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Case Report
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Abdominal Coarctation in a 9 Year Girl Mimicking Anxiety Attacks: Case Report

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Abstract

Background: Abdominal aortic coarctation is an uncommon vascular disease, representing approximately 2% of aortic coarctations, aortic coarctation most commonly occurs in the region immediately distal to the origin of the left subclavian artery. Clinical presentation of abdominal localization is variable especially when it occurs in children and may represent a rare cause of secondary arterial hypertension.

Case Report: We report a case of abdominal aortic coarctation diagnosed precociously in a 9 years old girl, who had showed non-specific symptoms. She complained dyspnea, retrosternal pain just before to fall asleep and palpitations and night fears, suggesting a diagnosis of anxiety attacks due to restarting of school time. We found accidentally increased blood pressure values in one evening measurement and confirmed by ambulatory blood pressure recording. So we started diagnostic process for differential diagnosis of hypertension causes and diagnosed abdominal coarctation, first suggested by an abnormal upper/lower extremities ratio and confirmed by CT and then by Angio-MRI.

Discussion: Aortic coarctation has a variable clinical presentation, depending on the severity and the site of obstruction. Abdominal aortic coarctations usually cause signs or symptoms during the second decade of life; the mean age of diagnosis is 22 years while our patient was just 9 years old. In our case, clinical presentation was not initially typical for aortic coarctation, because symptoms might suggest a generic diagnosis of anxiety attacks. Hypertension is not a frequent condition in children; when it is present, it's often a secondary form, so it needs a deep differential diagnosis process.

Conclusion: Abdominal aortic coarctation is a rare pediatric clinical condition, very difficult to identify because of its rarity and its variable symptoms. Essential was the occasional discovery of high blood pressure levels confirmed with ambulatory blood pressure monitoring. This element guided the diagnostic process to the research of causes of hypertension and allowed us to obtain the correct diagnosis.

Introduction

Abdominal aortic coarctation is a rare localization, the most common site of aortic coarctation occurs immediately distally to the origin of the left subclavian artery, while abdominal localization represents quite a an infrequent form that can be on a congenital basis [1], but may also be acquired.

Clinical presentation of abdominal localization is variable and may include arterial hypertension, heart failure, murmur, claudication, diminished femoral pulses and a leg blood pressure equal to or lower than arms [2].

We report a rare case of abdominal aortic coarctation diagnosed in a child who had showed apparently non-specific symptoms: anxiety attacks.

Case Presentation

When patient was admitted in our hospital, parents signed consent to the processing of personal data of the children.

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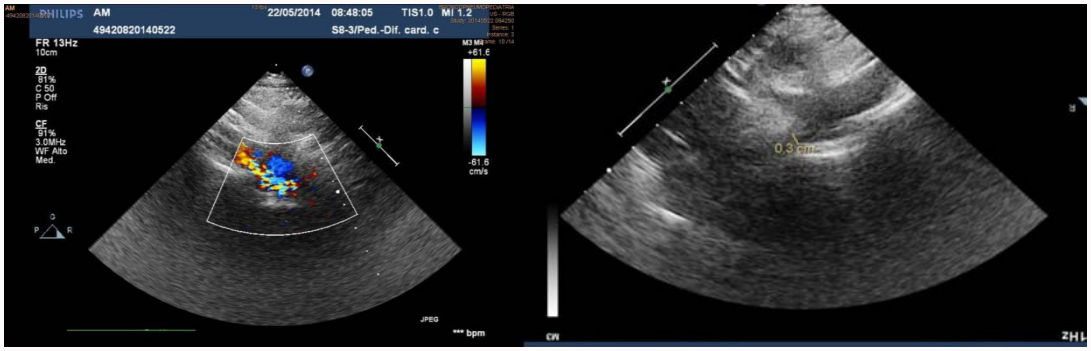

Figure 1: Vascular abdominal aorta colordoppler echography.

A 9 years old girl was referred to our hospital for dyspnea and retrosternal pain just before to fall asleep. The patient had a more recent history of recurring similar episodes, characterized by palpitations and night fears, in addiction to mild chest pain and short breathing difficulty; these symptoms should have suggested to run cardiac clinical and instrumental examinations but remote pathological anamnesis was characterized by transient patent ductus arteriosus with six-monthly cardiologic follow-up until the age of 5 years. For this reason only electrocardiogram was performed and a generic diagnosis of anxiety attacks due to restarting of school time had been the first diagnosis. Parents decided to ask a second opinion.

