CASE REPORT

Hypertension, Headache, Leg Pain: Aortic Coarctation in Child Revisited

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Abstract

Arterial hypertension in children up to 6 years of age is mostly secondary, and renal parenchymal disease is the leading cause of hypertension in children. Coarctation of the aorta is a rare cause and should be always excluded. A 6-year-old girl was referred to pediatric nephrology consultation for enuresis, high blood pressure, recurrent night headache, and sporadic pain in the lower extremities. On examination, a systolic murmur was detected, radiating to the carotids and the back, and the patient had weak femoral pulses as well. The echocardiogram confirmed a severe aortic coarctation. Surgical correction was performed and resulted in hypertension improvement and resolution of symptoms. Coarctation of the aorta may not be diagnosed in the neonatal period. The present case report highlights the importance of blood pressure monitoring in children and the need for a careful investigation of the secondary causes of hypertension and an early diagnosis of coarctation of the aorta for a timely surgical correction that improves prognosis.

Keywords: Aortic Coarctation/complications; Aortic Coarctation/diagnosis; Aortic Coarctation/therapy; Child; Hypertension/etiology

Keypoints

What is known:
- Coarctation of the aorta may not be detected early in life.
- In the etiological investigation of hypertension in the pediatric population, the exclusion of coarctation of the aorta is mandatory.
- An upper / lower extremities blood pressure systolic greater than 20 mmHg and decreased/absent femoral pulses leads to a probable diagnosis of coarctation of the aorta.

What is added:
- All children with repaired coarctation of the aorta need follow-up by pediatric cardiology.
- Regular assessment of blood pressure in children and/or pediatric follow-up is essential in primary health care.

Introduction

In recent years, there has been an increase in the prevalence of childhood high blood pressure. Currently, the prevalence of clinical hypertension in children and adolescents is about 3.5%, and the prevalence of high blood pressure is between 2.2%-3.5%.[1,2] preceded only by asthma and obesity out of all chronic childhood diseases.[3,4] In Portugal, the prevalence of hypertension in adolescents was estimated to be 13%.[5] American studies revealed that primary hypertension is currently more common. However, outside the United States of America, secondary causes continue to be more frequent in children up to 6 years of age, while essential hypertension begins to be predominant in older children.[3,6]

Kidney disease is the most common secondary cause of hypertension in children and adolescents.[7] Renal parenchymal disease and renal structural abnormalities are present in 34%-79% of children / adolescents with hypertension and 12%-13% of children with renovascular diseases.[8,9]

Regarding cardiac causes, coarctation of the aorta is rare and accounts for about 0.2% prevalence of hypertension in adults. However, it should always be excluded in the etiological investigation of hypertension in the pediatric age group.[10]

Coarctation of the aorta is a congenital defect present in 4%-6% of patients with congenital heart disease.[11] Although coarctation of the aorta is characterized by a segmental narrowing of the aorta, which is more frequent in the aortic isthmus, it can occur anywhere

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in the aorta. Coarctation of the aorta can be either simple or complex when accompanied by other cardiac malformations. However, the bicuspid aortic valve and the ventricular septal defect are the most common associated malformations. If the narrowing is not very severe, newborns and children may be asymptomatic or have subtle clinical manifestations, resulting in delayed diagnosis. In children with hypertension, coarctation of the aorta screening with a complete clinical examination is important and should include monitoring of arterial pressures in the four extremities and palpation of radial and femoral pulses. An echocardiogram confirms the diagnosis and provides detailed information on anatomy and hemodynamics.

Computed tomography angiography (angio-CT) and/or magnetic resonance imaging are important for the surgical or percutaneous therapeutic plan. Although survival increases after coarctation of the aorta correction, the survival rate in these patients is normally lower than in the general population. Re-coarctation and hypertension are the most frequent complications in patients with coarctation of the aorta.[12,13]

