Headaches and back pain

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A 39-year-old woman consulted her physician because of headaches and back pain. Physical examination revealed a systolic ejection murmur and decreased pulses in both legs. She

had a history of spontaneous subarachnoid hemorrhage secondary to a ruptured cerebral aneurysm. Radiographic and imaging studies are shown below (*Figures 1–4*).

Figure 1. Posteroanterior radiograph of the chest shows a localized indentation in the proximal portion of the descending thoracic aorta (arrow).

Figure 2. Detail view of the ribs demonstrates inferior notching of the middle and lateral parts of the posterior portions of multiple ribs (arrows).

Figure 3. Gradient echo image of the aorta in the sagittal plane shows a stenotic area at the aortic isthmus (arrow).

Figure 4. Similar gradient echo image of the aorta demonstrates enlarged internal mammary arteries (arrows) and bridging mediastinal vessels (arrowhead).

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DIAGNOSIS: Coarctation of the aorta.

DISCUSSION

Coarctation of the aorta refers to an area of narrowing or reduction in the aortic lumen that may be present anywhere in the thoracic or abdominal aorta. This condition was first described in Paris, France, in 1791 and was first demonstrated angiographically in 1941 (1). Coarctation occurs because of a primary defect in the media of the aortic wall, which causes an irregular stenosis in the aortic lumen. This phenomenon is most commonly found distal to the left subclavian artery near the ductus arteriosus or ligamentum arteriosum (*Figure 3*) (2). Coarctation of the aorta is more prevalent in males than females (4:1) and is extremely unusual in African Americans (3). This entity is usually divided into 2 subgroups: localized coarctation, also known as adult/postductal/juxtaductal type, and tubular hypoplasia, also known as infantile/preductal/diffuse type.

Localized coarctation of the aorta typically describes a short, discrete narrowing of the aortic isthmus close to the ligamentum arteriosum distal to a closed ductus arteriosus. Patients with this condition are adolescents or adults and are typically asymptomatic; coexistent cardiac anomalies, which are common in the tubular hypoplasia subgroup, are usually absent. Therefore, the diagnosis is often missed until a routine chest radiograph is performed. Patients with localized coarctation, such as this patient, have uniformly typical findings on chest radiograph.

The "figure 3" sign describes the abnormal appearance of the aortic knob and proximal descending aorta (2). Because of the focal narrowing of the thoracic aorta distal to the left subclavian artery and the pre- and poststenotic dilatation of the aortic knob and aortic isthmus, the left-sided mediastinal border on the posteroanterior chest radiograph can resemble the number 3 to an experienced observer (*Figure 1*). In addition, an enlarged left subclavian artery can sometimes be visualized extending from the mediastinum to the left lung apex (3).

In addition to the "figure 3" sign, rib notching (*Figure 2*) is a commonly identified abnormality in coarctation on a frontal chest film. Because of the focal aortic stenosis, numerous arterial collateral pathways route blood flow from the proximal to the distal descending aorta. These collateral pathways include the internal mammary, anterior spinal, lateral thoracic, and intercostal arteries. The intercostal arteries travel with the accompanying intercostal veins and nerves in the neurovascular grooves along the inferior border of the posterior ribs. As the intercostal arteries enlarge secondary to hypertension, they erode the inferior border of the third through eighth ribs, causing a notched appearance (*Figure 2*) (1). While this appearance is not limited to aortic coarctation, the finding of bilateral, symmetric rib notching in a patient >6 years of age, especially in association with an abnormal aortic contour as described above, is virtually diagnostic of coarctation of the aorta.

When coarctation of the aorta is radiographically suspected, especially in a previously undiagnosed patient, further evaluation with vascular imaging is strongly recommended. Traditionally, diagnostic angiography or cineangiography has been used. Today, magnetic resonance imaging with contrast-enhanced magnetic resonance angiography (MRA) is an accurate yet noninvasive diagnostic method. MRA with parasagittal imaging

in the left anterior oblique projection using cardiac gating can exquisitely demonstrate the appearance of the aortic coarctation, the nature and degree of vessel involvement, and the type and course of collateral pathways, as well as adequately provide pre- and postsurgical evaluation and comparison in coarctation patients.

While patients with the localized type of aortic coarctation are frequently asymptomatic, their anomalous vascular anatomy does result in aberrant physiology and abnormal clinical findings. These patients can complain of headaches (from hypertension) and are at increased risk of intracranial circle of Willis berry aneurysms from altered intracerebral vascular flow (3). These patients, therefore, are at increased risk of aneurysmal rupture and subsequent death. In addition, patients can describe claudication symptoms, especially while exercising, secondary to lower extremity hypoperfusion. Careful physical examination will typically elicit a noticeable difference in blood pressure between the bilateral upper extremities (above the stenosis) and lower extremities (below the stenosis) as well as a delayed lower extremity or groin pulse (3). Cardiac evaluation will usually demonstrate a midsystolic murmur, and electrocardiography or cardiac imaging will demonstrate left ventricular hypertrophy (*Figure 1*). Both of these findings are caused by the aortic luminal stenosis and resultant increased workload of the left ventricle.

The second subgroup in coarctation of the aorta, tubular hypoplasia, describes a long, fusiform narrowing of the aortic arch somewhere distal to the origin of the innominate artery. This type of aortic coarctation involves neonates and infants, and cardiac anomalies, such as a bicuspid aortic valve, ventricular septal defect, and patent ductus arteriosus, commonly coexist (1). Coarctation of the aorta is the second most common cause of congestive heart failure in a neonate (following hypoplastic left heart), and these neonates also present with lower extremity cyanosis, pulmonary venous hypertension, and left ventricular failure. Interpretation of the chest radiograph in this patient group can be difficult, as the abnormal mediastinal contour seen in adult patients is obscured by the overlying thymus, and rib notching is inapparent at this young age (1). Instead, the chest radiograph will reveal cardiomegaly and increased pulmonary vascularity, consistent with heart failure.

Treatment for patients with coarctation of the aorta involves intraoperative repair of the stenosis, either by resection and end-to-end anastomosis or by use of a patch or graft system to circumvent the affected segment (3). In addition, medical management of systemic hypertension, both before and after surgical repair, is strongly advocated. Postoperative recoarctation can occur, and this complication can be managed with percutaneous transluminal angioplasty and/or stent placement at the stenotic site.

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