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Aortic coarctation in a 49-year-old female

Although rarely diagnosed in adults, narrowing of the thoracic artery should be considered when patients present with difficult-to-control hypertension.

ABSTRACT: Aortic coarctation is characterized by localized narrowing of the thoracic aorta. While it is frequently diagnosed in infancy and childhood, first-time presentations in adults are rare. Unless significant hypertension exists, adult patients are usually asymptomatic. When aortic coarctation is not detected and repaired, survival averages 35 years, with mortality increasing to 75% by age 46. In a recent case, a 49-year-old female with cardiovascular risk factors presented with sudden onset dyspnea and was admitted to hospital for management of heart failure. Further investigation revealed a coarctation distal to the left subclavian artery. The patient subsequently underwent uncomplicated endovascular repair of her coarctation. Although uncommon in adults, aortic coarctation should be included in the differential diagnosis for difficult-to-control hypertension, since it is associated with significant mortality when untreated. Authors of a recent Cochrane review say more evidence is needed to determine the best treatment for coarctation of the thoracic aorta.

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Aortic coarctation (AC) is characterized by localized narrowing of the thoracic aorta. While acquired cases have been reported, most cases are congenital and occur in about 4 of 10 000 live births.[1-3] Diagnosis is most often made during infancy and childhood, and only rarely in adulthood. The coarctation lesion typically occurs distal to the left subclavian artery but can also be proximal to this artery, be within the abdominal aorta (rare), or be present as extended tubular hypoplasia. The pathophysiology of AC is largely unknown. Two theories exist and relate to (a) reduced intrauterine blood flow leading to underdevelopment of the aortic arch[4] and (b) migration of ductal tissue into the wall of the aortic arch during fetal development.[5-7]

Case data

A 49-year-old female presented to hospital with sudden onset dyspnea. Her medical history included hypertension, for which she had been seen by an internist and told there was a significant "white coat" component. She also had type 2 diabetes and benign supraventricular tachycardia. She had a 40-pack-a-year history of tobacco use. She was taking metoprolol (150 mg twice daily) and hydrochlorothiazide (12.5 mg daily).

Physical examination of the patient revealed significantly elevated blood pressure at 196/100 mm Hg bilaterally with a heart rate of 100 beats per minute. Oxygen saturation was 94% on 2 L of oxygen per minute via nasal cannula. The jugular venous pressure was elevated at 5 cm. There was a grade 2/6 mid-peaking systolic murmur appreciated over the left sternal border without radiation. Femoral pulses were present but weak. There was no significant lower limb edema. Diffuse rales were heard upon lung auscultation.

A chest X-ray revealed vascular redistribution in keeping with pulmonary edema, and an electrocardiogram showed a pre-existing left bundle branch block. She had a small increase in troponin. Based on these findings, the patient was admitted in to hospital for further investigation and management of new onset heart failure.

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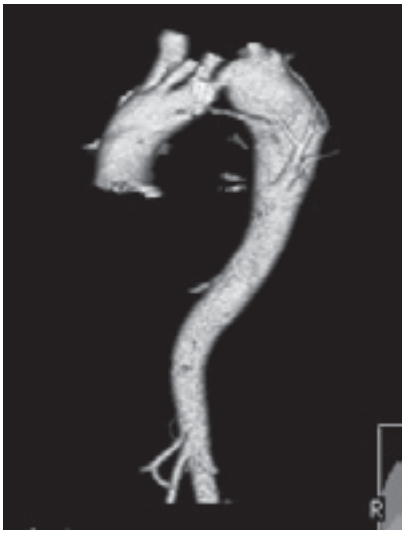

Figure 1. A reconstructed CT aortogram shows localized narrowing of the aorta immediately distal to the origin of the left subclavian artery. Postcoarctation dilatation can also be seen.

Investigation results

Transthoracic echocardiography showed diminished contractility of the septum. No vegetations, intraluminal masses, thrombi, or pericardial fluid were noted.

