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Pyoderma Gangrenosum and Behcet's Disease Overlap –A Case Report

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Abstract

Pyoderma gangrenosum is a rare, chronic, sterile,pustular and progressive ulcerative disease with 4 clinical and histological variants named as ulcerative, pustular, bullous and vegetative. Behcet's disease is a complex of multisystem disease characterized by orogenital ulceration and eye disease as a classic triad with involvement of other systems . BD and PG , both are neutrophilic dermatoses with oral and genital ulcerations having different frequency. The clinical features of mucosal ulcers are different to some degree between the two diseases, but there is a histopathological distinction between PG and BD . Association of BD and PG are rare but do occur . We report here a case of BD who subsequently developed genital ulcerative PG.

Keywords: Pyoderma gangrenosum; Behcet's disease; Neutrophilic dermatosis; Orogenital ulcers.

Introduction

Pyoderma gangrenosum (PG) is a rare, chronic, sterile,pustular and progressive ulcerative disease with unknown cause. Classical PG presents most commonly as an extremely painful erythematous lesion which rapidly progresses to a blistered or necrotic ulcer. There is often a ragged undermined edge with a violaceous/erythematous border and lower legs are most frequently affected.[1]

Behcet's disease (BD) is a complex of multisystem disease characterized by orogenital ulceration and eye disease as a classic triad with involvement of cardiovascular, gastrointestinal, musculoskeletal and central nervous systems also at times.[2]

There are few reported cases involving the diagnosis of BD and PG in the literature.[3] We report here a case of BD who subsequently developed genital ulcerative PG.

Case Report

A 30 year old, unmarried, female residing at Madhya pradesh, presented to skin OPD with complain of lesions over genitals since 3 months associated with fever, pain, itching, burning sensation and discharge which gradually turned into raw areas. Patient has taken medications from many private practitioners with no improvement . She complained of constipation, throat pain, recurrent oral ulcers and history of weight loss since past 6 months. History of redness, itching and watering from bilateral eyes since 2 years was present.

No history of seizure, unconsciousness, breathlessness, chest pain, abdominal pain, vomiting, diarrhoea, seasonal aggravation, any known food or drug allergy. No history of any sexual exposure was present.

Past history of similar lesions 3 years ago which

were associated with fever, pain, burning sensation and itching, She had Pulmonary tuberculosis 7 years ago and had taken AKT for 6 months. No history of similar skin lesions in family. The menstrual cycle was regular at every 28-30 days interval with 5-7 days of menstrual flow.

Cutaneous examination showed a single, well defined, annular ulcer approximately 2 cm in size with regular, rolled out violaceous margin, erythematous floor and pale granulation tissue over left lower part of labia majora. [Figure-1a] A single, well defined, oval, erythematous ulcer of size approximately 1 cm size with regular margin and single erythematous papule over right labia minora and majora respectively. [Figure-1b] A single, well defined, oval aphthous ulcer was present over left buccal mucosa [Figure-1c]. Hair and nail examinations were found to be normal. Looking at the history & examination provisional diagnosis kept was behcet's disease.