Case Report

A 6-year-old girl was referred by her primary care physician to pediatric nephrology consultation with complaints of recurrent night headaches and sporadic pain in both legs, which were more pronounced after physical exercise at school. On evaluation, the doctor also detected high systolic and diastolic blood pressure. There was no relevant family or personal history of the condition. Reportedly, the girl had undergone a neonatal echocardiogram after birth which did not reveal any heart disease. Observation showed persistent elevated arterial pressure in the upper and lower extremities (average 134/83 mmHg and 110/79 mmHg, respectively, and both higher than the 99th percentile, indicating significant systolic blood pressure gradient), stage 2 arterial hypertension,[14] a systolic murmur III/VI on the back of the chest radiated to cervical and dorsal areas, and bilateral weak femoral pulses. The patient was urgently referred to pediatric cardiology. An electrocardiogram showed criteria for left ventricle (LV) hypertrophy. An echocardiogram (Fig. 1) revealed the conserved global systolic function of left ventricle (LV diastolic diameter 44 mm, LV systolic diameter 8 mm, interventricular septum 9 mm, posterior wall 8 mm, and shortening fraction 36%), aortic valve with three asymmetric leaflets without functional changes, tortuous aortic arch, increased velocity (by continuous Doppler) with a corrected gradient of 80 mmHg flow with diastolic extension wave.

Thoracic angio-CT showed post-isthmic coarctation of the thoracic aorta with a very marked stenosis zone with a pencil tip configuration (Figs. 2, 3, 4), practically mimicking a site of aortic interruption, moderately hypoplastic aortic arch, the normal anatomy of the left aortic arch and supra-aortic trunks, left ventricle with dimensions at the upper limit of normal, slight concentric hypertrophy of the left ventricular myocardium, marked hypertrophy of the internal mammary arteries, intercostal and scapular arteries, indicating collateral circulation (Fig. 4).

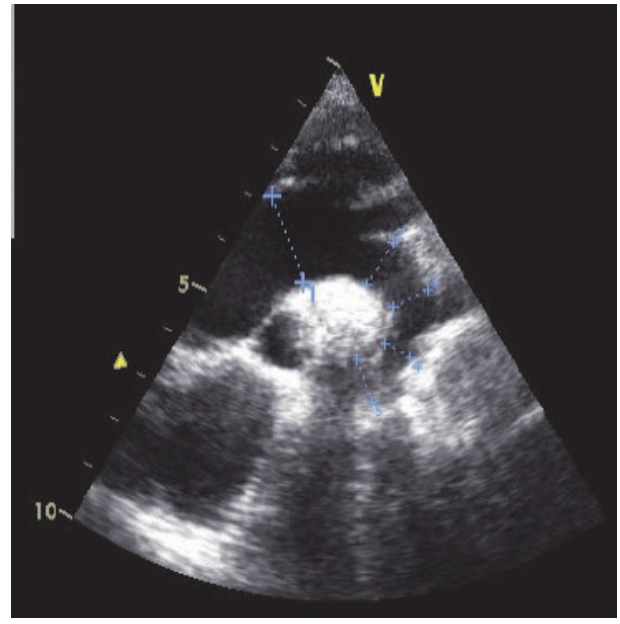

Figure 1. Transthoracic echocardiogram: Tortuous aortic arch from the isthmus region, with evident caliber decrease from the ascending aorta to the distal transverse arch.

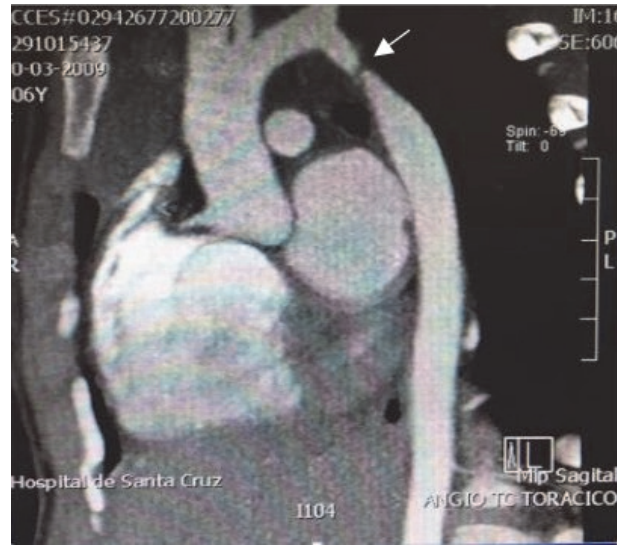

Figure 2. Computed tomography angiography: Post-isthmic coarctation of the thoracic aorta.

