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CASE REPORTS

Conditions Mimicking Coarctation of the Aorta

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Abstract Coarctation of the aorta is a cause of right arm hypertension in children and of heart failure in infants after ductal closure. We present two cases with these presentations that were initially thought to be coarctation of the aorta. They were subsequently diagnosed as Takayasu's arteritis in the older child and a large cerebral arteriovenous malformation in the infant. These conditions should be in the differential of right arm hypertension and of aortic flow reversal on echocardiography.

Keywords Takayasu's arteritis · Cerebral arteriovenous malformation

Introduction

Coarctation of the aorta is a treatable cause of hypertension in the pediatric population. The clinical presentation is that of increased blood pressure in the right arm with a decrease in the pressure and pulses of the other extremities and/or a heart murmur. In infants with a large patent ductus arteriosus (PDA), these findings may not occur until closure of the PDA. Rarely, other conditions that mimic typical coarctation of the aorta occur. Two cases are presented that were initially misdiagnosed as coarctation of the aorta.

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Case Reports

Case 1

An 11-year-old Caucasian girl presented with intermittent leg pain of 1-month duration. The pain, cramping in nature, started after walking a half a block. This was associated with a tingling sensation of the feet. There was a history of transient blurring of vision associated with dizziness, headache, and epigastric pain. Recently, her mother, who was in nursing school, noted that the patient's blood pressure was higher in the right arm than in the left arm.

Seven months earlier, the patient was diagnosed with systemic-onset juvenile rheumatoid arthritis after several weeks of fever, weight loss, rash, pericardial effusion, and fatigue. She was treated with prednisone, methotrexate, folic acid, and naprosyn.

On examination, she was afebrile with a heart rate of 125 beats per minute. Her respiratory rate was 20 breaths per minute and blood pressures (mmHg) were as follows: right arm, 147/61; left arm, 82/61; left leg, 66/38; and right leg, 72/48. She had bruits over both carotid arteries with a decreased pulse on the left side of the neck. She had a grade II/VI systolic murmur over the left upper sternal border. The right radial artery was well felt but the left radial, femoral, and dorsalis pedis pulses were barely palpable. An abdominal bruit was noted at the mid-epigastrium. The remainder of the examination was normal. These findings suggested the presence of a coarctation of the aorta.

Laboratory evaluation showed a hemoglobin of 10.4g/dl. Her C-reactive protein was 85 mg/L (normal, <9 mg/L) and the erythrocyte sedimentation rate was 79 mm/hr (normal, < 14 mm/hr).

An electrocardiogram showed sinus tachycardia. An echocardiogram showed normal ventricular function, nor-

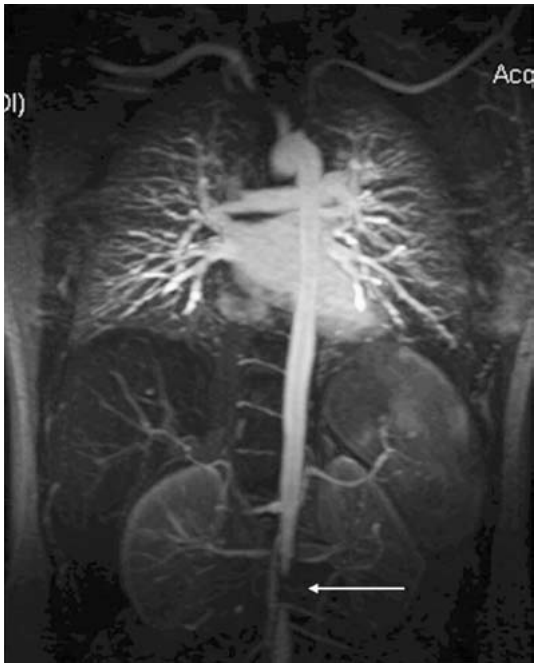

Fig. 1 Chest MRA. Anterioposterior view showing infrarenal abdominal aorta narrowing that measures 3 cm in length (*arrow*)

