A 16-year-old girl collapsed suddenly while on her way to school. When the rescue team arrived, they found her to be in asystole. Spontaneous circulation was restored after 14 minutes of cardiopulmonary resuscitation. A 12-lead electrocardiogram showed sinus tachycardia and diffuse repolarization abnormalities (Figure 1). An echocardiogram showed moderate aortic regurgitation with severely impaired left and right systolic function. Computed tomography showed increased thickness of the aortic wall extending from the sinotubular junction to the abdominal aorta (Figure 2). The arch branches, celiac trunk, and renal arteries were normal. The patient was transferred to the cardiac surgery department of our hospital. The chest radiograph showed a normal-size cardiac silhouette but with evidence of interstitial edema (Figure 3). Transesophageal echocardiography showed increased thickness of the ascending and descending aortic wall, with mild aortic regurgitation and severe depression of left and right systolic function. No intimal flap of aortic wall was evident. Six hours after the onset of symptoms, cardiac magnetic resonance imaging showed (1) severe impairment of left systolic function with left ventricular ejection fraction of 15% and akinesis of the anterolateral and apical segments (Movie I of the online-only Data Supplement) (2) moderate aortic regurgitation, (3) increased thickness of the ascending and descending aortic wall in gradient-echo steady-state free precession cine (Movie II of the online-only Data Supplement) in T2-weighted images and in delayed-enhancement images after gadolinium–diethylenetriaminepentaacetic acid contrast medium (Figure 4), (4) dilatation of the ascending aorta with extensive irregularities of the descending aortic wall in angiography (Figure 5 and Movie III of the online-only Data Supplement), (5) subendocardial delayed enhancement of the left ventricular anterolateral and apical segments (Figure 6), and (6) bilateral pulmonary edema in T2-weighted images after gadolinium–diethylenetriaminepentaacetic acid contrast medium (Figure 4C and 4D). A diagnosis of aortitis and acute myocardial infarction was made.

A significant rise of troponin (9.72 U/L), myoglobin (485 U/L), creatinine-kinase (6748 U/L), leukocyte count (14.2 10^9/L), and protein-C (15.8 mg/dL) was evident. The patient was given aspirin, angiotensin converting enzyme inhibitors, β-blockers, and prednisone at a 50-mg daily dose. After normalization of inflammation indexes, 2 weeks after admission, coronary angiography showed left main occlusion, ostial right coronary artery stenosis (Movie IV of the online-only Data Supplement), and the left anterior descending artery perfused by collaterals from the right coronary artery (Figure 7 and Movie V of the online-only Data Supplement). Echocardiography and cardiac magnetic resonance imaging showed improvement of left ventricular function (41%) and a large subendocardial infarction of the left anterior descending artery territory. Inasmuch as the transmural extent of the infarct was less than 50%, it was considered that substantial viability of the left anterior descending territory was present. Coronary artery bypass surgery was performed, with the right and left mammary arteries supplying the right coronary artery and the left anterior descending artery. The patient’s postsurgical course was uneventful. Histology and immunohistochemical analysis of biopsy specimens showed a vasculitis pattern of aorta characterized by intimal hyperplasia, mononuclear cell infiltration, smooth muscle cell proliferation and migration through the intima, elastic lamina destruction (Figure 8), and evidence of ascending aorta medial layer immunostaining for human leukocyte antigen–heterodimer, a marker of histiocyte activity (Figure 8D). The coronary arteries, subclavian arteries, and myocardial specimens showed normal patterns.

Takayasu arteritis is a rare disorder of young women, with an estimated annual incidence rate of 0.4 to 1 cases per million. The clinical presentation varies across a spectrum of symptoms and clinical signs, with the abdominal aorta considered the most common area of involvement, followed by the descending thoracic aorta and aortic arch. The cardiac pathological changes are directly related to aortic wall in-
flammation, with aortic regurgitation as a result of aortic root dilatation and coronary ostial stenosis resulting from aortitis.²,³ Specific diagnostic criteria for Takayasu arteritis have been established, including age < 40 years, claudication of extremities, decreased brachial artery pulse, blood pressure difference between arms > 10 mm Hg, bruit over subclavian arteries, and arteriogram abnormalities.⁴ The presence of 3 or more criteria has a sensitivity of 91% and a specificity of 98%.⁴ In this specific case, the most important clinical findings were aortic regurgitation as a result of aortic root dilatation and bilateral coronary ostial stenosis. Aortic arch branches were spared, as shown by the angiography and the selective arteriogram of the subclavian arteries and by biopsy specimens. Aortitis was evident in the abdominal aorta, with severe thickening of the aortic wall and lumen diameter reduction.

