A case of primarily facial pyoderma gangrenosum associated with Takayasu arteritis

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INTRODUCTION

Pyoderma gangrenosum (PG) is a type of neutrophilic dermatosis that shows noninfectious ulcers characterized by neutrophil infiltration of the skin.[1] PG is frequently associated with systemic diseases such as inflammatory bowel disease, rheumatoid arthritis, and hematologic disorders.[2] In Japan, PG is also a well-known complication of Takayasu arteritis (TA), yet only a few anecdotal reports are recorded worldwide.[3,4] Typically, patients with PG show skin lesions on the lower extremities; however, the PG lesions associated with TA are known to be more widespread than those without TA.[3] Ujiie et al[3] reported that 42.9% of the PG patients with TA (15 of 35) had lesions on the face and neck. We report a case of PG associated with TA that primarily affected the face and had been misdiagnosed as a facial skin infection.

Abbreviations used:

IL: interleukin
PG: pyoderma gangrenosum
TA: Takayasu arteritis

CASE REPORT

The patient was a 37-year-old Japanese man who had TA diagnosed 4 years before the skin manifestation. When he was 33 years old, he presented with left cervical pain followed by dizziness, a fainting episode, and fever that failed to respond to antimicrobial agents. Fluorodeoxyglucose-positron emission tomography/computed tomography and computed tomography imaging confirmed TA. Subsequently, the patient was prescribed systemic corticosteroids and immunosuppressive agents. Four years later he developed multiple pustules and subcutaneous abscesses within an erythematous plaque in the left buccal region. Facial skin infection was diagnosed at an outlying facility, and he was prescribed antimicrobial agents for approximately 6 months. However, his skin lesion increased and spread to the other cheek, trunk, and lower legs, after which he visited our institution. The major symptoms associated with TA were well controlled with oral betamethasone (1 mg/d) and tacrolimus (3 mg/d). Irregular-shaped reddish scarring plaques with some pustules and subcutaneous abscesses were present on each of his cheeks and on the lower jaw (Fig 1, *A*). In addition, a crusted ulcerative plaque with erythema was observed on the extensor surface of his right lower leg (Fig 1, *B*). Histopathologic examination of a biopsy section taken from the facial lesion found mild acanthosis, dilated capillaries in the papillary dermis, partial invagination of the epithelial component, and dense infiltrates of inflammatory cells in the dermis, most of which were neutrophils (Fig 2). Multiple bacterial cultures from facial pustules and abscesses showed no evidence of bacterial infection. Serologic tests were negative for rheumatoid factor and showed absence of a monoclonal gammopathy. Complete colonoscopy found no evidence of inflammatory bowel disease. We

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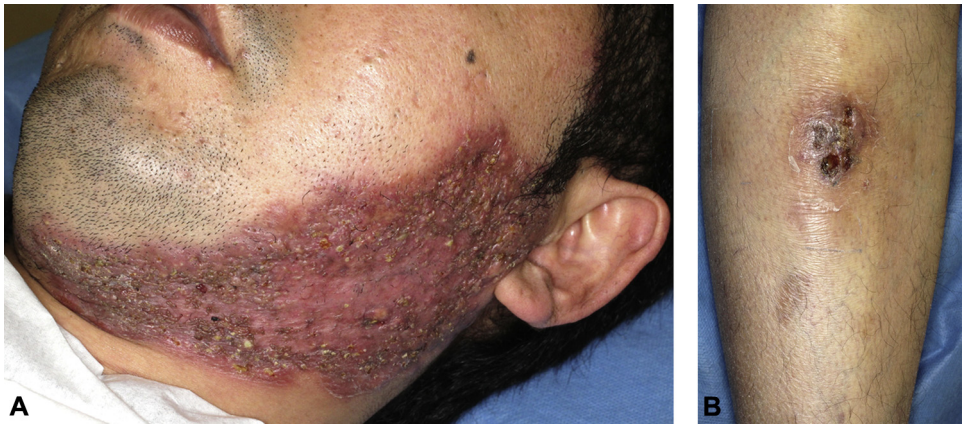

Fig 1. Clinical findings of the PG patient associated with TA. **A**, Irregular-shaped reddish scarring plaques with some pustules or subcutaneous abscess on the left cheek and lower jaw. **B**, Crusted ulcerative lesion with erythema on the extensor surfaces of the right lower leg.

