A rare case of multiple pyoderma gangrenosum co-mobilized with pseudoxanthoma elasticum

Dear Editor,

A 70-year-old woman had been suffered from clustering of rice-sized yellowish nodules and wrinkles on skin surface of her neck and axillary from early childhood under the diagnosis of pseudoxanthoma elasticum (PXE). Noninducible itching and pain arose on the bilateral groin 2 years before the consultation to our department followed by small erosions’ formation. Following 1 year later, because those erosions were aggravated to deep ulcers intractable against local treatment, she was referred to our department.

Walnut to palm-sized and well-defined painful ulcers surrounded by erythema was found on the both inguinal (Figure 1A), left thigh along with surgical linear scar (Figure 1B), and patella. On the other hand, the multiple yellow nodules were presented on the neck, axilla, medial elbows, lower abdomen, and groin, forming crepe-like atrophic plaques (Figure 1C). Skin biopsy from the margin of the right inguinal ulcer shown in Figure 1A revealed dense inflammatory cell infiltration from the upper dermis to the adipose tissue. Inflammatory cells were mainly neutrophils, histocytes, and lymphocytes. The necrotic ulcers were diagnosed as multiple pyoderma gangrenosum (PG). In the tissue of the yellow nodule on the right axilla, deposition of basophilic denatured elastic fibers and diffuse calcium on the reticular dermis were detected by EvG staining (Figure 1D) and Kossa staining (Figure 1E), respectively, and PXE was diagnosed. Since, daily administration of 25 mg (0.5 mg/kg) of oral prednisolone was insufficient, the dose increased to 50 mg/day, then, all necrotic ulcers began to noticeably shrink in response to treatment. No adverse event and recurrence were observed during the tapering off.

Pseudoxanthoma elasticum is a serious hereditary disorder of connective tissue that involves the elastic tissue in the skin, blood vessels, and eyes. In this case, severe longitudinal calcification of the retroperitoneal arteries and coronary arteries and angioid streaks of the eyes was involved as related diseases of PXE. There was no history of inflammatory bowel disease, blood dyscrasia, or RA that may influence in PG. The left buttock ulcer coincided with the incision in hip surgery, and the four ulcers in the groin coincided with the PXE-related eruption. 20%-30% of PG develops with minor trauma or external stimuli, which is called as pathergy. Indeed, the ulcerations in this case were attained on the friction or traumatic sites such as groin and patella in addition to a surgical wound on the thigh.

To our knowledge, no case that complicated PG and PXE has not been reported yet. Similar to the concept of pathergy, we expected the possibility on the skin fragility and dermal calcification on the PXE lesions could exacerbate traumatic skin damage followed by ulcer formation. Although skin biopsy specimen was obtained across the edge of the left inguinal ulcer in order to evaluate if PG was coincident with PXE, the dermal change of PXE was not overlapped with ulcer surface (Figure 1F).

We experienced here a rare case presenting both of PG and PXE lesions simultaneously. Further accumulation of similar cases helps us to early notice the development of pyoderma gangrenosum on PXE background.

DECLARATION SECTION

The written informed consent was obtained from the patient.

Informed Consent: Written informed consent was obtained from the patients.

Animal Studies: N/A.

CONFLICTS OF INTEREST

Dr Manabu Fujimoto is the Editor in Chief for the Journal of Cutaneous Immunology and Allergy. Management of the peer review process, and all editorial decision-making, for this article was undertaken by an Associate Editor.

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