

Takayasu Arteritis with Varied-size Vessel Involvement

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Small vessel vasculitis is a rare comorbidity of Takayasu arteritis (TA). This type of systemic vasculitis can be life-threatening. We present here a case involving small, medium-sized, and large vessels, and presumably microvessels.

The patient, a 30-year-old woman, presented with saddle nose (Figure 1A), scleritis, and multiple skin ulcers in her lower legs.

The patient's acute-phase reactant levels were elevated

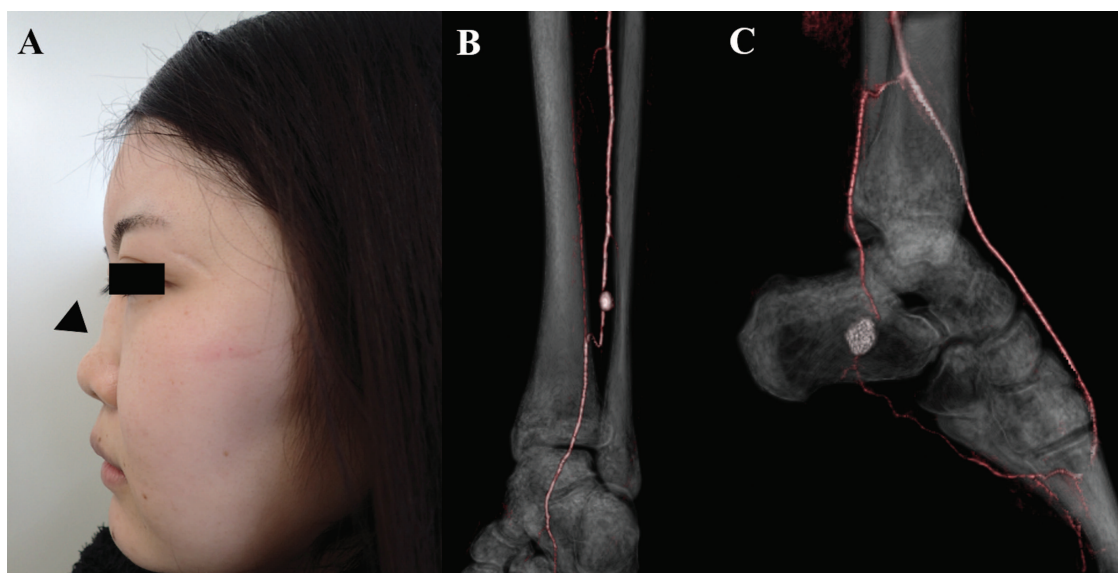


Figure 1. A. Saddle nose is observed (arrowhead). Computed tomography angiography reveals aneurysms in left posterior tibial artery (B) and right peroneal artery (C).

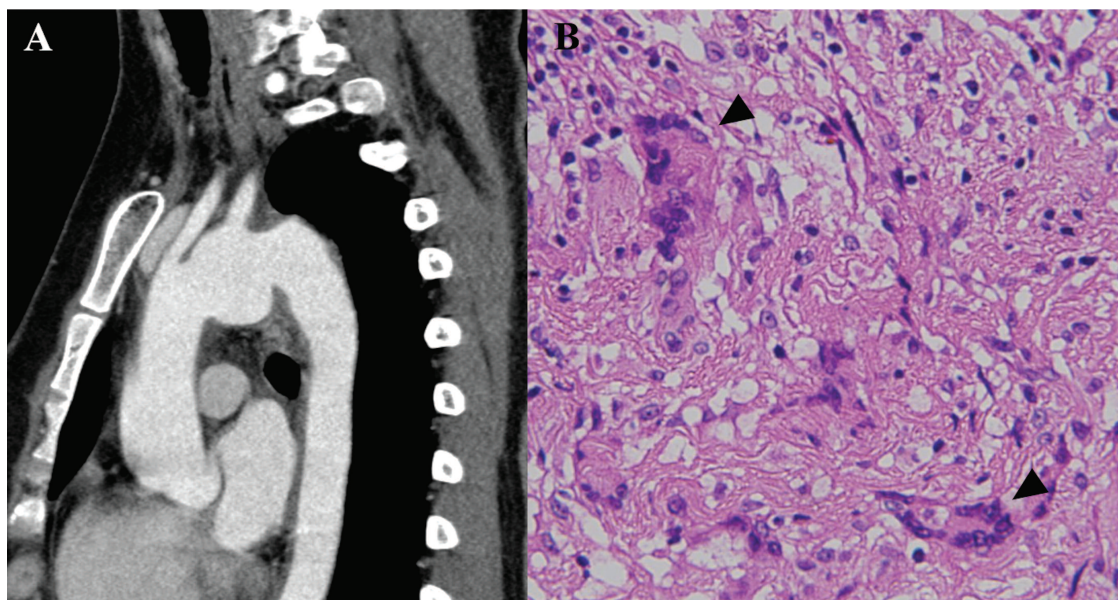


Figure 2. A. Computed tomography scan shows thoracic aortic aneurysm. B. Pathological findings of aortic aneurysm (arrowheads) reveal granulomatous inflammation with giant cells.

without autoantibodies or abnormal findings in contrast computed tomography (CT). Because small vessel vasculitis was strongly suspected, she was treated with immunosuppressants, resulting in clinical improvement.

One year later, she noticed a pulsatile mass in her left ankle. CT revealed aneurysms in the left posterior tibial artery (Figure 1B) and right peroneal artery (Figure 1C). Resection of the left aneurysm was done, but shortly after the surgery she presented with acute chest pain, and vascular bruit appeared on her chest and neck. A CT scan revealed a threatened rupture of a thoracic aortic aneurysm (Figure 2A). Emergency aortic root replacement was performed and pathological examination revealed granulomatous inflammation with giant cells (Figure 2B). This patient carried the HLA-B52 allele, which is frequently observed in cases of TA in the Japanese population¹. The patient was diagnosed with atypical TA involving not only large vessels but also small and medium-sized arteries, and presumably microvessels.

As a rare comorbidity of TA, several studies have shown small vessel vasculitis in the form of ocular lesions² or cutaneous manifestations³, which precede TA. This case at first developed clinical manifestations related to small vessel involvement and during the course, medium and large vessel vasculitis became apparent. Such a varied-size arteritis type of systemic vasculitis is quite rare and unique; however, it should be regarded as a progressive disease that can be fatal.

REFERENCES

1. Kimura A, Kitamura H, Date Y, Numano F. Comprehensive analysis of HLA genes in Takayasu arteritis in Japan. *Int J Cardiol Suppl* 1996;54 Suppl 1:61-9.
2. Noel N, Butel N, Le Hoang P, Koskas F, Costedoat-Chalumeau N, Wechsler B, et al. Small vessel involvement in Takayasu's arteritis. *Autoimmun Rev* 2013;12:355-62.
3. Rocha LK, Romitti R, Shinjo S, Neto ML, Carvalho J, Criado PR. Cutaneous manifestations and comorbidities in 60 cases of Takayasu arteritis. *J Rheumatol* 2013;40:734-8.