Pyoderma gangrenosum associated with mantle cell lymphoma

Sir,

Pyoderma gangrenosum (PG) is as an idiopathic, ulcerative, chronic inflammatory skin disease of uncertain etiology. We report a case of PG associated with mantle cell lymphoma (MCL).

A 40-year-old female patient presented with a large ulcer on the anterolateral aspect of left lower leg along with fever, weakness, cervical and axillary lymphadenopathy, severe anemia, and a history of repeated blood transfusion in the past. There was no history of any change in bowel habits or joint pain or history of any drug intake. The cutaneous lesion started as a small nodule, which subsequently eroded to form an ulcer of size 12 cm × 10 cm within 20 days. The ulcer was indurated, tender with an elevated bluish border, undermined edge, and multiple sieve-like openings discharging serosanguinous fluid [Figure 1a and b]. Except splenomegaly, findings of other systemic examinations were unremarkable. Laboratory investigation revealed the following: Hb - 4 gm%, total leukocyte count - 90,000/mm^3, predominantly normocytic normochromic red blood cells, thrombocytopenia, and marked leukocytosis with presence of 70% blast cells. Fine-Needle Aspiration cytology of cervical lymph node showed monomorphic population of small lymphoid cells with mitosis and numerous small lymphocytes with scanty cytoplasm and round nuclei, suggestive of the cytological picture of MCL. Mantle cells were also found in the biopsy of axillary lymph node [Figure 2c and d]. Flow cytometric analysis detected an atypical population of lymphoid cells (73% of the total) showing coexpression of CD5 and CD19 with kappa light chain restriction, expressing CD20, CD22, CD79a, CD79b, surface IgM, and CD45 and was negative for CD23, CD10, and CD103 [Figure 3]. Overall findings were consistent with a B-cell lymphoproliferative disorder, the leukemic phase of MCL.

Punch biopsy from the edge of the ulcer showed a dense diffuse infiltration of acute and chronic inflammatory cells mostly neutrophils along with necrosis of the epithelium [Figure 2a and b]. On the basis of laboratory investigation and histopathological study, a diagnosis of PG secondary to

MCL was made. The patient was treated with oral dapsone 100 mg/day (G6PD was normal); prednisolone 40 mg/day; topical clobetasol propionate ointment, and moist vaseline gauze dressing along with three units of blood transfusion. The ulcer started healing and decreased in size within

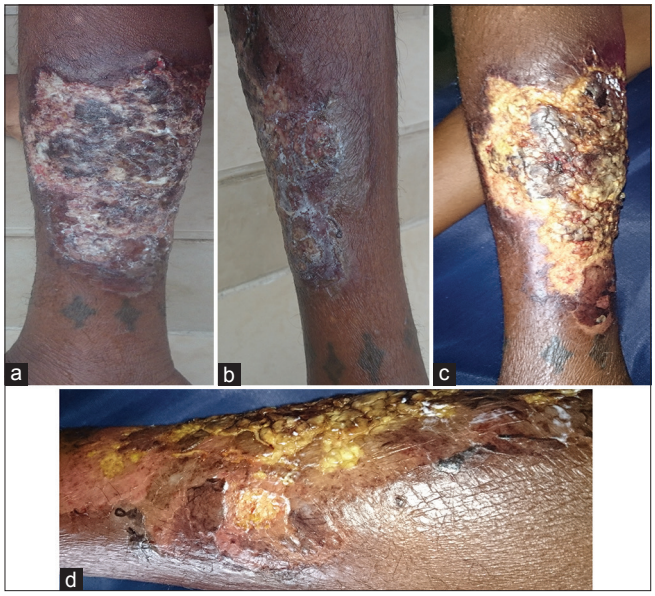

Figure 1: (a and b) Ulcer with elevated bluish border, undermined edge, and multiple sieve-like openings discharging serosanguinous fluid. (c and d) Healing of ulcer after 7-10 days of treatment

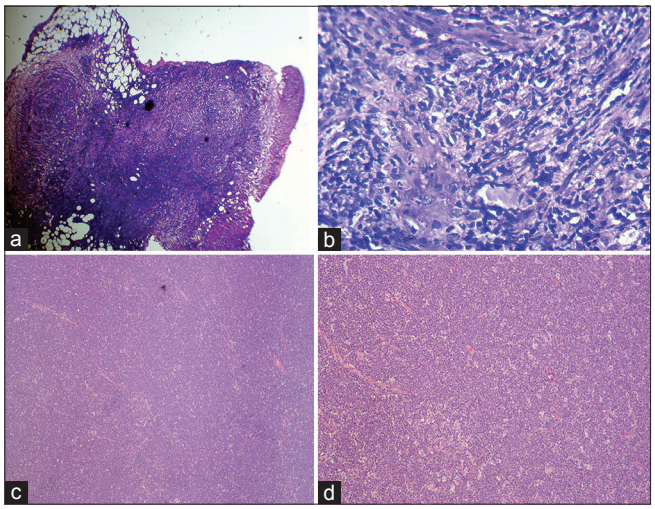

Figure 2: (a and b) Histopathology of ulcer ×10 and ×40, respectively, showing dense diffuse infiltration of acute and chronic inflammatory cells. (c and d) Histopathology of lymph node ×40 and ×100, respectively, showing monomorphic population of small lymphoid cells with mitosis and mantle cells. H and E Stain