On admission, her general conditions were good. Heart auscultation noted a 1/6 systolic murmur at base of heart. Laboratory exams, spirometry, electrocardiogram and echocardiography were normal. During the first day of hospitalization we found an increased arterial blood pressure value in the evening and for this reason an ambulatory blood pressure recording was performed that finally revealed a before now undiagnosed hypertension. We completed diagnostic process for differential diagnosis of hypertension causes. The exams showed a normal renal function, a regular circadian rhythm of plasmatic cortisol levels and normal aldosterone blood levels; higher renin levels were found in clinostatism and in orthostatism (respectively 47,10 uUI/ml and 125,40 uUI/ml). In the while we assessed an abnormal difference in blood pressures between the upper and lower extremities: arterial blood pressure was 120/84 mmHg at right arm and 90/73 mmHg at right leg. A new Echocardiography excluded isthmic aortic coarctation; in effect there was a much rarer, especially in child, abdominal localization of aortic coarctation. Doppler vascular abdominal echography and contrasted-enhanced CT of chest and abdomen showed an isolate significant stenosis of abdominal aorta, with the stenotic tract originating vertebral arteries; this picture was confirmed by MRI and Angiography (Figure 1 and 2).

Once made the diagnosis, the patient has started a Ca-antagonist treatment to manage hypertension, while waiting for the choice of when and which kind of bypass correction.

Discussion

Coarctation of aorta comprises 5% to 10% of congenital cardiac lesions [2-4] but if we consider only coarctation involving the abdominal aorta they are much rarer with a rate of 2% of all coarctations [5].

Abdominal aortic coarctations clinical presentation is variable, depending on the severity and the site of obstruction. Usually cause signs or symptoms during the second decade of life; the mean age of diagnosis is 22 years while our patient was just 9 years old. If untreated, this condition has been associated with stroke, progressive left ventricular hypertrophy with congestive heart failure and flash pulmonary edema, and less often with renal insufficiency. In one review, 55% of untreated patients died at a mean age of 34 years [5].

Symptoms of aortic coarctations are related to the kind and site: in the neonatal isthmic severe localization there are early symptoms and signs of heart failure and respiratory distress such as tachypnea, grunting, retractions, pale skin , heavy sweating, diminished or absent lower extremity pulses, increased upper extremity pulses, difficulty in feeding But in the adult post isthmic form patients may remain for long time almost asymptomatic or may present headache, systemic hypertension, nose bleeding, muscle weakness, cold feet, leg cramps and claudication; in effect our patient after diagnosis realized to have suffered of leg cramps. Physical examination can note strong upper body pulses and diminished or absent leg pulses, which are corroborated by a difference in blood pressures between the upper and lower extremities. After infancy, the systolic blood pressure in the lower extremities is usually higher (5 to 20 mm Hg) [2-6] than in the upper extremities as a result of the standing wave effect. If the leg systolic blood pressure is equal to or lower than the arm pressure, the diagnosis of aortic coarctation should be considered [2-7], like in our patient, who presented a systolic pressure gradient of about 30 mmHg in legs.

In our case, clinical presentation and the history of frequent "normal Echocardiograms" contributed to mislead an early diagnosis also because specific symptoms began to be evident just before school restarting suggesting a generic diagnosis of anxiety attacks that, according to parents' feelings, conformed with the irritable disposition of the preadolescent girl. Panic or anxiety disorders may be characterized by recurrent unexpected episodes of severe anxiety, which typically reach their peak within 10 minutes and last around 30-45 minutes [8]. These symptoms can look similar to those referred by our patient, who lamented an anxiety state associated to dyspnea and heart rate acceleration. Luckily in this case a new complete extended evaluation of the patient, including blood pressure ambulatory monitoring, thanks to the individuation of high blood pressure quickly redirected the diagnosis: in fact hypertension is not a frequent condition in children with a prevalence ranging from 4% to 17% [9-10]. The probability of a secondary hypertension is inversely proportional to the age of the patient and directly proportional to blood pressure levels [11]; but a rare abdominal aortic coarctation caused just an apparently mild hypertension in this girl.

