An Interesting Case of Recurrent Hemorrhagic Blisters

Sir,

Pyoderma gangrenosum is an uncommon sterile neutrophilic disease under the broad group of neutrophilic dermatoses which includes other dermatoses like—Sweet syndrome, Behcet disease, rheumatoid neutrophilic dermatoses, neutrophilic eccrine hidradenitis, bowel associated dermatitis arthritis syndrome etc.[1] Although classical ulcerative PG is the most common presentation, several variants have been described like pustular, vegetative, parastomal, granulomatous, and bullous.[2] Bullous PG is a rare variant that has been reported to be associated with myeloproliferative disorders and presents with recurrent hemorrhagic bullae.

A 62 years old female, a known case of chronic myeloid leukemia (CML) for the past 5 years, was referred to dermatology OPD with recurrent painless blood-filled blisters over the body for the last 2 years duration. She was diagnosed with a case of CML 5 years back and she was started on Imatinib since then with intermittent accelerated as well as a couple of phases of blast crisis requiring systemic steroids. The lesions which mostly coincided with accelerated phase and/or blast crisis would begin as blood-filled blisters approximately the size of a pea which would grow to approximately 4–6 cm and rupture spontaneously to release the contents and heal with dark spots. They were not associated with any itching or pain. She also gives the history of similar lesions in the oral cavity with a similar course. Her vital parameters and systemic examination were within normal limits. Dermatological examination revealed multiple polysized discrete flaccid as well as tense hemorrhagic bullae in different stages of evolution, distributed over both upper and lower limbs, largest measuring approximately 4 × 4 cm located over the dorsum of the left hand. [Figure 1a–c] Oral and genital mucosa were uninvolved. Nikolsky’s sign, bulla-spread sign, and test for pathergy were negative. All routine blood tests were within normal range. Biopsy from the bulla from the dorsum of the left hand revealed hyperkeratosis, acanthosis, and sub-epidermal split filled with fibrin and neutrophils. Dermis showed dense peri-vascular and peri-adnexal inflammatory infiltrate mostly comprising of neutrophils and occasional lymphocytes and plasma cells. [Figure 2a–d] However, there were no acantholytic cells, atypical malignant cells, granulomas, or features of vasculitis. Direct immunofluorescence was negative for IgG, IgA, and C3. The clinical and histopathological features were suggestive of bullous pyoderma gangrenosum. Considering the use and benefit of systemic steroids especially in blast crises of CML as well as the drug of choice in PG, we started her on 0.5 mg/kg/day of prednisolone. Emphasis was given on topical therapy consisting of potassium permanganate soaks followed by the application of 0.05% fluticasone cream. The lesions healed in a week with healthy crusts, scaling, and post-inflammatory hyperpigmentation. [Figure 3a and b] Prednisolone was slowly tapered and the patient is presently on 5 mg prednisolone per day and she is in remission since then.

Pyoderma gangrenosum was first described by Brocq in 1916 and further characterized in 1930 by Brunsting et al.[3] They believed that it was caused due to Streptococcus which led to gangrene and hence the misnomer- pyoderma gangrenosum. Bullous PG was described much later in 1972 by Perry and Winkelmann as hemorrhagic bullae rapidly progressing to superficial ulcers.[2] Bullous PG is a rare variant that has many differentiating features from...
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classical PG. Bennet et al.\(^4\) considered bullous PG as “Atypical Pyoderma Gangrenosum” which is characterized by hemorrhagic bullae and were present mainly on upper extremities as compared to classical ulcerative PG which commonly affects legs.\(^5\) As compared to destructive, deep ulcers in ulcerative PG, the lesions in the bullous variant are superficial. Bullous PG has been seen to be associated with hematological malignancies. The most commonly reported hematological malignancy with PG is myelodysplastic syndrome (MDS) followed by monoclonal gammopathy of undetermined significance (MGUS) and acute myeloid leukemia (AML).\(^6\) Although Bullous PG is often discussed in association with AML; ulcerative PG is the most common type of PG associated.\(^6\) Although bullous PG mostly presents after the malignancy has been diagnosed, rarely bullous PG may be the presenting symptom of the underlying malignancy.\(^7,10\) This fact underlines that any patient with bullous PG should be investigated to rule out hematological malignancies. Although there may be many clinical and histopathological overlaps between features of malignancy-associated PG and Sweet syndrome; they should be considered as diseases belonging to the same spectrum.\(^5\) Treatment of underlying hematological malignancy is the cornerstone of management of bullous PG. In addition, systemic and/or topical corticosteroids at doses ranging from prednisolone equivalent of 0.3 to 1 mg/kg/day may be needed for varying duration for relapses.\(^2\) We report this interesting case for its rarity.

**Declaration of patient consent**

The authors certify that they have obtained all appropriate patient consent forms. In the form, the parent has given the consent for images and other clinical information to be reported in the journal. The parent understands that names and initials will not be published and due efforts will be made to conceal patient identity, but anonymity cannot be guaranteed.

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Nil.

**Conflicts of interest**

There are no conflicts of interest.

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