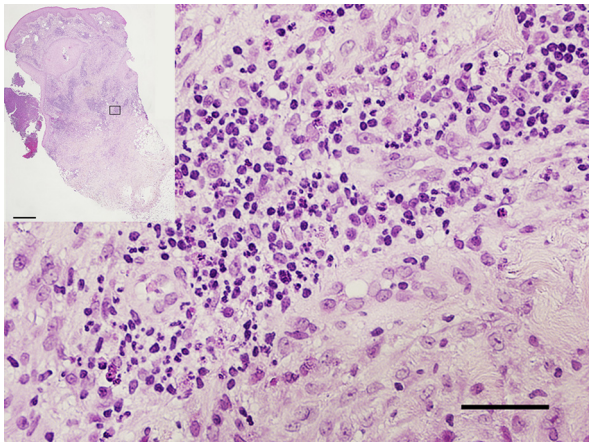

Fig 2. Histopathologic findings. Magnification of the rectangle in the low-power field image (inset) is shown in the center. A skin biopsy from the facial lesion found mild acanthosis, dilation of capillaries in the papillary dermis, partial invagination of the epithelial component, and infiltration of neutrophil-rich inflammatory cells to the dermis. (Hematoxylin-eosin stain; bar indicates 500 μm [inset] and 50 μm [magnified image]).

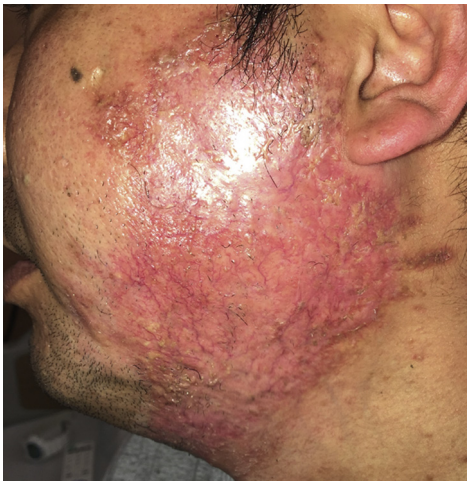

Fig 3. A clinical image after treatment with high-dose corticosteroids. Rose-pink scar with telangiectasia persisted on his face.

diagnosed him with PG. After increasing the dose of systemic corticosteroid dose to 50 mg/d of prednisolone the purulent inflammation of the facial plaque rapidly resolved, and his leg ulcer epithelized within a month. However, rose-pink scars with telangiectasia persisted on his face (Fig 3). The prednisolone dose was gradually decreased while ensuring that it did not cause flare up of the PG.

DISCUSSION

The case report presented here describes a male patient who had TA followed by PG, which had been misdiagnosed as a skin infection for 6 months.

TA is a rare, systemic vasculitis that predominantly involves the aorta and its branches; cutaneous lesions occur in up to 28% of patients.[5] Although the pathogenesis of TA remains unknown, upregulated proinflammatory cytokines, such as interleukin (IL)-6, IL-8, IL-18, IL-23, and tumor necrosis factor-α in patients with TA has been shown.[6] A very recent genomewide association study also found some susceptible genes associated with immunoregulatory pathways, including *IL6*.[7] On the other hand, significant upregulation of IL-6 and IL-8 has been reported in PG.[8] Overexpression of IL-8, IL-17, IL-23, and tumor necrosis factor-α has also been found in PG lesions.[2]

Considering the previously reported cytokine profiles in patients with TA and PG, the coexistence of the 2 diseases seems to be reasonable. Currently, the patient has facial scars. Early diagnosis and treatment is prudent to prevent this untoward complication.

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