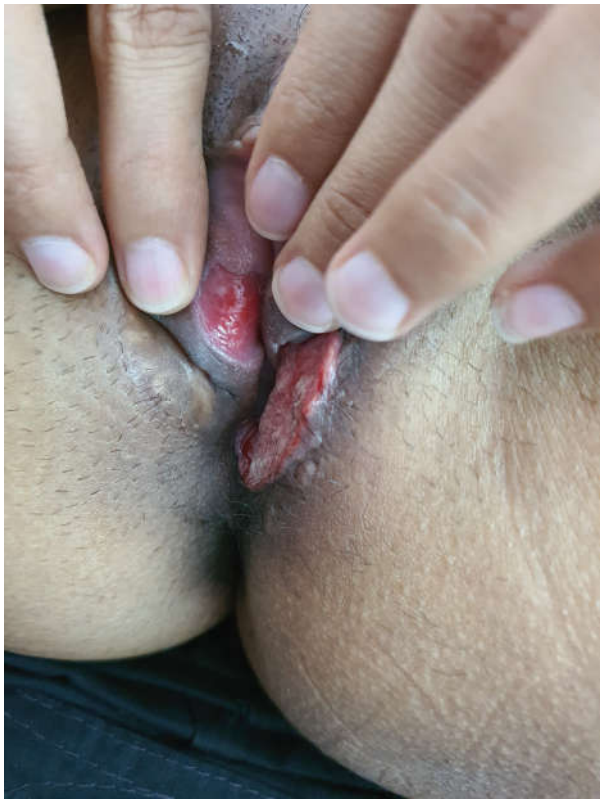

Fig. 1a: Single, well defined,annular ulcer approximately 2 cm in size with regular, rolled out violaceous margin, erythematous floor and pale granulation tissue over left lower part of labia majora.

Laboratory examination of blood showed raised ESR and CRP. Serology for HIV and Syphilis were non reactive. Pathergy and Montox test were negative. Histopathological examination of skin biopsy taken from ulcer over left labia majora showed focally hyperplastic epithelium. The

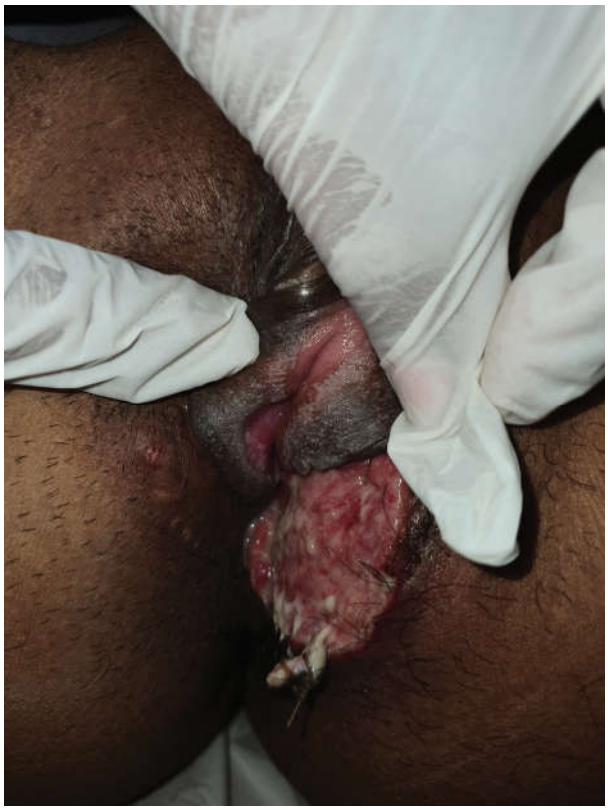

Fig. 1b: Single, well defined, oval, erythematous ulcer of size approximately 1 cm size with regular margin over right labia minora and single erythematous papule over right labia majora.

