Correspondence Takayasu's arteritis with pyoderma gangrenosum and erythema induratum

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The patient was a 21-year-old Japanese woman. At 16 years of age, she noticed ulcer with pain on her left lower leg. At 19 years of age, she visited our hospital for evaluation of the cutaneous lesion. As the clinical appearance and the histopathological findings (Fig. 1a, b), the lesion was diagnosed pyoderma gangrenosum. Because previous treatment of topical steroids had not been effective to the lesion, treatment with topical tacrolimus was tried to be successful. The lesion was healed with cribriform scarring after seven months therapy. While laboratory tests were performed, underlying disorder was not revealed at that time.

At 21 years of age, she noticed an erythematous nodule with pain on her left shin. She had fever after one week and the lesion became enlarged (Fig. 1c). Cefcapene (300mg per day) administration was not effective for the condition. Histopathological findings showed lobular panniculitis with necrotizing vasculitis in the deep dermis (Fig 1d). Direct immunofluorescence microscopy showed deposits of IgG and C3 in the blood vessel walls of the dermis. Bacterial cultures from the lesion were negative. Laboratory findings showed increase of white blood cells $8.77 \times 10^3 \mu L$ ($3.04 \sim 8.72 \times 10^3 \mu L$), C-reactive protein $2.70 \, \text{mg/dl}$ ($0.00 \sim 0.10 \, \text{mg/dl}$), IgA 552 mg/dl ($110 \sim 410 \, \text{mg/dl}$) and IgG 2224 mg/dl ($110 \sim 410 \, \text{mg/dl}$). Anti nuclear antibodies, antineutrophil cytoplasmic antibodies or rheumatoid factor was negative. The cutaneous lesion was diagnosed erythema induratum based on the histopathological findings. This lesion was healed with pigmentation after 2 weeks of loxoprofen (180mg per day) administration.

Because she had suffered from pyoderma gangrenosum and erythema induratum, TA was suspicious. Physical examinations disclosed the diminished left radial pulse and carotid bruit. Blood pressure discrepancy of 26 mm Hg was recognized between the left and right arms. She complained dysesthesia of arms as well as episodic general fatigue and low-grade fever. The findings of CT scanning and MR angiography established the diagnosis of TA. Therapy with systemic corticosteroid, predonizolone (30mg per day), and aspirin led to the rapid resolution of her symptoms. Her dose of predonizolone was decreased after her symptoms improved. After one year, systemic corticosteroid was stopped because the C-reactive protein level was decreased to the normal range. No recurrence of general fatigue, low-grade fever or cutaneous lesions was observed to 2 years.

Cutaneous lesions accompany by 2.8% to 28% of TA patients.¹ We reviewed the Japanese cases of TA with cutaneous lesions and compared with western cases (Table 1).¹⁻³ In Japanese, pyoderma gangrenosum was the most prevalent cutaneous lesion associated with TA. Twenty to 30% of the patients with pyoderma gangrenosum had TA.⁴ By contrast, in western cases, the most prevalent cutaneous lesion associated with TA is clinically erythema nodosum-like and necrotizing vasculitis in histopathology. Although an etiology of relationship between cutaneous lesions and TA is unclear, racial difference might be one of the factors which cause the discrepancy of cutaneous lesions between Japanese and western cases of TA. To our knowledge, there were only two cases of TA including our case had both pyoderma gangrenosum and erythema induratum.¹

<u>In conclusion</u>, early treatment for TA may lead to a better prognosis.⁵ To know cutaneous manifestations of TA is essential for early diagnosis and therapy.

References

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Legends

Figure 1 (a) Pyoderma gangrenosum on the left lower leg at 19 years of age. A brownish macule, 10×7 cm in diameter, covered with necrotic crusts. (b) The pathological findings of Fig. 1 (a). Dense neutrophilic infiltration in the dermis, especially around the destroyed follicles. (hematoxylin & eosin, (b) \times 400). (c) Erythema nodosum-like lesion (arrow) at 21 years of age. The lesion was over the left shin with pain and heat. The scar was due to pyoderma gangrenosum. (d) The pathological findings of Fig. 1 (c). Lobular panniculitis with necrotizing vasculitis in the deep dermis. (hematoxylin & eosin, (d) \times 400).

Table 1 Comparison of TA cases with cutaneous lesions in Japanese and western literatures.

Histopathological findings	The number of cases	
	In Japanese literatures	In western literatures
Pyoderma gangrenosum	35<	4
Necrotizing vasculitis	1	11
Panniculitis	9	3
Granuloma	0	2
Granulomatous vasculitis	0	2

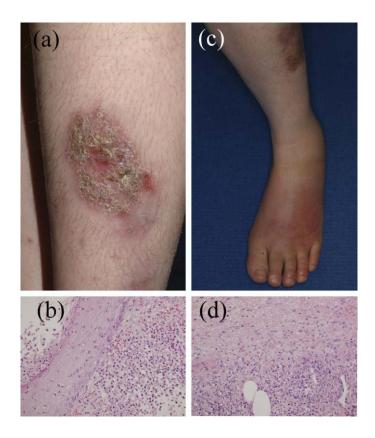


Figure 1