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LETTER TO THE EDITOR

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WILEY

Cutaneous ulcer resembling pyoderma gangrenosum in a patient with antiphospholipid syndrome

Dear Editor,

Cutaneous manifestations of antiphospholipid syndrome (APS) vary from livedo reticularis to cutaneous necrosis, with the appearance of associate cutaneous ulcers with sharp margins, typically over the legs.[1] Classic pyoderma gangrenosum (PG), also known as ulcerative type, is characterized by rapidly progressive ulcers with violaceous

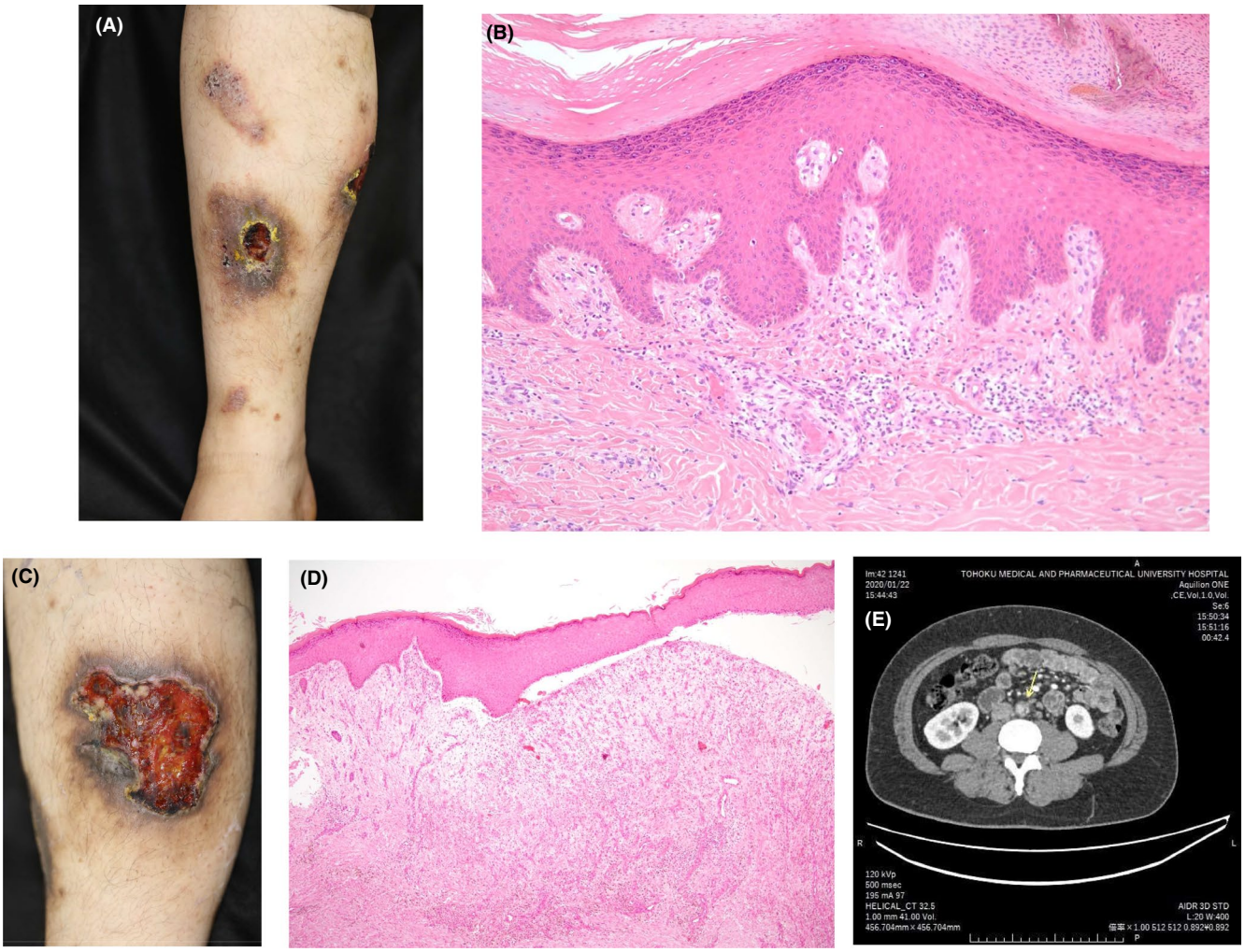

FIGURE 1 A, Necrotic ulcers with dense brown pigmentation on the patient's right leg. B, Microscopic findings show thrombi with cell inflammation and capillary dilatation in the upper to middle dermis. C, Classical and rapidly progressive pyoderma gangrenosum ulcer showed an elevated, violaceous, undermined border, with a necrotic and hemorrhagic base on his right leg. D, Neutrophils and lymphocytic cells were identified around the dermal vessels with capillary dilatation and thrombi in the dermis to subcutaneous fat tissue. E, Abdominal contrast-enhanced computed tomography revealed arterial thrombosis. White arrow indicates thrombi in the abdominal aorta

indurated borders. The diagnosis remains challenging due to the lack of a gold standard diagnostic test.[2]

A 39-year-old man presented with a reddish necrotic and erosive lesion and cutaneous ulcers all over his legs that had appeared one month earlier. Physical examination revealed various sized necrotic ulcers include papules, vesicles, and pustules over both his legs, along with dense brown pigmentation (Figure 1A). Microscopic findings showed dermal fibrosis, intravascular hyalinized material of fibrin-containing thrombi with cell inflammation and capillary dilatation in the upper to middle dermis (Figure 1B). Antibiotic treatment alone was insufficient, and the lesion continued to gradually increase in size. He had abrupt appearance of rapidly enlarging and hemorrhagic ulcerations on his right leg. Cutaneous ulcers in the lateral femoral region of the right leg were found with undermined, overhanging, dusky purple edges, and surrounding induration and erythema (Figure 1C). Histopathological examination revealed a severe dermal infiltrate comprised of neutrophils and lymphocytic cells and capillary dilatation and thrombi in the dermis to subcutaneous fat tissue (Figure 1D). The clinical features of the cutaneous ulcer suggested a diagnosis of PG. In contrast, histopathological findings showed thrombi and vascular damage in the dermal capillary vessels with neutrophilic infiltrate.

