is very rare, the association of these two neutrophilic dermatoses; vulvar PG and BD, was reported in the literature[2].

The differential diagnosis between CD and BD is a real challenge[3], although the association between these two entities can exist suggesting different aspects of the same spectrum of Chronic Inflammatory Systemic Diseases. The clinical, radiological, endoscopic, histological data may be similar with superimposable EDM. This symptomatic and histological proximity suggests a close pathogenetic link between the two entities. The overlap or even the CD-BD association with PG[4] as the revealing initial lesion was the diagnosis retained in our patient.

Conclusion

Vulvar PG is very rare. It requires to rule out a sexual infection and malignant neoplasia. Its diagnosis is a real challenge, especially when it reveals a Systemic Inflammatory Disease; the most common being the CD. A difficult differential diagnosis, the BD, can also realize an overlap with superimposable clinical, endomorphological, histopathological data.

Conflicts of Interest: None

References

1. Bommireddipally J, Broussard K, Osman H, Uhlhorn A, Loganantharaj N. Gastrointestinal: A rare case of concurrent vulvar Crohn's disease and pyoderma gangrenosum. J Gastroenterol Hepatol. 2024;39:417-419.

2. Ozuguz P, Kacar SD, Manav V, Karaca S, Aktepe F, Ulu S. Genital Ulcerative Pyoderma Gangrenosum in Behçet's Disease: A Case Report and Review of the Literature. Indian J Dermatol. 2015;60:105.

3. Evereklioglu C. Diagnostic dilemma between intestinal Behçet disease and inflammatory bowel disease with pyoderma gangrenosum. World J Gastroenterol. 2006;12:5748-5751.

4. Akay N, Boyvat A, Heper AO, et al. Behçet's disease-like presentation of bullous pyoderma gangrenosum associated with Crohn's disease. Clin Exp Dermatol. 2006;31:384-386.