Renal ultrasound with Doppler was suggestive of upstream obstruction, with intra-renal circulation and renal arteries with low resistance, maintained peak systolic speeds resistance indexes together with an increase in systolic times. The laboratory evaluation of blood count, renal function, ionogram, lipid profile, and thyroid function was unremarkable. The patient started propranolol for blood pressure control and underwent coarctation of the aorta correction with a Gore-Tex patch two months after diagnosis. A postoperative echocardiogram revealed a hypertrophied left ventricle with maintained systolic function and a corrected gradient of 40 mmHg without diastolic extension. The child, currently 11 years old, is asymptomatic and her hypertension is controlled by propranolol. She remains under surveillance.

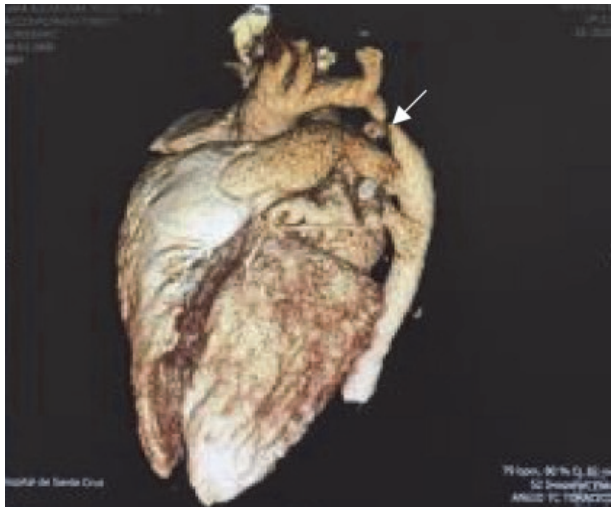

Figure 3. Computed tomography angiography (3D): Coarctation of the thoracic aorta immediately post-isthmic (white arrow), and concentric hypertrophy of the left ventricular myocardium.

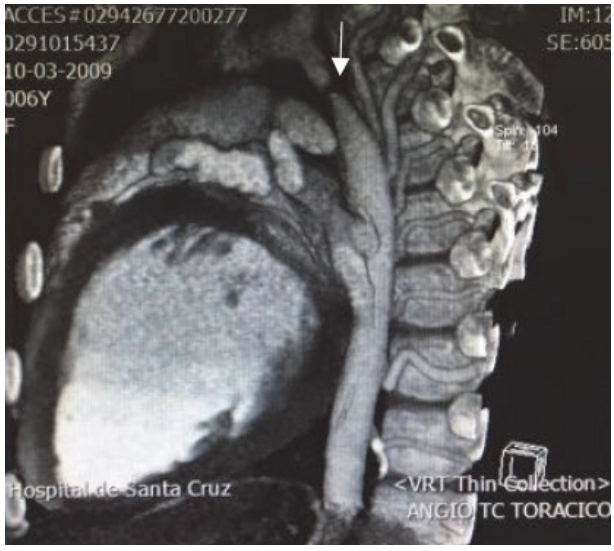

Figure 4. Computed tomography angiography: Coarctation of the thoracic aorta (white arrow), with exuberant collateral circulation, with highly developed intercostal breast arteries.

Discussion

The prevalence of hypertension in pediatric age has been increasing. Studies have shown that high blood pressure in childhood correlates with hypertension in adulthood, an earlier onset of hypertension, and an increased risk of metabolic syndrome.[15] Left ventricular hypertrophy is the most prominent target organ damage caused by hypertension with a prevalence of 20%- 41%.[16]

Coarctation of the aorta is present in less than 1% of hypertension with known causes and is more frequent in males. The age of diagnosis depends on the severity of the stenosis. Cases of acquired diseases have been already described, but they generally correspond to congenital pathology. Although rare, coarctation of the aorta must be included in the etiological investigation of hypertension in pediatric age, as it may not be detected in the pre- and neonatal period. Clinical manifestations vary according to age, and three age groups can be distinguished.

