Anaplastic Large Cell Lymphoma Presenting as Pyoderma Gangrenosum

Dear Editor,

Pyoderma gangrenosum is a rare, noninfectious neutrophilic dermatosis commonly associated with underlying systemic diseases. Estimates vary, but the most frequent disease associations include inflammatory bowel disease (IBD) (20–30%), rheumatoid arthritis (10%), hematological malignancies or monoclonal gammopathy (5%), and other visceral malignancies (5%).[1-3]

We report a recalcitrant case diagnosed as pyoderma gangrenosum showing no response to conventional treatment. On further investigations, it was unveiled that the patient had anaplastic large cell lymphoma (ALCL).

A 21-year-old female presented to us with complaints of multiple painful nodules and ulcers over her abdomen and lower limbs for 5 months [Figure 1a]. On reporting to us, a complete investigation panel (complete blood picture, X-ray chest, and the Mantoux test) was done, and no significant findings were noted. Histopathology showed diffuse interstitial infiltrates involving the entire dermis composed of lymphocytes, histiocytes, numerous neutrophils, nuclear debris, and plasma cells, findings suggestive of pyoderma gangrenosum [Figure 2a], for which the patient was treated with oral prednisolone 40 mg/day and ulcer care, but showed no response.

Following computed tomography (CT) and magnetic resonance imaging (MRI), multidepartmental consultations advised us to continue treating as pyoderma gangrenosum; however, the patient was lost to follow-up. Three months later she presented with an insidious onset of excruciatingly painful swelling over the right knee and left ankle joint of size (9 × 8 cm^2) and (3 × 2 cm^2), respectively, and aggravation of preexisting lesions [Figures 1b and c]. Routine investigations and radiological examinations were done in which necrotic lymph nodes (LN) were seen on contrast-enhanced computed tomography of the abdomen, pelvis, and chest. Ultrasonography (USG) of the swelling suggested an abscess. MRI suggested the same with the popliteal artery pushed to the lateral side, threatening vascular compromise, for which incision and drainage was done that showed necrotic fatty discharge.

