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CASE REPORT

Contribution of imaging modalities in the diagnosis of middle aortic syndrome

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1. Introduction

The coarctation of abdominal aorta, also known as middle aortic syndrome (MAS) is a rare vascular disease considered to be a life-threatening condition due to associated severe hypertension. It remains a challenging situation for clinicians because of its low prevalence. Imaging modalities represent essential tools for the diagnosis.

We reported two cases of MAS in children focusing on imaging features allowing positive diagnosis.

2. Case report 1

An 8-year old boy was referred to our department for uncontrolled hypertension. He was symptomatic of headaches. On examination blood pressure was 160/110 mmHg in the upper limbs and 100/60 in lower limbs. Femoral pulses were weak and an abdominal murmur was found in the aorto-renal region. Blood inflammatory markers, urinary and blood ionogram, creatinine serum level and proteinuria were negatives.

In order to investigate secondary hypertension causes, urinary catecholamines and their methoxylated derivatives as well as renin and aldosterone serum levels were performed. Only rennin serum level was high (71.5 pg/mg), while no abnormalities were observed in other urine and blood tests.

Echocardiography showed eccentric hypertrophy of the left ventricle, while left ventricular ejection fraction was normal. There was no left ventricular outflow obstruction. The aortic valve and the ascending aorta were normal and the aortic arch was well developed with a normal flow (Fig. 1A). However, subcostal views revealed severe narrowing of the abdominal aorta with reduction of its diameter from 12 mm to 5 mm. Doppler confirmed the presence of high velocity abdominal aortic flow with diastolic flow extension (Fig. 1B and C).

The multidetector computed tomography angiography (MDCTA) showed a narrowing of inter and infra renal abdominal aorta with a minimum caliber of $3 * 2.5$ mm, a repermeabilized occlusion of left renal artery, and a severe stenosis of the right renal artery. Aortic arch and its proximal branches were normal, mesenteric artery was spared (Fig. 2A and B) and there were no periaortic inflammatory signs.

Based on clinical examination and imaging investigations, the diagnosis of congenital MAS with bilateral stenosis of renal arteries was confirmed. An anti-hypertensive treatment was initially prescribed included full doses of betablockers, calcium channel inhibitors, and Thiazide diuretics, however hypertension was unsuccessfully controlled. Therefore, patient was referred to surgical treatment, consisting on prosthetic aorto-aortic bypass with renal arteries reimplantation.

3. Case report no. 2

A 11 year old girl was admitted for systemic arterial hypertension. The physical examination showed a low-grade fever (37.9 °C). Blood pressure at right arm was160/110 mmhg, while 90/50 mm Hg and 80/40 mm Hg were measured at left arm and lower limbs, respectively. In addition, upper peripheral pulses were asymmetrical. Abdominal exam was normal. Blood tests revealed an inflammatory syndrome with high erythrocyte sedimentation rate and elevated C reactive protein level.

Echocardiography showed no left ventricle hypertrophy Valvular aortic stenosis and isthmus aortic coarctation were ruled out, however there was a stenotic flow at the level of the abdominal aorta on subcostal views (Fig. 3A and B).

MDCTA revealed hypodense and extensive circumferential thickening of the descending thoracic and abdominal aorta. This thickening began within the origin of the left subclavian artery and ended with the celiac trunk. It was accentuated at the level of the abdominal aorta creating a severe stenosis (Fig. 4).

Furthermore, there was a significant stenosis of the origin of the left subclavian artery (Fig. 5) in addition to an occlusion of the superior mesenteric artery The other extra aortic vessels were normal.

Based on clinical and biological findings, absence of left ventricular hypertrophy and multifocal arterial lesions at MDCT, the diagnosis of MAS caused by Takayasu's disease was retained. The child was treated with the corticosteroids and immunosuppressive therapy for 13 weeks normalization of inflammatory syndrome. Despite combined antihypertensive therapy (full doses of betablockers, calcium channel inhibitors, and Thiazide diuretics),

