Case Report

Suprarenal Abdominal Aortic Coarctation Diagnosed During Pregnancy

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Coarctation of the abdominal aorta is an extremely rare vascular defect in which congenital or acquired etiologies have been described. This case concerns a 30-year-old pregnant woman with 15-years history of uncontrolled hypertension and lower limb claudication presented with worsened hypertension during her first pregnancy. Magnetic resonance angiography study of aorta revealed a stenosis in abdominal aorta about 12mm from the origin of celiac axis accompanied by left sided aortic arch and right aberrant subclavian artery. This case highlights the importance of a throughout physical examination in patients presented with hypertension and it emphasizes considering the coarctation of the abdominal aorta during the diagnostic workup of hypertension, especially in young patients. In such cases magnetic resonance angiography of the aorta is a useful tool to reach a definitive diagnosis especially in pregnant women. Also to our knowledge, this patient is the first one found to have aortic arch malformation combined with an abdominal coarctation.

Introduction

Coarctation of the abdominal aorta is an extremely rare vascular defect in which congenital or acquired etiologies have been described. Typical symptoms are hypertension and lower limb and visceral ischemia which mostly occur later in life. We describe a 30-year-old pregnant woman with 15-years history of uncontrolled hypertension and lower limb claudication found to have abdominal aortic coarctation during evaluation for worsened hypertension during her first pregnancy.

Case presentation

A 30-year-old pregnant woman was admitted in her 24th week of gestation with an uncontrolled hypertension. Her medical history showed poorly controlled hypertension, which had been first noted nearly 15 years previously, and an intermittent claudication from 5 years ago.

On physical examination blood pressure was 200/120 mmHg in her arms and 100/90 mmHg in the legs. Pulse rate was 80/ min, radial pulses were strong, however femoral and dorsalis pedis pulses were weak and absent, respectively. The rest of physical examination was unremarkable.

Chest x-ray and ECG were normal. Echocardiography showed mild left ventricular hypertrophy. Although missed by multiple previous echocardiographic examinations abdominal aortic flow was compatible with coarctation but no gradient was seen in distal aorta due to severe stenosis or prominent collaterals.

Pulsed Doppler studies of renal arcuate arteries showed normal resistance index (resistance index of 0.62 in the right and 0.61 in the left). (Fig. 1)

Magnetic resonance angiographic study of aorta using pre-contrast and post-contrast TURBO FLASH 3D images with gadolinium revealed a stenosis in abdominal aorta about 12mm from the origin of celiac axis. Aortic arch was left sided and right aberrant subclavian artery was seen. No sign of aneurysmal dilatation was detected. (Fig. 2,3)

Discussion

Abdominal aortic coarctation was first described

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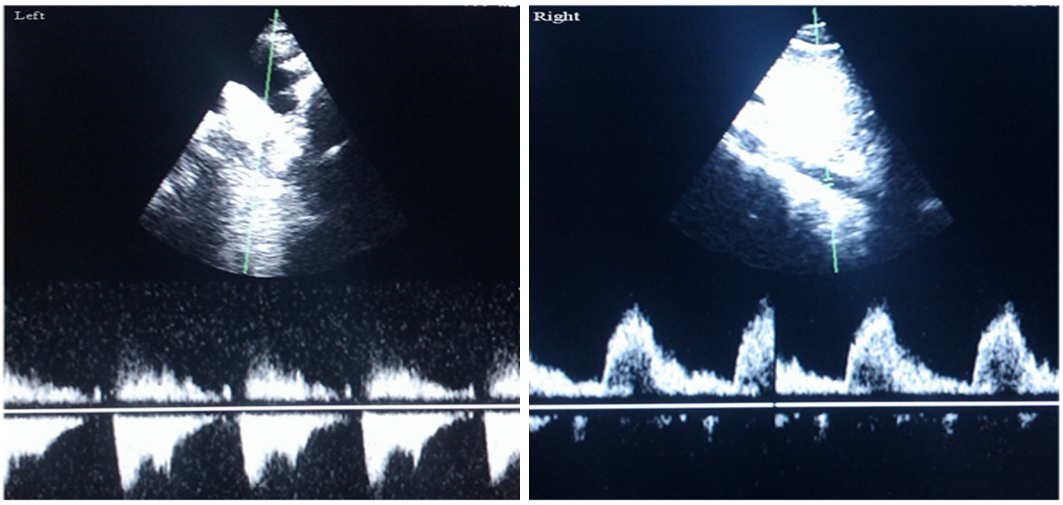

Figure 1. Continuous wave Doppler echocardiography of descending aorta shows no significant gradient (left). Pulsed wave Doppler echocardiography of abdominal aorta shows continuous flow with low velocity diastolic tail suggestive of abdominal coarctation of aorta (right).

