

Single Case

# Pyoderma Gangrenosum and Erythema Nodosum Revealing Takayasu's Arteritis

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## Keywords

Erythema nodosum · Pyoderma gangrenosum · Takayasu's arteritis · Vasculitis

## Abstract

We report a Caucasian female who presented with simultaneous erythema nodosum and pyoderma gangrenosum due to underlying Takayasu's arteritis. Takayasu's arteritis is a chronic large vessel vasculitis of unknown cause. The disease has a worldwide distribution but is most commonly seen in Asian populations. There is a strong predilection for young females. The clinical presentation is variable, but mostly derives from stenosis or occlusion of affected arteries, resulting in claudication and ischemia. Skin manifestations are observed in up to 28% of patients with Takayasu's arteritis, with erythema nodosum reported more frequently in Caucasians. Pyoderma gangrenosum is more common in Asian patients. This report demonstrates the importance to exclude Takayasu's arteritis in patients with such skin lesions.

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## Case Report

A 43-year-old Caucasian woman presented with a history of recurrent painful subcutaneous nodules at the lower extremities (Fig. 1a). Skin biopsy had revealed lymphocytic lobular panniculitis and led to the diagnosis of erythema nodosum. In addition, the patient had

recurrent ulcers of the calves in which biopsies had demonstrated neutrophilic infiltrates without vasculitis, leading to the diagnosis of pyoderma gangrenosum (Fig. 1b). Three months before referral to us, the patient had experienced a new episode of pyoderma gangrenosum, the C-reactive protein (CRP) was 72 mg/L and prednisone had been started (50 mg).

At admission, the patient had multiple ulcers at the calves but was in good general condition. There was no pulse at the left ulnar and radial arteries. The blood pressure was 168/102 mm Hg at the right upper arm, and 117/94 mm Hg at the left upper arm. Magnetic resonance angiography revealed a proximal occlusion of the left subclavian artery (Fig. 1c), and T1-weighted magnetic resonance images showed a circular contrast-enhanced thickening of the wall of the aortic arch and the subclavian artery (Fig. 1d). IgG4 was within normal limits and thrombophilia was excluded. According to the criteria of the American College of Rheumatology, the vasculitis in this woman was classified as Takayasu's arteritis [1]. Cyclosporine and methotrexate were added to prednisone. After 6 months of immunosuppressive combination therapy, the pyoderma gangrenosum lesions had vanished and the aortic inflammation had regressed.

## Discussion

Takayasu's arteritis is a chronic granulomatous large-vessel vasculitis affecting predominantly the aorta and its main branches. The majority of cases are female; the age of onset is mostly in the second and third decades. Takayasu's arteritis has a worldwide distribution but the greatest prevalence was observed in Asians [2]. The initial symptoms of Takayasu's arteritis result from systemic inflammation and consist mainly of constitutional symptoms, such as malaise, fever, and weight loss. In later stages, narrowing and occlusion of the arteries cause symptoms such as limb claudication, stroke, or congestive cardiac failure. Characteristic clinical findings consist of diminished or absent pulses, asymmetric blood pressures between right and left limbs, or vascular bruits. The systemic inflammation of the active vasculitis may be reflected by increased CRP levels or increased erythrocyte sedimentation rates, but there are no laboratory parameters that are specific for Takayasu's arteritis.

Cutaneous manifestations have been observed in up to 28% of patients with Takayasu's arteritis [3]. They include pyoderma gangrenosum (mainly in Asian patients), erythema nodosum (predominantly in Caucasian patients), as well as erythema induratum. Papulonecrotic eruptions, ulcerated nodular lesions, facial eruptions resembling systemic lupus erythematosus and Raynaud's phenomenon are observed more rarely [4]. On skin biopsy, granulomatous lesions and small vessel vasculitis may also be seen [5].

The pathogenesis of Takayasu's arteritis and its relationship to the inflammatory skin manifestations is not understood. Gamma/delta T lymphocytes have been observed to infiltrate the aortic wall [6]. It is interesting in this context that T cells appear to also play an integral role in pyoderma gangrenosum [7] and erythema nodosum [8].

