‘Lead pipe’-like stiff aorta with grossly widened pulse pressure in burned-out Takayasu arteritis

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A 67-year-old Japanese woman was referred for weakness of the left radial pulse and intermittent claudication in the left leg. The four extremities showed apparent discrepancies in the blood pressure (BP): 254/58, 130/54, 110/46, and 60/31 mmHg at the right brachium, left brachium, right ankle, and left ankle, respectively (Panel A). The central aortic pressure estimated from right radial pulse waveform was 218/57 mmHg with enhanced wave reflection (Panel B). Chest X-ray and computed tomography (CT) showed diffuse calcification in the aortic wall, which had become like a ‘lead pipe’ (Panels C–E). Vascular examinations revealed multiple stenoses with calcification in the left subclavian artery, middle cerebral artery, aorta, and left femoral artery (Panels F and G, MR; Panels H and I, CT) without renal artery stenosis, although the patient had no diabetes, obesity, smoking habit nor severe dyslipidaemia. The vascular lesions were considered to represent a burned-out phase of Takayasu arteritis, which is chronic vasculitis affecting large elastic arteries with the greatest prevalence in Asians. Because intensified BP-lowering therapy caused transient aphasia suggestive of cerebral ischaemia, the BP was maintained between 180–200/40–50 mmHg at the right brachium.

This case showed grossly widened pulse pressure (150–200 mmHg) without associated aortic regurgitation. Since the aortic elasticity is responsible for cushioning cardiac ejection and creating adequate diastolic perfusion, the loss of elasticity in the ‘lead pipe’ caused exceptionally lowered diastolic BP with elevated systolic BP. These findings represent an extreme example of the relationship between increased aortic stiffness and wide pulse pressure. In patients with stiff aorta, reduced diastolic perfusion is associated with ischaemia in vital organs even without obstructive vascular lesions. Thus, in such cases, BP control should be carefully performed with monitoring of the perfusion in the vital organs, especially those with stenotic lesions.

Conflict of interest: None declared.

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