mal proximal coronary arteries, and no evidence of coarctation of the thoracic aorta. A sonographic evaluation showed circumferential wall thickening throughout the course of the left common carotid artery (LCCA) and a narrowed left internal carotid artery. A systolic pressure gradient was documented between the right and left brachial arteries of 76 mmHg. Magnetic resonance angiography (MRA) revealed normal caliber of the aortic arch and proximal descending aorta without evidence of thoracic coarctation. There was an infrarenal fusiform 60–70% narrowing of the abdominal aorta, measuring 3 cm in length (Fig. 1). It also showed a narrowing at the origin of the LCCA, the proximal portion of the left subclavian artery, and the origin of the right CCA. The right subclavian artery appeared normal. Angiography was performed to better delineate the cranial vasculature, and it revealed severe narrowing of the LCCA, left subclavian artery, the vertebrobasilar system bilaterally, the origins of the celiac, superior mesenteric artery, and both renal arteries with a severe infrarenal aortic stenosis (Fig. 2).

The patient was admitted to the hospital, where she received intravenous followed by oral steroids. The patient improved and was discharged home on day 9 of admission on prednisone, amlodipine, hydralazine, baby aspirin, etanercept (tumor necrosis factor inhibitor), and cyclophosphamide treatment.

On follow-up 3 months later, the patient reported being able to walk three blocks without pain. Her resting heart rate was still increased at 135 per minute with a blood pressure of 135/64 in the right arm. Her peripheral pulses were palpable in the right arm and right side of neck but were still reduced in the rest of her extremities. No bruits were heard. Her laboratory evaluation showed normalization of her acute phase reactants.

Case 2

A newborn infant was transferred to our hospital with a suspected diagnosis of interrupted aortic arch. The infant was born full term. Apgar scores were 9 at both 1 and 5 minutes. Delivery was complicated by pregnancy-induced hypertension in the mother. No abnormalities were detected on prenatal ultrasound examinations.

The infant developed cyanosis soon after birth and was noted to have low blood pressures. An echocardiogram suggested an interrupted aortic arch. The child was started on a prostaglandin (PGE_1) infusion at 0.05 μg/kg/min and was transferred to our institute for further evaluation. Physical examination on arrival revealed normal blood pressures in all extremities and good peripheral perfusion. All peripheral pulses were well felt. The precordium and the neck vessels were found to be bounding. No bruits were heard over the head, neck, or abdomen, and a head ultrasound was normal.

A repeat echocardiogram revealed normal intracardiac anatomy. The aortic arch was hypoplastic at the isthmus and had a tortuous appearance. A large PDA was present. Doppler echocardiography demonstrated retrograde blood flow through the transverse aortic arch (Fig. 3). Cardiac catheterization was performed on day 3 of life. Surprisingly, the right heart catheterization revealed high oxygen saturations of 96% in the superior vena cava (SVC) with the child breathing room air. Angiography in the ascending aorta filled only the arch vessels, with no contrast flowing into the descending aorta. This was followed by quick return of the contrast to the SVC. Injection in the descending aorta confirmed retrograde flow into the transverse arch. All these factors raised the suspicion of an arteriovenous malformation (AVM) in the head.

An MRA of the head and neck was obtained on day 5, revealing a dural-based AVM in the region of the torcula and posterior saggital sinus, with predominant blood supply from the external carotid arteries. Embolization of the feeder vessels was attempted; however, the external carotid artery was found to have a very tortuous course, which precluded this. Therefore, surgical ligation of both external carotid arteries was performed.

