A 61-year-old male with a past medical history of chronic, uncontrolled hypertension received a non-contrasted computed tomogram (CT) of the chest and abdomen to investigate for possible Conn syndrome. This noncontrast study showed some areas of nodularity around the vertebral bodies bilaterally and extending into the posterior mediastinal region. A CT of the chest with intravenous contrast, and 3D reconstruction were then obtained.

**What is your diagnosis?**

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RADIOLOGIC DIAGNOSIS: Aortic coarctation. There is narrowing in the region of the aortic isthmus (arrows, Figures 2 and 4). The extent of collateral arterial dilation (arrows, Figures 1 and 3) suggests this to be a hemodynamically significant finding.

INTERPRETATION OF IMAGES

The noted abnormalities on the CT scan are shown to represent dilated vessels (arrows in Figure 3). The ascending aorta measures 4.3 cm x 4.3 cm in size but narrows to 1.5 cm (arrows in Figures 1, 2, and 4). Just beyond the narrowing it measures 2.1 cm x 2.1 cm. The previously noted paravertebral soft tissue densities represent dilated intercostal arteries. There are collaterals seen in the upper posterior mediastinum in the paraspinal region, and one of the collateral vessels is posterior to the esophagus. Additional internal mammary artery collaterals are seen.

DISCUSSION

Coarctation of the aorta (coarctation) is most commonly a congenital abnormality that involves stenosis of the proximal descending thoracic aorta. The majority of these abnormalities are congenitally acquired; however, there have been case reports of severe atherosclerosis, umbilical artery catheterization, or surgical aortotomy causing the stenosis. This process was first described in 1760 and accounts for about 7% of all congenital heart diseases. It occurs twice as frequently in men and should always be ruled out if other conditions like bicuspid aortic valve, Turner syndrome, Shone syndrome, ventricular septal defect, or aneurysms of the circle of Willis are present because of the high association of these defects with coarctation. Up to 40% of patients with bicuspid aortic valve will also have coarctation of the aorta. Diagnosis of the disease is critical, as the mortality rate is high in poorly managed patients. For unoperated cases, the average survival is approximately 35 years with a 75% mortality by age 46.

The pathologic changes that lead to this disease are rooted in the wall of the aorta. These changes become so severe that the lumen of the vessel narrows, resulting in poor perfusion of systemic organs. The pathophysiologic changes in the congenital cases involve medial thickening and hyperplasia of the intima. The etiology of this thickening is believed to result from fetal aortic arch underdevelopment and constricted ducial tissue. If antegrade intrauterine blood flow is reduced and ducial tissue extends into the thoracic aorta, then the fetal aortic arch will not develop fully, and this excess tissue will cause constriction just distal to the aortic valve. For clinicians, the end result is a few tell-tale physical examination findings. The major manifestation is a difference in systolic blood pressure between the upper and lower extremities (usually upper extremity hypertension) with little difference in the diastolic pressures. In addition to this, renal hypoperfusion will increase renin secretion, resulting in volume expansion and further hypertension. Other more specific clinical findings include a delayed femoral compared to brachial pulse or greatly diminished blood pressure in the lower extremities.

With the advancements in imaging techniques in recent years, radiologic assessments have become the predominant tool with which to diagnose an aortic coarctation. A chest radiograph can aid in the diagnosis, as the engorged collateral vessels will show notching of posterior ribs. This vessel erosion is most commonly seen in the third to eighth ribs. Echocardiography is also used as a noninvasive technique to better visualize suspected coarctation. With the addition of Doppler, transthoracic echocardiography can allow for full identification of the severity of the disease, as well as any associated cardiac defects. The gold standard, however, for diagnosing aortic coarctation using imaging is with either magnetic resonance imaging (MRI) or computed tomography (CT). As with our patient, these modalities allow for clear visualization of the severity of the coarctation, as well as any collateral vessels that have developed. From our experience, a CT with intravenous contrast should always be performed if this abnormality is suspected.

Treatment of this anomaly depends largely on age at presentation, severity of illness, and degree of collaterals surrounding the area. Neonates and infants generally get surgical correction of the anatomy while adult treatment includes the option of percutaneous stenting. Although less than 1% of patients have surgical complications, these complications can be devastating and include nerve paralysis, spinal cord ischemia, or bowel infarction. Currently, there is not enough evidence to recommend stenting versus surgical correction as the optimal treatment for coarctation in adults. Stenting appears to have better short-term improvement, while the long-term affects have not been studied well enough to make a recommendation. Monitoring of the patient, regardless of intervention, is similar. At least one MRI or CT of the head should be performed at or near diagnosis to exclude intracranial aneurysm formation. In addition to this, all aortic coarctation patients need close monitoring and follow-up by a cardiologist at yearly intervals. These visits should include evaluation of blood pressure in all extremities, as well as aggressive treatment if hypertension is found. Imaging (MRI or CT) of the chest should be performed at five-year intervals with the purpose of catching progression of the disease before a catastrophic event occurs.

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