About two-thirds of cases with Takayasu's arteritis and simultaneous skin lesions can be treated successfully with systemic corticosteroids alone, while one-third of cases were resistant to this therapy [9]. We chose cyclosporine A as an adjunctive treatment because this agent was demonstrated to be effective in pyoderma gangrenosum [10] and possibly also in Takayasu's arteritis [9]. We also added methotrexate as an immunosuppressant, as methotrexate was suggested to suppress the relapse rate of the large-vessel vasculitis and to reduce the cumulative corticosteroid requirements [11]. Methotrexate has also a good safety

profile when given in combination with cyclosporine A [12]. About 3 months after the initiation of immunosuppressive therapy, the cutaneous lesions had healed and the large vessel vasculitis regressed.

The association of Takayasu's arteritis with pyoderma gangrenosum has been described mainly in Japanese patients and only rarely in the Western literature [9]. To our knowledge, this is the first case featuring the combination of Takayasu's arteritis with both cutaneous manifestations. Our case also illustrates the need to actively search and exclude the presence of concomitant Takayasu's arteritis not only in Japanese, but also in Caucasian patients.

### Statement of Ethics

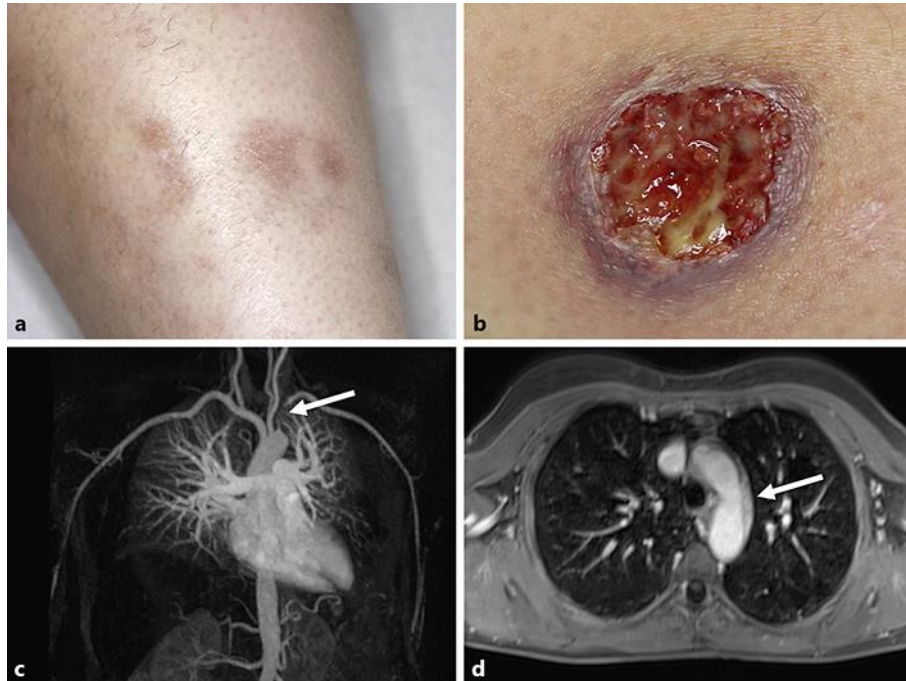
The authors have no ethical conflicts to disclose.

### Disclosure Statement

The authors have no conflicts of interest to disclose.

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**Fig. 1.** A 43-year-old Caucasian female with erythema nodosum as painful, erythematous, and subcutaneous nodules on both lower extremities (a), and pyoderma gangrenosum with undermined ulcers on the calves (b). Magnetic resonance angiography revealed a proximal occlusion of the left subclavian artery (c), and T1-weighted magnetic resonance images showed a contrast-enhanced thickening of the wall of the aortic arch (d). This large vessel vasculitis was classified as Takayasu's arteritis.