Right heart catheterization revealed moderate pulmonary hypertension, while left heart catheterization revealed a ventricular pressure of 200/30 mm Hg. Left ventricular function was minimally diminished and there was significant hypertrophy. No aortic valvular gradient was demonstrated. No significant coronary artery disease was noted. A significant aortic pressure drop was measured from the ascending to the descending aorta, and a supraventricular aortogram revealed a coarctation distal to the left subclavian artery with postductal dilatation. The aortic pressure was 200/90 mm Hg before the AC lesion and 130/90 mm Hg after.

The narrowest point of the AC measured approximately 7 mm in diameter (**Figure 1**). The distal descending thoracic aorta was unremarkable, and reasonable collateral circulation was supplied by the left subclavian artery branches.

Treatment

The patient was discharged from hospital following diuresis and optimization of blood pressure management. She was referred to a centre specializing in percutaneous coarctation repair and subsequently underwent uncomplicated endovascular repair and stenting (**Figure 2**).

Discussion

Classically, aortic coarctation manifests as systolic blood pressure differences between the upper and lower extremities.[8] Hypertension is noted in the upper extremities, while the lower extremities exhibit lower or unobtainable blood pressures as well as delayed pulses (brachial-femoral delay).[8] For cases where the origin of the subclavian artery is distal to the coarctation, the left brachial pulse is diminished and equal to the femoral pulse. In the 3% to 4% of cases where the right and left subclavian arteries originate below the area of coarctation, blood pressures and pulses are symmetrically decreased in all four extremities.[8]

Natural history

When aortic coarctation is not detected and repaired, survival averages 35 years, with mortality increasing to 75% by age 46.[9] In addition to systemic hypertension, common complications of untreated AC include expedited coronary artery disease, stroke, aortic dissection, and heart failure. Aortic stenosis or regurgitation can occur in patients with associated bicuspid aortic valve. Cause of death in untreated AC is variable, but heart failure, aortic rupture, aortic dissection, endocarditis, endarteritis, intracranial hemorrhage, and MI have all been documented.[9,10]

Diagnosis

Undiagnosed adults are commonly hypertensive but otherwise asymptomatic, unless significant hypertension exists, in which case, headache,

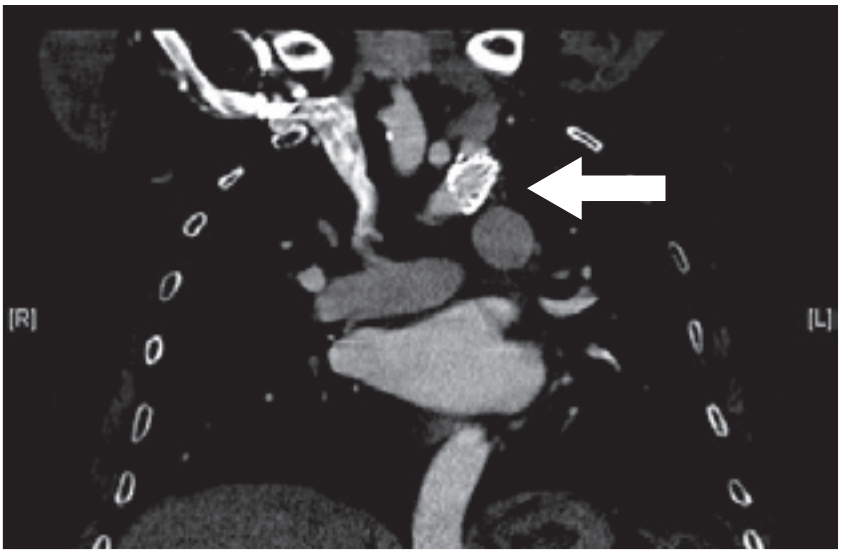

Figure 2. A postoperative CT scan with contrast shows a portion of the percutaneously inserted stent within the aortic arch (arrow).

epistaxis, heart failure, aortic dissection, or lower extremity claudication can be present. Physical examination may show continuous murmurs originating from collateral vessels or systolic murmurs from coexisting cardiac defects such as patent ductus arteriosus, ventricular septal defect, or aortic stenosis.[8]

When aortic coarctation is not detected and repaired, survival averages 35 years, with mortality increasing to 75% by age 46.

