Acute Heart Failure Due to Midaortic Occlusion as the Initial Manifestation of Takayasu Arteritis

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An afebrile 8-year-old Japanese girl was referred with hypotensive shock and respiratory distress. She presented with a cough of 1-week duration followed by lethargy, dyspnea, and anuria, with which she was admitted to a previous hospital. She was intubated and given intravenous catecholamines, which did not improve her symptoms. Then, she was transferred to our hospital. Physical examination revealed a right arm blood pressure of 128/64 mm Hg and a left arm blood pressure of 94/56 mm Hg while on 4.6 μg/kg per min of dopamine and dobutamine. However, no blood pressure was obtained in the lower limbs. The heart sounds were distant with no murmur audible. Coarse crackles were prominent in both lungs. Laboratory data showed the following values: creatine kinase of 1392 U/L, creatine kinase-MB of 31 U/L, aspartate aminotransferase of 268 U/L, lactate dehydrogenase of 1284 U/L, plasma renin activity of 0.64 μg/dL, prothrombin time international normalized ratio of 3.42, partial thromboplastin time of 53.6 s, D-dimer of 4.8 μg/mL, C-reactive protein of 0.64 mg/dL, and plasma renin activity of 54 ng/mL. An ECG showed sinus tachycardia with a biphasic P wave in V1 and flat T waves in II, III, aVF, and V6 (Figure 1). A transthoracic echocardiogram showed significantly decreased ventricular wall motion (left ventricular ejection fraction of 24%) and left ventricular hypertrophy (left ventricular mass index of 64 g/m²) without cardiac anomaly, valvular abnormalities, or coronary lesions. Enhanced computed tomography showed complete occlusion of the descending aorta from the distal part of the aortic arch to the supraceliac aorta (Figure 2). Stenosis was found in the left common carotid artery, left carotid bifurcation, proximal superior mesenteric artery, pararenal aorta, and bilateral renal arteries. Vascular wall thickening was observed in the distal part of the aortic arch and left common carotid artery (Figure 3). Dilatation was found in the bilateral internal mammary arteries, superior and inferior epigastric arteries, and anterior spinal artery, all of which were thought to serve as collateral arteries (Figure 2). A color Doppler ultrasonography showed retrograde blood flow in the femoral artery, external iliac artery, common iliac artery, and abdominal aorta. Based on these findings, the patient was diagnosed with Takayasu arteritis and transferred to another hospital for bypass grafting. A complete medical history did not reveal any early disease manifestation such as unexplained fever, night sweats, weight loss, fatigue, myalgia, or arthritis. However, a review of a previous chest radiograph showed dilatation of the descending aorta, which appeared 16 months before admission (Figure 4). The narrowing in the descending aorta, abdominal aorta, or both is called middle aortic syndrome, which is often caused by Takayasu arteritis or fibromuscular dysplasia. Middle aortic syndrome due to Takayasu arteritis can cause cardiac failure even in childhood.

Disclosures
None.

Reference
Figure 1. An ECG is shown. Note a biphasic P wave in V1 and flat T waves in II, III, aVF, and V6.

Figure 2. A 3D computed tomogram is shown. The frontal view is shown in A and the lateral view is shown in B. Note complete occlusion of the descending aorta, measuring 12 cm in length. The bilateral internal mammary arteries and superior and inferior epigastric arteries are dilated. A reconstructed parasagittal enhanced computed tomogram is shown in C. Complete occlusion of the descending aorta and stenosis of the pararenal aorta are shown.
Figure 3. Cervical ultrasonography shows the left common carotid artery and left internal jugular vein. Note stenosis and vascular wall thickening in the left common carotid artery.

Figure 4. Chest radiographs are shown 16 months (right) and 18 months (left) before admission. Note the irregularity in the lateral margin of the descending aorta (arrow).
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