A repeat echocardiogram performed postoperatively revealed antegrade flow through the aortic arch with some residual shunting to the cerebral blood vessels. After the PGE_1 was stopped, the infant continued to have a stable clinical course, with good peripheral perfusion and femoral pulsations and no difference between the upper and lower

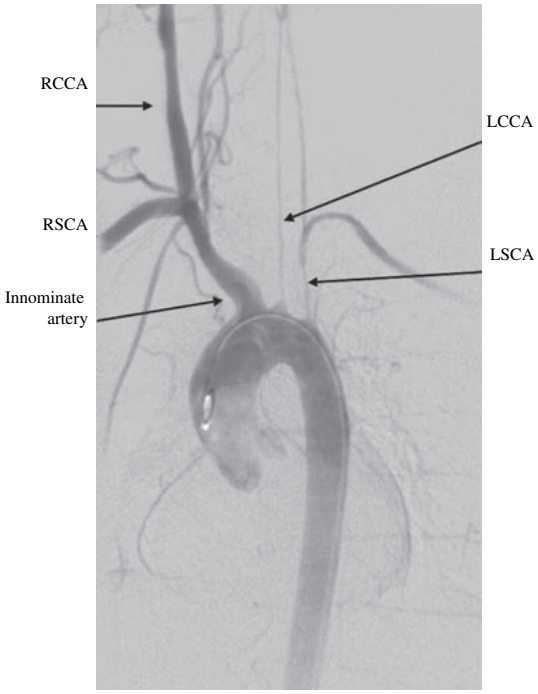

Fig. 2 Arch aortography reveals a patent aortic arch. Severe diffuse narrowing of the left common carotid and left subclavian arteries is shown. *LCCA*, left common carotid artery; *LSCA*, left subclavian artery; *RCCA*, right common carotid artery; *RSCA*, right subclavian artery

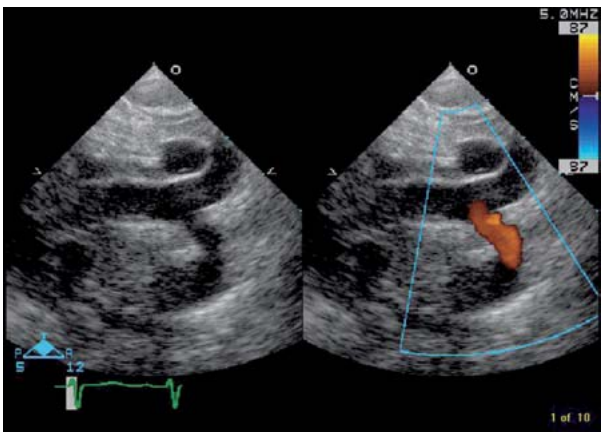

Fig. 3 Suprasternal echocardiographic image of the aortic arch reveals mild hypoplasia of the transverse aortic arch with flow reversal in the aortic arch

extremity blood pressures. She developed a new finding of ST segment depression and T wave inversion predominantly in the precordial chest leads. The etiology of these ECG changes was unclear. Repeat cardiac catheterization performed on day 20 of life demonstrated a pressure gradient of 10 mmHg from the transverse arch to the descending aorta. An angiogram revealed a tortuous arch with mild tapering toward the descending aorta.

The ECG changes normalized within a few days and the infant was discharged from the hospital on day 27 of life. There were no neurological deficits.

Discussion

We describe two cases that initially suggested coarctation of the aorta, one through the physical findings and one through echocardiographic findings. The first patient presented with hypertension of the right arm and a decrease in the pressure and pulses of the other extremities. Imaging studies revealed multiple sites of narrowing at the origin of the left subclavian artery, the abdominal aorta just below the renal arteries, as well as the renal arteries, thus explaining the findings of isolated right arm hypertension. The constellation of these findings pointed to the diagnosis of Takayasu's arteritis.

Takayasu's arteritis, also known as the pulseless disease, is a form of vasculitis that involves the large blood vessels. It typically affects the aortic arch and its major branches. It affects women more than men, and the onset is usually between 10 and 40 years of age [4].