Findings from an electrocardiogram tend to vary with age and severity. In adults the tracing may be normal or show left ventricular hypertrophy with increased voltage and ST and T wave changes in the left precordial leads.[11] Chest X-rays in adults can show notching of the posterior one-third of the third to eighth ribs secondary to erosion by large collateral arteries,[8] and indentation of the aortic wall at the site of coarctation with pre- and postcoarctation dilatation producing a figure 3 sign.[11]

Transthoracic echocardiography, including suprasternal notch views, can allow for hemodynamic evaluation in suspected cases of AC.[12] Doppler echocardiography can establish the diagnosis, characterize the severity of coarctation, and detect associated cardiac defects.[13,14]

Current guidelines recommend that every patient with AC, repaired or not, should have at least one cardiovascular MRI or CT scan to determine the location and severity of coarctation as well as to evaluate collateral flow.[12] Cardiac catheterization is indicated when coronary artery disease is suspected and surgery is planned.[12]

Management

Adults with aortic coarctation should undergo intervention in the following circumstances: (a) when the gradient across the coarctation is greater than or equal to 20 mm Hg or (b) the peak-to-peak coarctation gradient is less than 20 mm Hg but anatomic imaging shows significant coarctation as well as marked collateral flow.[12] While correction of coarctation can sometimes be delayed in children, in adults correction should be performed at the time of diagnosis.

Because aortic coarctation is uncommon in adults, data comparing treatment options in this population are limited. Forbes and colleagues[15] conducted a multicentre observational study comparing surgery, stent, and balloon angioplasty for the treatment of native coarctation of the aorta. The authors found that stent patients had a lower rate of periprocedural complications than surgery patients. Surgery was associated with severe hypertension and atrial fibrillation. Balloon angioplasty was associated with aortic wall injury. At short-term and intermediate-term follow-up, stent and surgery patients had better hemodynamic outcomes than their counterparts who received balloon angioplasty. As well as having lower blood pressure gradients between upper and lower extremities, a large proportion of stent and surgery patients had normal systolic blood pressures postoperatively.

Stent placement after balloon angioplasty has been reported to reduce complications associated with angioplasty or surgery, minimize residual gradient, act to improve luminal diameter, and sustain hemodynamic benefit.[9,10,15-17] The use of covered stents has been associated with lower risk of aneurysm.[18-20]

In patients with suboptimal anatomy such as vessel tortuosity and transverse arch hypoplasia, stenting may be undesirable and the choice of procedure must be made on an individual basis by the clinical team.[21]

Complications following surgery or balloon angioplasty include recoarctation, aortic aneurysm, aortic dissection, and systemic hypertension.[22] Patients should be followed closely and regularly after whatever procedure is performed.

A recent Cochrane review performed by Padua and colleagues[23] evaluated several studies comparing endovascular stenting and surgical repair in the treatment of aortic coarctation. They concluded that there is insufficient evidence to determine the best treatment for coarctation of the thoracic aorta and emphasized the need for a randomized controlled clinical trial with high-quality samples and long-term follow-up.

Summary

Although uncommon in adults, aortic coarctation can have significant mortality and should be included in the differential diagnosis for difficult-to-control hypertension.

When a 49-year-old female presented to hospital with sudden onset dyspnea, examination revealed significantly elevated blood pressure, and she was admitted to hospital for further investigations. Subsequently, a CT aortogram revealed a coarctation lesion with involvement of the thoracic aorta.

After diuresis and optimization of blood pressure management, the patient was discharged and referred to a centre specializing in percutaneous coarctation repair. The patient later underwent angioplasty and stenting.

While the aortic coarctation was repaired successfully in this case, more evidence is still needed to determine the best treatment for coarctation of the thoracic aorta.

Competing interests

None declared.

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