Appropriate imaging modalities are crucial to establish the diagnosis. Ultrasonography of the aortic arch branches permits accurate evaluation of the arterial wall. A typical finding is the circumferential thickening of the arterial wall of the carotid arteries—the “macaroni sign.”⁵ Echocardiography plays a key role in the assessment of the aortic root and aortic valve in the setting of aortitis of the ascending thoracic aorta associated with aortic insufficiency.⁶ In addition, transesophageal echocardiography allows evaluation of the thickness of the aortic wall and the exclusion of aortic pathological changes that may resemble acute aortitis, including aortic dissection.⁷ Computed tomography in the setting of acute aortitis permits accurate assessment of the aortic wall thickening and the rapid exclusion of aortic pathological conditions, including intramural hematoma and aortic dissection. Cardiac magnetic resonance imaging offers a comprehensive evaluation of aortitis with specific sequences (T2-weighted and gadolinium contrast enhancement) for vessel wall edema characterization⁸,⁹ and an accurate evaluation of cardiac function without radiation exposure or iodinated contrast media.

Figure 1. Electrocardiogram at time of admission, demonstrating sinus tachycardia and precordial ST elevation.

Figure 2. Computed tomography scan on admission, showing increased wall thickness of ascending and descending aorta, interpreted as intramural hematoma.
For the sake of safety, simplicity, and accessibility, in emergency settings ultrasound and computed tomography are considered first-choice diagnostic tools. In this specific experience, cardiac magnetic resonance imaging showed feasibility and safety for a patient in an extremely unstable clinical condition, with incremental diagnostic value over ultrasound and computed tomography.

Treatment of coronary ostial stenosis of Takayasu arteritis is still controversial. Coronary angioplasty and the implantation of drug-eluting stents are affected by a high rate of restenosis, whereas cardiac surgery can be hampered by aortic arch involvement excluding the mammary artery grafting and by ascending aorta aneurysm. In this specific case, the evidence of angina resulting in marked limitation of physical activity, the significant amount of viable myocardium as shown by cardiac magnetic resonance imaging, and the absence of involvement of the subclavian arteries oriented the treating physicians to the surgical option.

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Disclosures
None.

References
Figure 4. Oblique sagittal cardiac magnetic resonance images of thoracic aorta with gradient echo (A) and delayed enhancement (B). Transverse images of thoracic aorta with T2-weighted black blood showing increased wall thickness of ascending and descending aorta (white arrows) (C) and delayed enhancement of descending aorta (white arrows) (D), equivalent to the “macaroni sign,” the ultrasonographic evidence of circumferential thickening of the arterial wall of the carotid arteries. Notice bilateral lung edema.

Figure 5. Aortic angiography showing ascending aorta dilatation (A) and widespread restrictive pathological changes of descending aorta (B).
Figure 6. Cardiac magnetic resonance images showing subendocardial delayed enhancement of anterolateral segment of left ventricle. A, Four-chamber view. B, Medioventricular short axis view.

Figure 7. Coronary angiography showing ostial stenosis of right coronary artery with collaterals to left coronary artery (A) and left main ostial occlusion (B).
Figure 8. A, Massive intimal hyperplasia of aortic bulb (INT=intima, MED=media). B, Ascending aorta media layer with evidence of monocytic infiltrate (black arrows). C, Elastic fiber necrosis of ascending aorta media layer (white arrows) with smooth muscle fiber proliferation and collagen deposition. D, Ascending aorta media layer immunostaining for human leukocyte antigen-heterodimer (HLA-DR) (black arrows) showing activation of histiocytes.
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