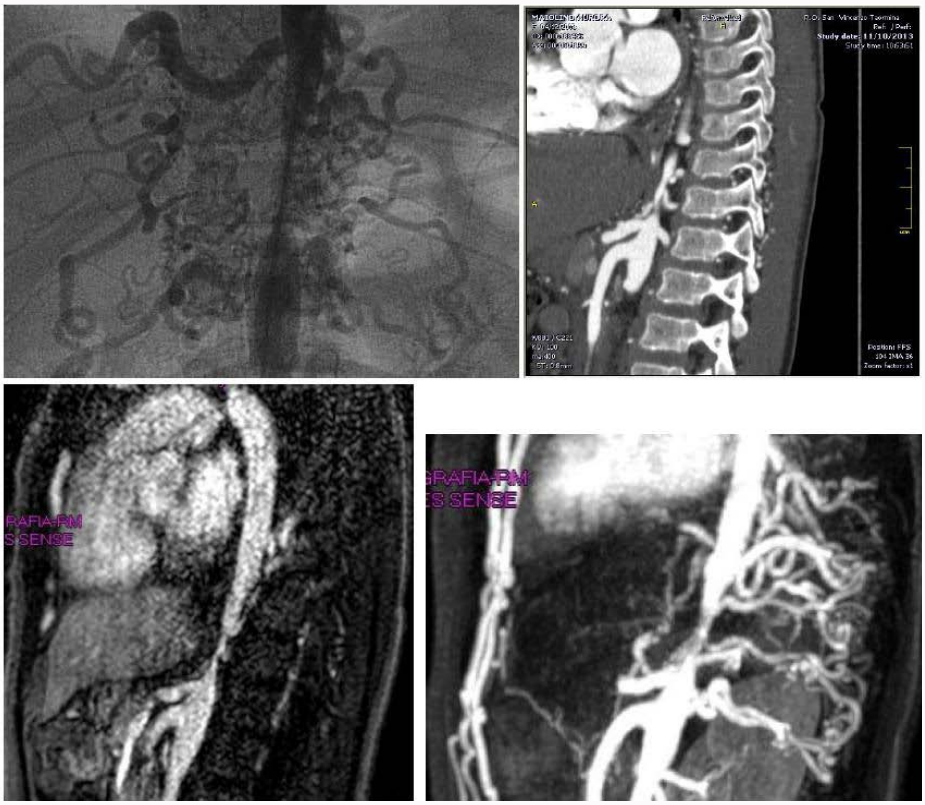

Figure 2: Angiography, CT angiogram and MR of the site of coarctation.

Regarding abdominal coarctation, we can suppose to have diagnosed a congenital form. Considering the acquired forms, they have been associated with pathologies such as Takayasu syndrome, William's syndrome, neurofibromatosis, fibromuscular dysplasia, retroperitoneal fibrosis, and mucopolysaccharidosis [12]. In our case we could exclude neurofibromatosis, mucopolysaccharidosis and Williams's syndrome because of absence of typical stigmata; so the differential considerations are limited to Takayasu arteritis and non-inflammatory aorto-arteriopathy, such as fibromuscular dysplasia [13]. Takayasu arteritis usually is multi distrectual and it can be differentiated from fibromuscular dysplasia for the general inflammatory state and by histologic examination [14]. The only therapeutic differences between the two conditions would be the use of anti-inflammatory therapy in Takayasu arteritis. D'Souza et al. [14] with a study in 1998 conclude that differentiating between Takayasu arteritis and fibromuscular dysplasia may not be as important as the management itself of aortic coarctation [13].

Management decisions for patients with coarctation of the aorta depend upon patient age, presentation, and the severity of the lesion. Medical therapy consists in hypertension management with antihypertensive drugs (beta-blockers, ACE inhibitors, angiotensin-receptor blockers or Ca Antagonist), waiting to choose the best surgical option. There are different methods for the treatment of aortic coarctation, including surgical repair or percutaneous balloon angioplasty with or without stent placement [15]. Surgical repair of coarctation can be achieved by several techniques: resection with end-to-end anastomosis, subclavian flap aortoplasty in infants with long-segment coarctation, a bypass graft across the area of coarctation when the distance to be bridged is too long for an end-to-end repair or prosthetic patch aortoplasty. Balloon angioplasty has been recommended as the preferred treatment for children and adults with native coarctation or recoarctation after surgery [15]. It has been suggested that stenting after balloon angioplasty lowers the risk for complications and has a beneficial effect on long-term survival. However, primary stenting has an important limitation which is the failure to adapt to the growing vessel in a child. To overcome this problem, redilatation during follow-up has been described [16]. In our case stent implantation can't be realized because vertebral arteries originate from the stenotic tract, so we have to exclude this option. She will need to bypass the stenotic tract with the addition of all the implications connected with the growth of the girl. In this while the patient has begun antihypertensive therapy with Ca-antagonists, in order to manage arterial blood hypertension.

Conclusion

Abdominal aortic coarctation is a rare pediatric clinical condition that can be quite difficult to identify because of its rarity and its variable symptomatology. Anamnesis and physical examination are the first steps to approach any patient. Our case presented initially non-specific clinical features. Essential was the occasional discovery of high blood pressure levels confirmed with ambulatory blood pressure monitoring. This element guided the diagnostic process to the research of causes of hypertension and allowed us to obtain the correct diagnosis.

Author's Contribution

Conception and design: Pietro Sciacca, Paolo Guccione.

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Draft of the manuscript: Ileana Stella, Francesco Sciacca.

Critical revision of the manuscript and important intellectual

content: Pietro Sciacca, Salvatore Leonardi.

Final approval: Pietro Sciacca.

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