The diagnosis is based on a diagnostic criteria set by the American College of Rheumatology. It has six elements, and patients are diagnosed with Takayasu arteritis if at least three of the six criteria are present with a sensitivity and specificity of 90.5 and 97.8%, respectively. The elements are (1) age younger than 40 years at the time of diagnosis; (2) claudication of the extremities; (3) decreased pulsation of one or both brachial artery; (4) bruit over one or both subclavian arteries or the abdominal aorta; (5) 10 mmHg systolic blood pressure difference or more between the two arms; and (6) an arteriogram showing a narrowing/occlusion of the aorta, its primary branches, or large arteries of extremities, not due to atherosclerosis, fibromuscular dysplasia, or other causes [1, 2]. Our patient met all six criteria.

The second case represents an unusual presentation of a cerebral AVM presenting with cyanosis and shock, mimicking a severe coarctation on echocardiography. This AVM, located outside the cerebral hemispheres, was not detected by auscultation or on routine head ultrasound. The cardiac catheterization results confirmed the echo findings of reversal of flow in the aortic arch but demonstrated a markedly elevated SVC saturation, which raised the suspicion of an AVM.

In this case of a large AVM off the external carotid artery system, the absence of antegrade flow across the aortic arch mimicked an interrupted aortic arch. Ligation of the external carotid arteries returned normal antegrade flow across the aortic arch.

An association of congenital heart disease with a vein of Galen malformation has been previously described in a

case series review of 23 patients by McElhinney et al. [6]. Coarctation of the aorta was found in 9 of the 23 patients in their group. It may be the diversion of blood flow from the developing aortic isthmus to the AVM that leads to the aortic hypoplasia and/or coarctation.

Garcio-Monaco et al. [3] reviewed endovascular management of 30 children with congestive cardiac manifestations from cerebral AV shunts [3]. They noted that only 5% of children with cerebral AV malformations presented with cardiac manifestations. External carotid arterial, vertebrovertebral, and vein of Galen malformations were most frequently associated with congestive cardiac symptoms. They found that the younger the child at presentation, the higher the chances of having cardiac manifestations. The prognosis of severe congestive heart failure of cranial origin in the newborn period was very poor. They reported an overall mortality of 20% in their series. Beneficial changes in cardiac hemodynamics and symptomatic relief were obtained even following partial embolization.

There have been several reports of patients misdiagnosed initially as coarctation of the aorta on a clinical or echocardiographic basis. Rinelli et al. [8] reported three cases that were misdiagnosed as coarctation. The first case was a patent ductus arteriosus, the second case was an arteriovenous malformation, and the third case was a low thoracic coarctation. Onuzo et al. [7] reported two cases that were misdiagnosed as coarctation of the aorta. In both cases, hypertension and radiofemoral delay were noted. An echocardiogram revealed severe coarctation. Cardiac catheterization was done for balloon dilatation of the narrowing but revealed interruption of aorta. Both cases were surgically repaired [7]. Waikar et al. [11] reported a case that was misdiagnosed as coarctation of the aorta with aneurysm formation. The patient had a hematemesis and was operated on urgently. After cross-clamping of the distal aorta, he had a massive hemorrhage and died. On autopsy, an aortoesophageal fistula was seen and histology revealed Takayasu's arteritis.

Coarctation of the aorta has been previously reported as a complication of Takayasu's arteritis. Kashani et al. [5] described an 11-year-old boy who had Takayasu's arteritis that was complicated by coarctation of the abdominal aorta. He underwent balloon dilatation that was complicated by aneurysm formation. One year later, healing of the aneurysm and persistence of relief was shown. Sugawara et al. [9, 10] described two patients with Takayasu's arteritis that was complicated by coarctation of the aorta and occlusion of the arch vessels. Both patients were successfully surgically repaired.

Coarctation of the aorta is an important cause of hypertension in the pediatric population and of neonatal congestive heart failure after the ductus arteriosus closes. These cases demonstrate conditions that may mimic coarctation of the aorta by physical findings or by echocardiography. Takayasu's arteritis and cerebral arteriovenous malformation should be in the differential when evaluating such children.

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