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CORRESPONDENCE

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 axillar 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).[1] 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.[2] 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.[3] There was no history of inflammatory bowel disease, blood dyscrasia, or RA that may influence in PG.[4] 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.[5] 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.

Approval of the research protocol: N/A.

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

Registry and the Registration No. of the study/trial: N/A.

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.

Saki Iga
Atsushi Tanemura
Takumi Kamitani
Madoka Takafuji
Yuma Hanaoka
Noriko Arase
Manabu Fujimoto

Department of Dermatology, Osaka University Graduate School of Medicine, Suita, Japan

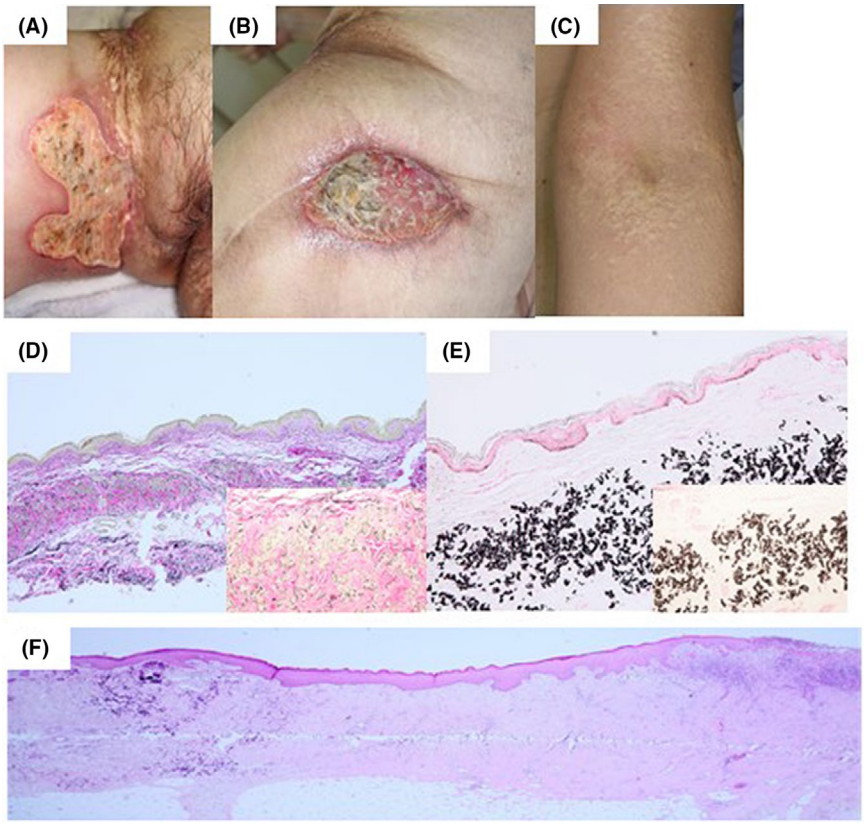

FIGURE 1 (A, B) Clinical features of PG lesions at the 1st visit. The undermining ulcers with yellow-to-black turbid necrotic tissue developed on the right inguinal and left thigh along with surgical scar. (C) Clinical feature of PXE lesion at the 1st visit. The yellow nodules developed on the medial elbows, forming a crepe-like atrophy plaque. (D) Histopathological feature under EvG staining (×20). Inbox indicates ×200 enhance to show fragmentation of dermal elastic fibers. (E) Histopathological feature under Kossa staining (×40). Inbox indicates ×200 enhance to show calcification of dermal elastic fibers. (F) Low power magnification of H-E staining shared both of PG and PXE lesions

Correspondence
Atsushi Tanemura, M.D., Ph.D., Department of Dermatology Integrated Medicine, Osaka University Graduate School of Medicine, 2-2 Yamadaoka, Suita, Osaka 565-0871, Japan
Email: tanemura@derma.med.osaka-u.ac.jp

ORCID

Saki Iga https://orcid.org/0000-0001-9285-4334
Atsushi Tanemura https://orcid.org/0000-0002-5239-8474
Noriko Arase https://orcid.org/0000-0003-3966-3437
Manabu Fujimoto https://orcid.org/0000-0002-3062-4872

REFERENCES

1. Von den Driesch P. Pyoderma gangrenosum: a report of 44 cases with follow-up.Br. J Dermatol. 1997;137:1000–5.
2. Uitto J, Jiang Q, Varadi A, et al. Pseudoxanthoma elasticum: Diagnostic features, classification, and treatment. Expert Opin Orphan Drugs. 2014;2(6):567–77.
3. Marconi B, Bodyr I, Campanati A, et al. Pseudoxanthoma elasticum and skin: Clinical manifestations, histopathology, pathomechanism, perspectives of treatment. Intractable Rare Dis Res. 2015;4(3):113–22.
4. Yamamoto T. Pyoderma gangrenosum: an important dermatologic condition occasionally associated with rhematic disease. World J Rheumatol. 2015;5:101–7.
5. Callen JP, Jackson JM. Pyoderma gangrenosum: an update. Rheum Dis Clin North Am. 2007;33:787–802.

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