Middle aortic syndrome, severe hypertension, and endovascular repair

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A 55-year-old woman presented to the emergency room with dyspnoea, epigastric pain, and dizziness. She was severely hypertensive. Physical examination revealed a systolic murmur in the left sternal edge (II/VI), bilateral basal rales, and absence of pulses on both lower limbs. The chest radiography supported the diagnosis of acute pulmonary oedema. The 12-lead electrocardiogram showed sinus tachycardia with non-specific changes of repolarization. Echocardiography showed severe concentric left ventricular hypertrophy and low ejection fraction. Thoraco-abdominal CT was performed showing a severe calcified stenosis in the transition from the thoracic aorta to the abdominal aorta. Aortography confirmed the presence of severe stenosis of descending aorta with a gradient of 160 mmHg. The patient underwent urgent percutaneous stent implantation with an acceptable luminal gain and a residual gradient of 30 mmHg (Panels C, D, and E). The patient’s haemodynamic condition improved during the next few hours following the endovascular procedure, and she was discharged asymptomatic 1 week later. Three months after hospital discharge, her blood pressure remains normal.

Middle aortic syndrome is a diffuse narrowing of the distal thoracic and abdominal aorta that commonly involves the visceral and renal arteries. This condition typically presents as severe hypertension in young patients who have weak or absent femoral pulses and an abdominal or lower back bruise. The aetiology of the middle aortic syndrome is controversial. The diagnosis of this rare syndrome is made by computed tomography, magnetic resonance imaging, or angiography. The natural history of this rare syndrome is unknown. Most authors agree that these patients should undergo revascularization whenever feasible. Aorto-aortic bypass, patch aortoplasty, and percutaneous techniques are the most frequent approaches used for the treatment of patients with middle aortic syndrome.

Panel A. Non-contrast computer tomography shows a severe calcified stenosis in the transition from the thoracic aorta to the abdominal aorta.

Panel B. The same lesion at a coronal view.

Panel C. The sequence of percutaneous intervention (pre- and post-intervention).

Panels D and E. Multidetector computer tomography showing the implanted stent at axial and sagittal views.

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