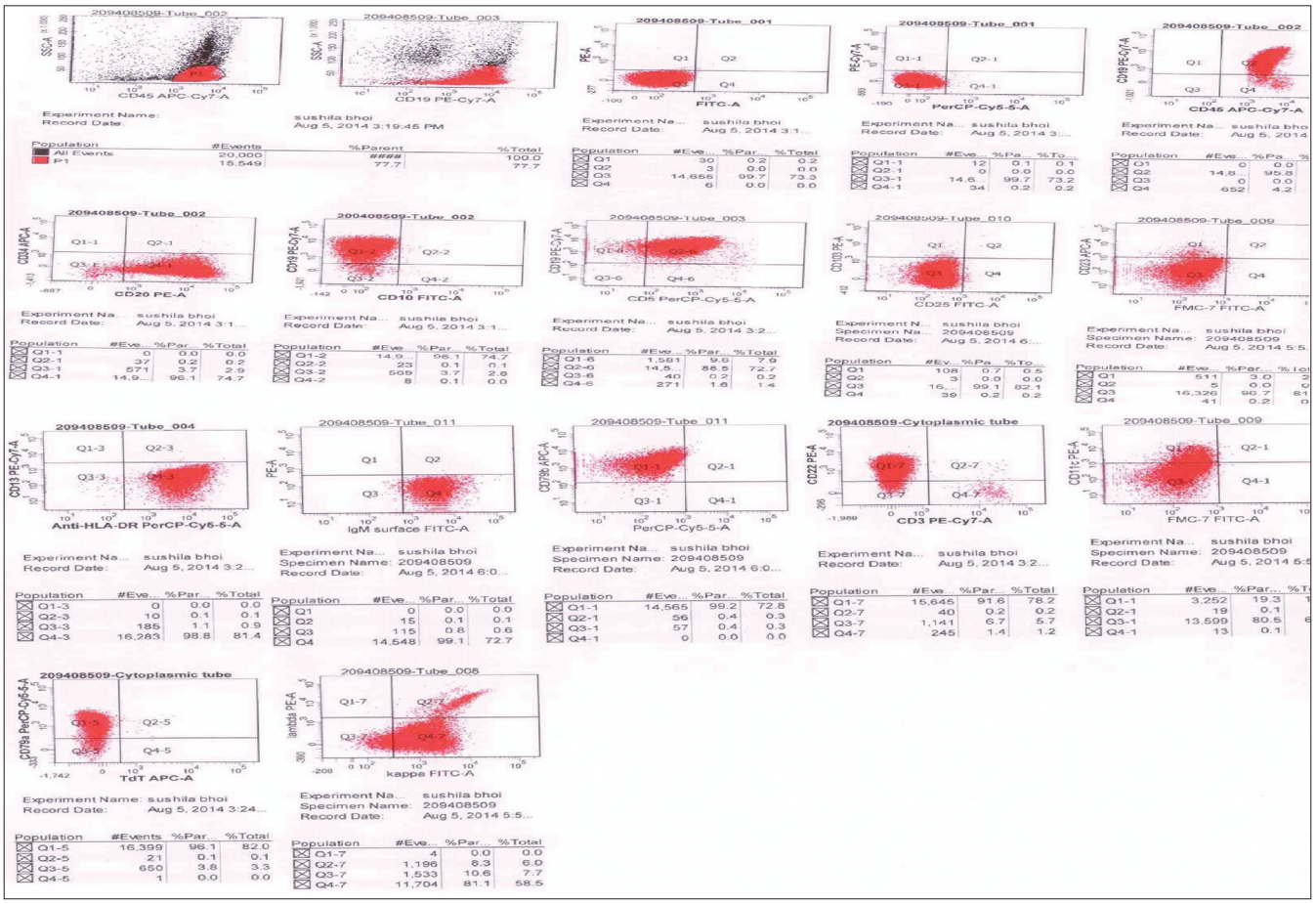

Figure 3: Flow cytometry analysis report

7–10 days [Figure 1c and d] and the patient was referred to a higher center for further management of MCL.

PG is a rare chronic destructive inflammatory skin disease, which begins as a tender erythematous nodule, then forms a pustule, followed by rapid central necrosis characterized by edematous and dusky overhanging borders with a surrounding margin of erythema. It is often associated with various systemic diseases, most notably inflammatory bowel disease (IBD) and also appears in the setting of various malignancies.[1] The most common reported malignancy is acute myeloid leukemia and the association with solid tumors has also been reported.[2]

The etiology of PG is unknown but tumor necrosis factor-α (TNF-α) is believed to play a major role, possibly through its stimulatory action on the neutrophil chemoattractant, IL-8.[3,4] Similarly, higher than normal levels of cytokines such as TNF-α, IL-8, and vascular endothelial growth factor (VEGF) are often reported in various hematological malignancies.

The recommended firstline treatment in PG is steroids or other immunosuppressive drugs.[5]In our patient it was the ulcerative variant of PG secondary to MCL, which responded dramatically to oral prednisolone and dapsone. As she was referred to an oncology center, we lost her to follow up and could not ascertain what therapy she received and whether the ulcer recurred.

The co-existence of PG and MCL in our case could be attributed to common cytokine milieu such as TNF-α, IL-8, and VEGF in both these conditions and hence the presence of MCL might have induced the occurrence of PG. We report the case for its rarity as PG occurring in the setting of MCL has not been reported in the scientific literature.

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Conflicts of interest

There are no conflicts of interest.

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