Laboratory examinations showed increased anti-cardiolipin antibodies (112 U/mL; normal range, <9 U/mL) and anti-β2 glycoprotein I antibodies (>125 U/mL; normal range 0-3.4 U/mL), and the lupus anticoagulant was detected. Abdominal contrast-enhanced computed tomography revealed thrombi, contraction, and occlusion of the abdominal aorta and iliac arteries (Figure 1E). Ultrasonography of his veins showed thrombi in the femoral veins, popliteal veins, and flounder veins. Based on these findings, he was diagnosed with APS. He responded to anticoagulation treatment with warfarin alone and his cutaneous ulcer that had initially been diagnosed as PG improved.

Classic ulcerative PG is characterized by a peripheral erythematous inflammatory halo and the edges are erythematous, raised, and sometimes necrotic, with an undermining border, the size of which determines how rapidly the ulcer edge will evolve.[2,3] The present case presented with abrupt appearance of rapidly enlarging ulcerations over his legs that resembled PG with no atypical histopathological findings such as dermal infiltrate showing numerous neutrophils. There have been other reports of PG-like ulcers in patients with APS.[3,4]

Recent studies have also suggested that lupus anticoagulant and anti-cardiolipin antibodies could be an important risk factor for venous ulcers of the leg.[5] These antiphospholipid antibodies potentially cause repeated thrombi which lead to chronic damage of the skin and eventually to poorly healing venous ulcers over the long term. We propose that the pathogenesis of PG-like cutaneous ulcer in the present case could be related to some abnormalities in the coagulation/fibrinolysis pathways and an abnormal immune condition that involves antiphospholipid antibodies.

CONFLICT OF INTEREST

The authors declares no conflict of interest.

DECLARATION

Approval of the research protocol and Informed Consent were performed by Tohoku Medical and Pharmaceutical University, Sendai, Japan.

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

Animal Studies: N/A

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