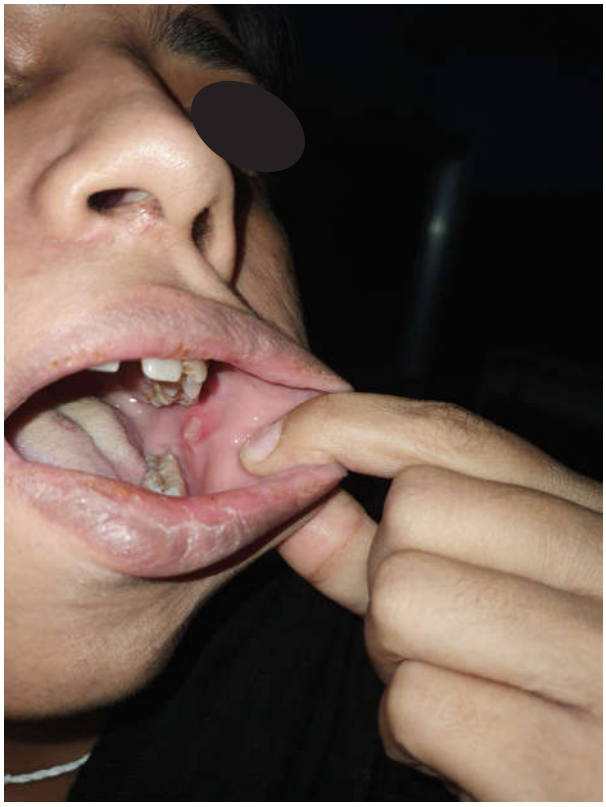

Fig. 1c: A single, well defined, oval aphthous ulcer over left buccal mucosa.

underlying stroma contains granulation tissue comprised of plenty of polymorphs, lymphocytes, plasma cells and macrophages with vascular proliferation and hemorrhage with no epitheloid granuloma. [Figure-2] Findings with suppurative inflammation were suggestive of pyoderma gangrenosum .The gram stain examination showed moderate number of gram negative bacilli and plenty of gram positive bacilli. No Donovan bodies were found. Chest X-ray showed no significant findings .Patient was diagnosed as a case of behcet's disease with ulcer of pyoderma gangrenosum over the genitals.

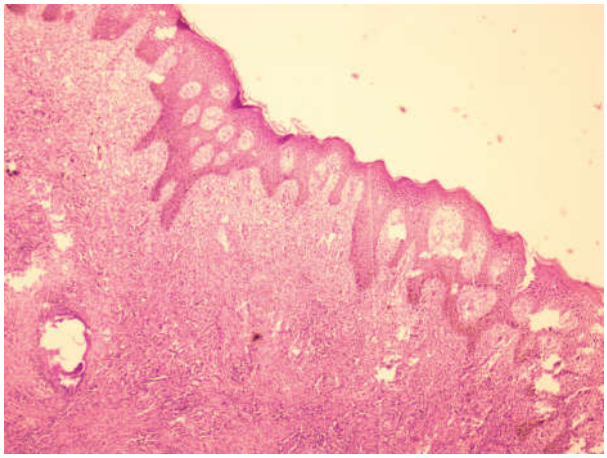

Fig. 2: Histopathology showed focally hyperplastic epithelium. Dermis showed granulation tissue comprised of plenty of polymorphs, lymphocytes, plasma cells and macrophages with vascular proliferation and hemorrhage.

Patient was treated with tab Dapsone 100 mg HS, tab Doxycyclin 100 mg BD, tab Prednisolone 40 mg OD to start with , which was gradually tapered and stopped after 1 month. The lesions improved with scars and showed no recurrences at the end of 4 months [Figure-3]

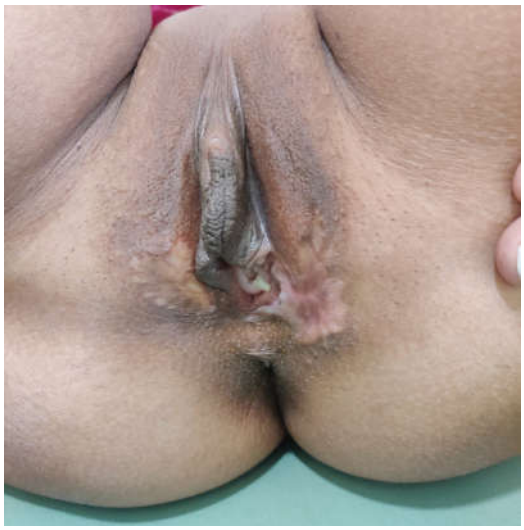

Fig. 3: Healed scar at the end of 4 months.