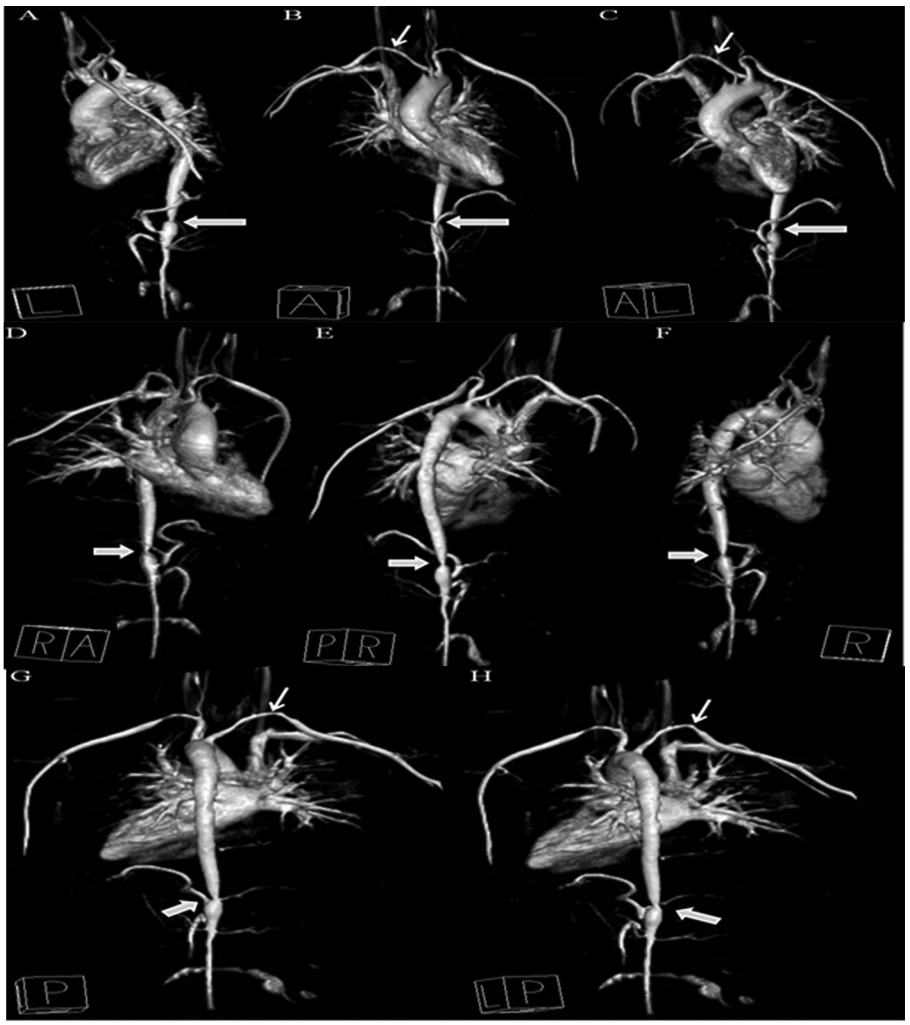

Figure 2. Magnetic resonance angiography of aorta shows suprarenal stenosis of abdominal aorta, about 12mm from the celiac axis (thick arrow). Left sided aortic arch is seen. Right aberrant subclavian artery is shown with the arrow. Each view is shown in the yellow box on the bottom of each figure; A, anterior; L, left; P, posterior; R, right.

in 1847 by Quain.[1]

Coarctation of the abdominal aorta, also known as middle aortic syndrome or mid-aortic dysplastic syndrome, is a rare vascular pathology caused by localized or extended narrowing of the abdominal or distal descending thoracic aorta secondary either to a congenital anomaly in the development of the abdominal aorta or to one of several acquired conditions such as infection, obliterative panarteritis, neurofibromatosis, retroperitoneal fibrosis, fibromuscular dysplasia, mucopolysaccharidosis and Takayasu's arteritis.[2-4] Most patients are young, with a mean age of 22 at diagnosis.[5]

Coarctations of abdominal aorta have different locations and lengths. More than 160 cases of abdominal coarctation have been described, with many anatomic variations.[6] Based on its locations four types of aortic coarctation have been described. The coarctation sites in types I and III is suprarenal and in types II and IV is infrarenal. Whereas types I and III are associated with renal artery stenosis, types II and IV are not.[7]

Patients with renal artery stenosis typically present with uncontrolled hypertension in the first and third decades of life.[7] However, our patient has been suffering from uncontrolled hypertension without signs of renal artery stenosis.

It is believed that claudication is a less common symptom in cases of suprarenal hypoplasia.[7] However, one of the major symptoms of our patient was lower limb claudication despite her lesion site which was suprarenal. Considering the presence of these two unusual symptoms in our patient it can be concluded that predicting the presenting symptoms based on the location of the coarctation site alone may seem illogical. Because in the presence