Coarctation of the aorta does not cause hemodynamic changes *in utero*. However, after birth, with the closure of the *ductus arteriosus* and *foramen oval*, manifestations may appear in the first weeks of life. Newborns and infants with patent *ductus arteriosus* or *foramen oval* may be asymptomatic, unlike heart failure, in which these channels are closed and there is a manifestation of low output. In children, the diagnosis is usually deferred since the majority are asymptomatic and have subtle clinical manifestations. Some children may report chest pain, cold extremities, and claudication with physical exertion. Adolescents and adults typically appear with signs / symptoms originating from hypertension.

Clinical diagnosis is suggested and should include careful evaluation of blood pressure and palpation of the pulses in the four extremities. Examination shows lower systolic blood pressure in the lower extremities compared to the upper extremities, a broad pulse in the brachial or radial arteries, and decreased or absent pulse in the femoral artery.

According to the guidelines of the American Academy of Pediatrics,[14] European Society of Hypertension,[5] and American Heart Association,[17] all children with confirmed hypertension must undergo complete clinical examination including the assessment of blood pressure in all four extremities and palpation of the radial and femoral pulses. An upper / lower extremities blood pressure differential greater than 20 mmHg and decreased / absent femoral pulses leads to a probable diagnosis of coarctation of the aorta. Different treatment approaches are available based on the age group, morphological and functional characteristics, and medical-surgical decisions.

Correction should be carried out as early as possible, preferably in childhood, to prevent the development of hypertension and minimize target organ damage. Hypertension immediately after correction of coarctation of the aorta is frequent, justifying pharmacological therapy and returning to normal blood pressure in most children.[18] The basic mechanisms of paradoxical hypertension include sympathetic activation and renin stimulation, both of which can be targeted using beta-blockers.[19] The role of prophylactic propranolol has been described in two prospective trials,[20,21] showing that two weeks of prophylactic propranolol and one week of postoperative propranolol significantly reduced the rise in post-operative blood pressure. After correction, survival increases compared to unrepaired coarctations (being 90% at 20 years of age and 74% at 30 years of age). However, the survival rate remains lower in these patients compared to that in general population.[12] All children with repaired coarctation of the aorta need follow-up by pediatric cardiology and should undergo regular imaging examination.[22] There is a risk of hypertension, re-coarctation, aortic aneurysm / pseudoaneurysm, and sudden death after correction.[13] This clinical case highlights the fundamental role of primary health care in blood pressure surveillance in child health consultations which allows for early detection, treatment, and prognosis of the condition.

Author Contribuitions

RRA participated in acquisition of data. RRA, AT and SD participated in the analysis or interpretation of data. RRA participated in the drafting of the manuscript. CPG, AT and SD participated in the critical revision of the manuscript. All authors approved the final manuscript and are accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved.

Conflicts of Interest

The authors declare that there were no conflicts of interest in conducting this work.

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Protection of human and animal subjects

The authors declare that the procedures followed were in accordance with the regulations of the relevant clinical research ethics committee and with those of the Code of Ethics of the World Medical Association (Declaration of Helsinki 2013).

Provenance and peer review

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Confidentiality of data

The authors declare that they have followed the protocols of their work centre on the publication of patient data.

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Hipertensão, Cefaleia, Dor nas Pernas: Coartação da Aorta numa Criança Revisitada

Resumo:
A hipertensão arterial em crianças com idade inferior a 6 anos é principalmente secundária, e a doença do parênquima renal é a causa secundária mais comum de hipertensão em crianças. A coartação da aorta é uma causa rara, mas deve ser sempre excluída. Uma menina de 6 anos foi encaminhada para consulta de nefrologia pediátrica por hipertensão, cefaleia noturna recorrente e dor esporádica nos membros inferiores. No exame físico, detetou-se sopro sistólico com irradiação para as carótidas e dorso, com pulsos femorais fracos. O ecocardiograma confirmou uma coartação de aorta grave. A correção cirúrgica foi realizada com melhoria da hipertensão e resolução dos sintomas. A coartação da aorta pode não ser diagnosticada no período neonatal. Este caso destaca a importância da vigilância da pressão arterial na infância, a necessidade de uma investigação cuidadosa das causas secundárias para hipertensão e, para a coartação da aorta, um diagnóstico precoce, para uma correção cirúrgica precoce, melhorando o prognóstico.

Palavras-Chave: Coartação Aórtica/complicações; Coartação Aórtica/diagnóstico; Coartação Aórtica/tratamento; Criança; Hipertensão/etiologia