Discussion

PG was first described by Brocq in 1916 as "phagedenisme geometrique" later named by Brunsting, Goeckerman and O'Leary in 1930, It is a misnomer also known as dermatitis gangrenosa and phagedenic pyoderma.[4]

Etiology of PG includes neutrophil dysfunction, immunological & genetic factors, infection with Chlamydia pneumoniae, drugs like propylthiouracil, tyrosine kinase inhibitors, isotretinoin, TNFα inhibitors, epidermal growth factor receptor inhibitor and granulocyte-colony stimulating factor .About 50% cases are associated with inflammatory bowel disease such as ulcerative colitis or Crohn disease, arthritis, monoclonal gammopathy, malignancy, myeloproliferative disorder[4] and rarely Behcet's disease.[3]

PG usually starts as an inflamed nodule which progresses rapidly to an ulcer with an indurated and undermined purplish edges. The borders of ulcers extends peripherally in rough, serpiginous configuration. It is surrounded by crops of small, discrete pustules with inflammatory areola. Within a few days,the centre of the pustule softens and becomes an ulcer as seen in our patient. Clinically, PG is classified into the following four varieties:ulcerative,pustular, bullous, vegetative, peristomal and malignant.

The skin biopsy is done to exclude other causes of cutaneous ulceration from the active border of ulcer and to allow specimens to be sent for bacterial, mycobacterial and fungal cultures.

Table 1: Diagnostic criteria for Behçet's syndrome.

Criterion	Required features
Recurrent oral ulceration	Aphthous (idiopathic) ulceration, observed by clinician or patient, with at least three episodes in any 12-month period
Plus any two of the following:	
Recurrent genital ulceration	Aphthous ulceration or scarring, observed by clinician or patient
Eye lesions	Anterior or posterior uveitis cells in vitreous in slit-lamp examination; or retinal vasculitis documented by ophthalmologist
Skin lesions	Erythema nodosum-like lesions observed by clinician or patient; papulopustular skin lesions or pseudofolliculitis with characteristic acneiform nodules observed by clinician
Pathergy test	Interpreted at 24 to 48 hours by clinician

BD and PG, both are neutrophilic dermatoses. Oral and genital ulcerations are seen in both diseases.[5] but the frequency is absolutely different.

Orogenital ulcers are commonly seen in BD, while is rare in PG.[6] In BD, oral ulcers tend to be small and round, with well circumscribed margins, erythematous halo and yellow to grey floor. The lesions resemble those of recurrent aphthous stomatitis. Genital ulcers are similar to oral ulcers and can cause scar formation.There is no pathognomonic laboratory tests in Behçet syndrome; thus the diagnosis is made on the basis of the clinical findings or on diagnostic criteria from International Study Group for Behcet's Disease[7] (table-1) Our patient was diagnosed as Bechet's disease fulling the criteria in the form of recurrent oral ulceration , eye complains and genital ulcer.

Pustular lesions can be the initial skin manifestation and skin hyperreactivity or pathergy is often found in both diseases. In PG, the ulcers of the genital mucosa appear similar to those skin, which are comprised of a shallow burrowing ulcers with irregular margins and a ragged purple overhanging edges,which was seen in our case. Rapid local destruction may occur in PG.

In PG, vascular involvement ranges from none to fibroid necrosis, and in most cases a neutrophilic infiltrate is present with limited vascular damage. Conversely, in BD, mononuclear cell vasculitis with variable fibrin deposition or leukocytoclastic vasculitis is usually found.

There is a second thought that there is a clinical and histological overlap between PG and BD.[5] Skin lesions of PG and BD may show similar range of histological appearances, including a neutrophilic vascular reaction. Our case showed no necrotic changes with fibrinoid material in the vessel wall favouring PG.

Glucocorticoids, sulfa drugs, dapsone and immunosuppressive agents, such as cyclosporine, azathioprine or cyclophosphamide are therapeutic agents used to treat both PG and BD.[8] Our patient responded with dapsone and had no recurrences after 4 months.

According to history of patient, she is labeled as case of BD who concurrently developed lesions of PG thus it can be accepted as the overlap of PG and BD.

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