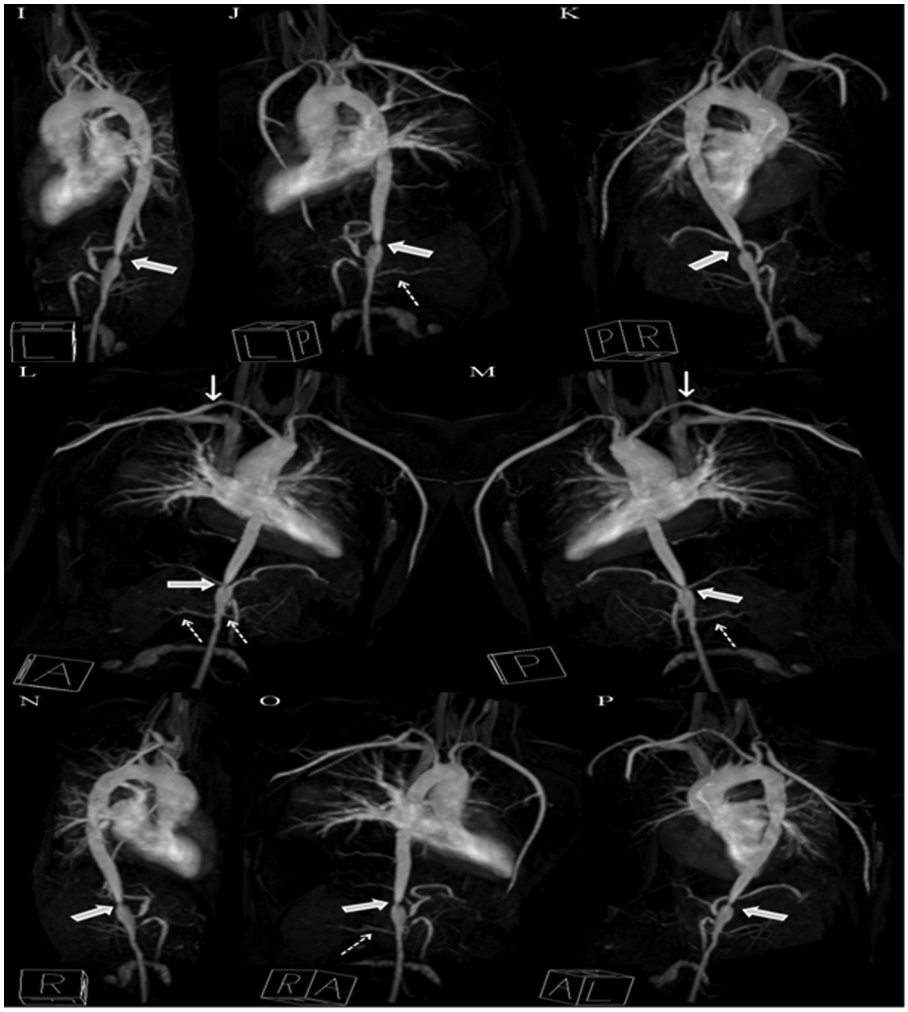

Figure 3. Magnetic resonance angiography of aorta; Suprarenal stenosis of abdominal aorta is located above the celiac axis (thick arrow). Right aberrant subclavian artery is shown with arrow. Renal arteries are shown with dashed arrow. Each view is shown in the box on the bottom of each figure; A, anterior; L, left; P, posterior; R, right.

of sufficiently narrow coarctation, signs and symptoms of both renovascular hypertension and vascular insufficiency in lower extremities may be seen even when the stenotic lesion is located in suprarenal portion of abdominal aorta. The presence of collateral vessels may explain why our patient with a stenosis above the celiac axis did not have symptoms of visceral involvement.

Hypoplasia of the major aortic branches distant from the coarctation, aneurysms of renal artery, atherosclerosis proximal to the lesion, intimal proliferation about the aortic branches are some of the vascular lesions reported to be associated with some cases of abdominal aorbi coarctation.[8] To best our knowledge, our patient is the first case of abdominal coarctation that is associated with aortic arch malformations including left-sided aortic arch and right aberrant subclavian artery suggesting possible congenital nature of the lesion.

The presence of these accompanied lesions may help to differentiate the congenital from acquired cases of abdominal aortic coarctations.

Coarctation of the abdominal aorta is an unusual cause of hypertension in pregnancy and there have been reported a limited number of hypertensive pregnant women diagnosed with abdominal coarctation.[9-12]

Maternal mortality associated with coarctation of the aorta approaches 4%. Deaths are usually related to aortic rupture or dissection, often associated with hypertension.

In summary, this case highlights the importance of a throughout physical examination, which in this case led to the discovery of an abdominal bruit and weak femoral pulses that raised the suspicion of aortic coarctation or renal artery stenosis as the cause of hypertension. Also it emphasizes that coarctation of the abdominal aorta, although rare (2% of all coarctations of the aorta) can be the cause of secondary hypertension and should be considered during the diagnostic workup of hypertension, especially in young patients. Moreover our case showed that in conventional echocardiography abdominal aortic flow may show evidences of presence of an anatomic pathology in the abdominal aorta. This manifested low velocity diastolic tail, as in our patient, thereby gives rise to use more sensitive and specific modalities of imaging to provide a definitive diagnosis such as magnetic resonance angiography or conventional aortoraphy. In the similar situations where the presence of abdominal coarctation is suspected in a pregnant woman, magnetic resonance angiography of the aorta can provide a useful tool to reach a definitive diagnosis. Since it is radiation free and can be done safely in pregnancy with no adverse effect on the fetus.

Acknowledgement

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