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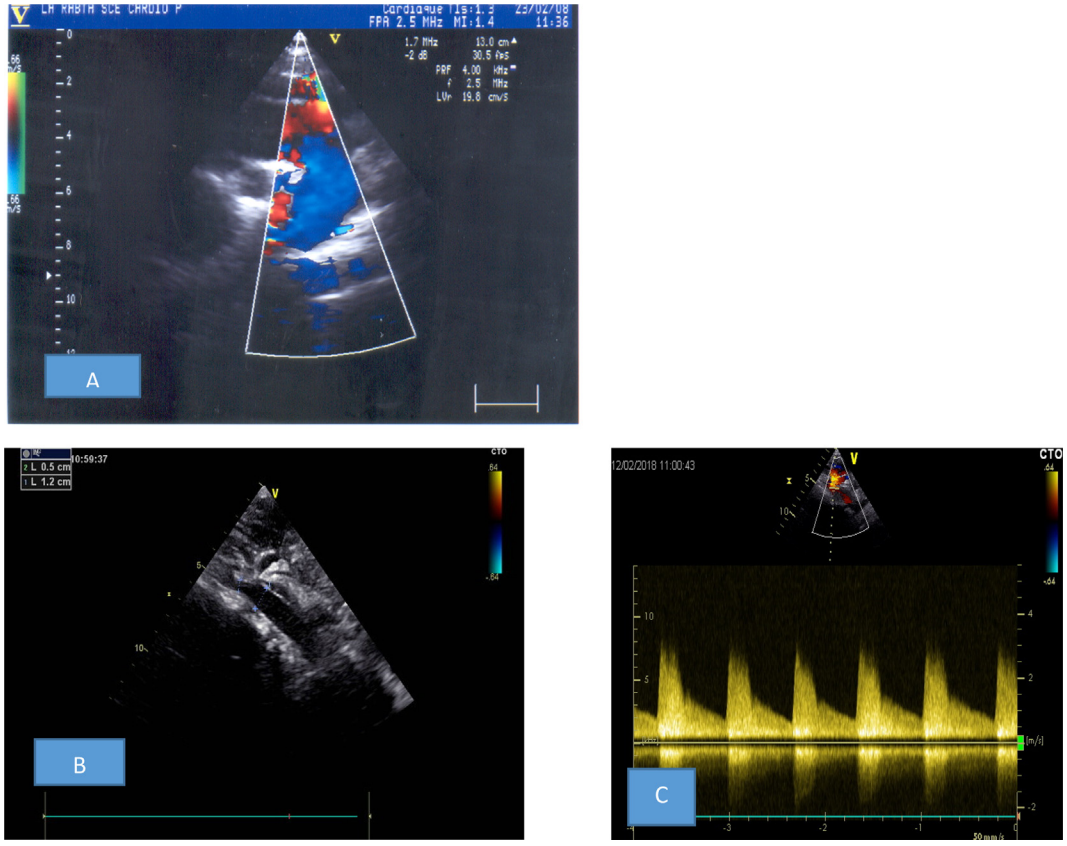

Fig. 1. echocardiography showed abdominal aortic coarctation. (A) Normal aortic arch in suprasternal view. (B) Narrowing of the abdominal aorta on a subcostal view. (C) A high velocity abdominal aortic flow on continuous wave Doppler.

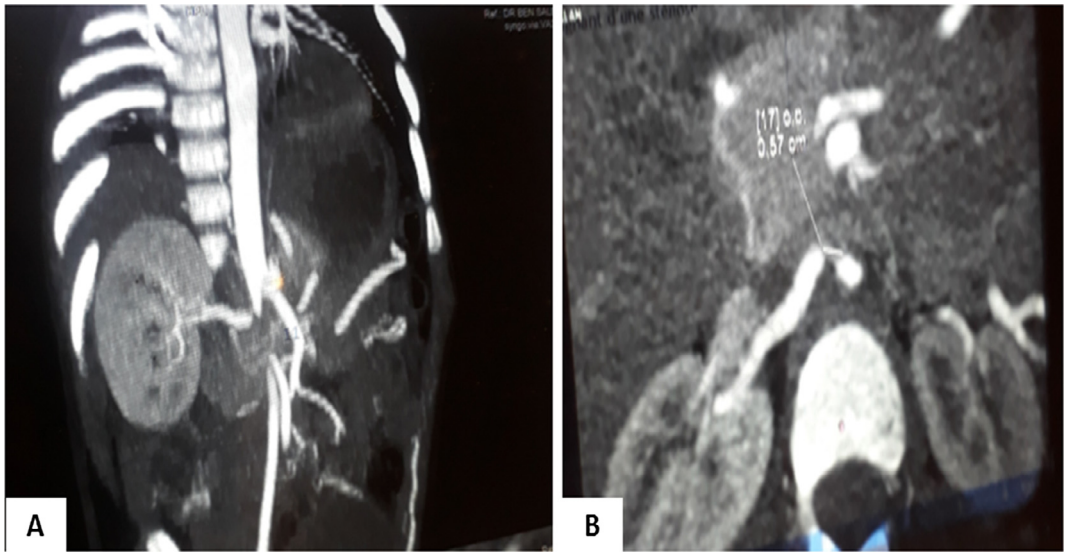

Fig. 2. The multidetector scan showed. (A) Narrowing of abdominal aorta. (B) An occlusion of left renal artery and a severe stenosis of the right renal artery streteched about 5.7 mm.

blood pressure remained uncontrolled. Therefore, aorto-aortic bypass was performed with with good short- and mid-term outcome.

4. Discussion

Aorta coarctation is commonly juxtaductal and classically located in the thoracic aorta distal to the origin of the left subclavian artery at about the level of the ductal structure.[1] However, the coarctation may involve the descending thoracic or abdominal aorta and is called MAS.[1] This clinical entity is rare, representing 0.5–2% of all cases of aortic coarctation.[2] MAS can be acquired or congenital. Acquired causes of MAS include Takayasu's disease, neurofibromatose and fibromuscular dysplasia.[3] Whereas congenital MAS is due to a developmental anomaly in the fusion and maturation of the paired embryonic dorsal aortas.[4]

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