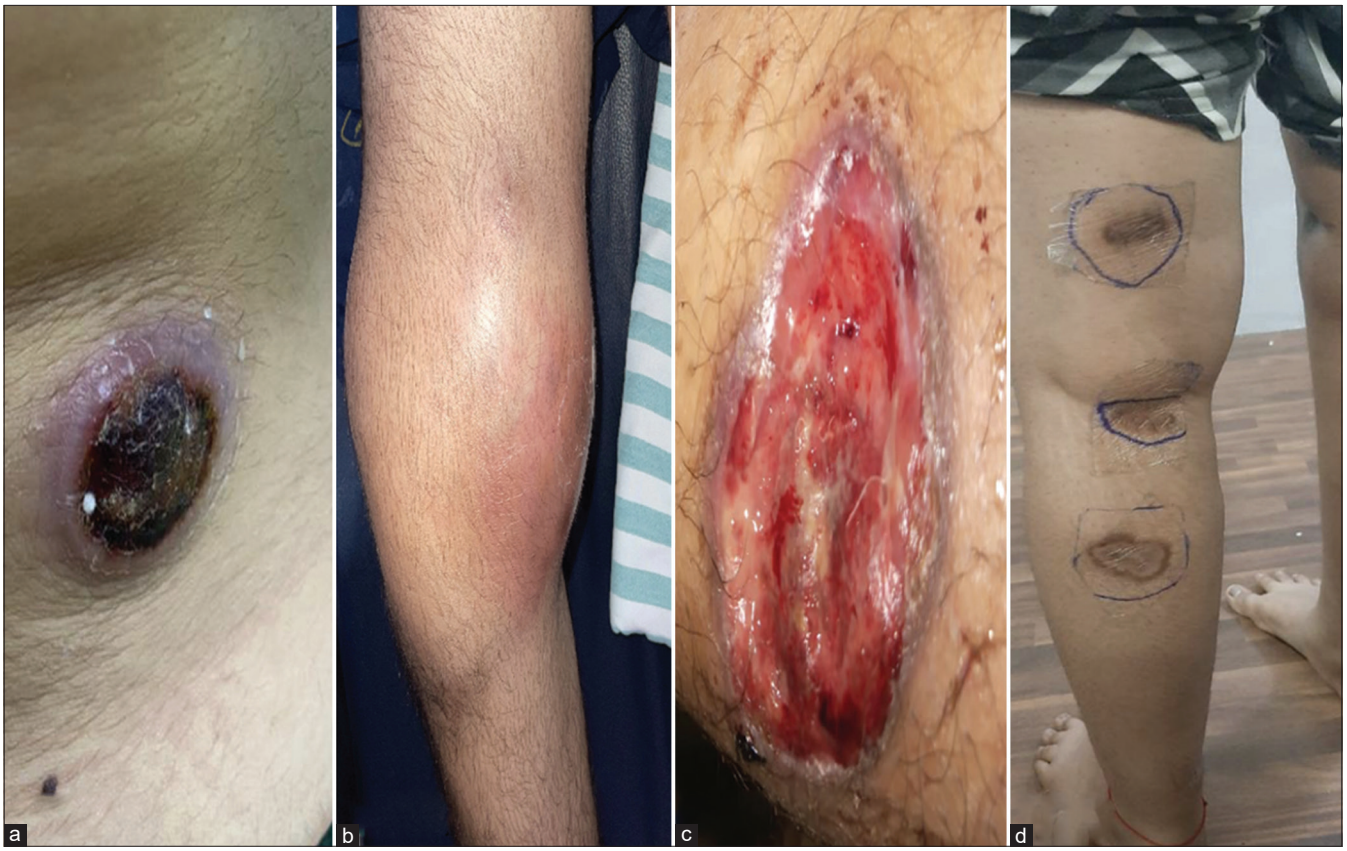

Figure 1: (a) Initial lesions showing an ulcer of 2 × 2 cm^2 with everted margins and hemorrhagic crusting; (b) Insidious swelling over the right knee of size (9x8 cm^2) associated with erythema and excruciating pain; (c) Flared up previous lesions; and (d) Complete recovery of lesions after the fourth cycle of CHOEP

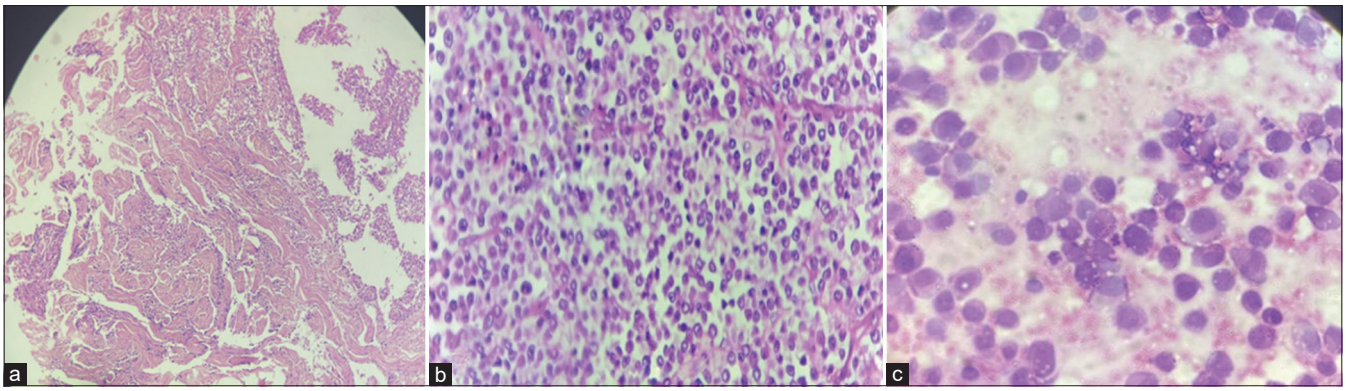

Figure 2: (a) Diffuse interstitial infiltrate involving the entire dermis. Infiltrate composed of lymphocytes, histiocytes, numerous neutrophils, nuclear debris, plasma cells findings suggestive of pyoderma gangrenosum (H &E, 10x); (b) Incision and drainage sample showing diffuse proliferation of cells which exhibited moderate cytoplasm, large nuclei, dense nuclear membrane, and prominent nucleoli. About 4–5 mitotic figures were noted per field. Features suggesting high-grade lymphoma (H &E, 40x); and (c) Right inguinal lymphnode fine needle aspiration cytology showed monomorphic lymphoblasts which were suggestive of lymphoma (H & E, 50x)

Histopathological examination showed a proliferation of cells that exhibited moderate cytoplasm, large nuclei, dense nuclear membrane, and prominent nucleoli. About 4–5 mitotic figures per field suggested high-grade lymphoma [Figure 2b]. On the right inguinal lymph node, fine needle aspiration cytology was done, which showed monomorphic lymphoblasts that were suggestive of lymphoma [Figure 2c].

The diagnosis of anaplastic large cell lymphoma ALK (+) stage III was confirmed with immune histochemistry, which showed diffuse and strong staining for CD30 and positive anaplastic lymphoma kinase. The patient was started on the CHOEP regime inj. cyclophosphamide 800 mg + inj. doxorubicin 40 mg + inj. vincristine 2 mg + tab.etoposide 150 mg+ tab. prednisolone 60 mg ; four cycles after the commencement of the treatment, complete rapid resolution of ulcers was noted [Figure 1d].

Curth's criterion is used for the diagnosis of cutaneous paraneoplastic syndromes; in our case, it was used to derive the disease pathology.[4] Also, the absence of atypical T cells in the histopathology of our case supported the diagnosis of paraneoplastic pyoderma gangrenosum and not an ulcer secondary to ALCL.

In the present case, our diagnosis was ALK-positive ALCL with systemic involvement (Lymph node (LN) involvement as seen in MRI);[5] however, it is hard to comment on the simultaneous course of ALCL and pyoderma gangrenosum, as the only initial presenting complaints were the ulcers, and their treatment was backed by histopathology might have been a reason for the delay in thorough systemic investigations.

The presentation of ALCL as neoplastic pyoderma gangrenosum is rather rare and lacks reports. To the best of our knowledge, this is the second report after Saito S *et al.*, where similar complaints were reported in a 5-year-old girl.[6]

With this letter, we would like to state that even though rare, paraneoplastic dermatoses should be kept in mind when there is a treatment-resistant disease, out-of-proportion symptoms of a disease, and sudden changes in the patient's chief complaints. Hence, as dermatologists, we must be vigilant and proactive in the management of treatment-resistant conditions.

Declaration of patient consent

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

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Conflicts of interest

There are no conflicts of interest.

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