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Case Report

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Coarctation of the aorta: a misdiagnosed cause of hypertension

Keywords: Coarctation of the aorta, congenital heart disease, hypertension

Abbréviations: CoA, coarctation of the aorta; CTA, computed tomography angiography; TTE, transthoracic echocardiography

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Introduction

Coarctation of the aorta (CoA) is a congenital segmental narrowing of the aortic wall, affecting up to 6% of all new-born with congenital heart disease. This condition may persist asymptomatic for many years after birth explaining late diagnoses in up to 60% of patients. In these patients, a long-term overload of the left ventricle results in hemodynamic repercussions with an increased risk of secondary hypertension, aortic aneurism and myocardial infarction. In fact, unoperated patients have a life-expectancy of as low as 35 years old. Clinically, patients may remain asymptomatic for years, although up to half may present lack of lower limbs pulse, typical systolic-diastolic murmur and an increased pressure gradient by echocardiography, which is required for the diagnosis. This method allows measuring the aortic gradient, but eventually a computed tomography angiography (CTA) of the aorta may be also necessary. Treatment involves clinical management of hypertension and heart failure symptoms, followed by surgical or balloon angioplasty for the correction of the narrowed segment. Though the surgical correction is often successful, life-long clinical and echocardiographic follow-up is required as re-coarctation occurs in one-third of patients.

Patient information

A 20years-old female patient was found to be hypertensive in a check-up visit in 2006. She had no personal or family history of cardiovascular disease and, at the time, was not under any medication. At the first visit, she had a 140x90mmHg blood pressure, and her physical exam showed a systolic aortic heart murmur and no other finding was reported. Echocardiographic and biochemical analyses were performed with negative results for pheochromocytoma and coarctation of the aorta. The patient had a prescription for thiazide diuretic and initiate annual clinical follow-up. In the following visits, her blood pressure levels had increased progressively reaching 180x110mmHg in spite of regular use of 4 different classes of antihypertensive drugs.

In 2015, she decided to seek a second medical opinion. At this visit, she had absence of lower limbs pulse, shortness of breath on exertion (NYHA II), lower limbs edema and uncontrolled blood pressure. Echocardiogram showed an elevated aortic gradient of 70mmHg, and an AngioCT demonstrated unequivocally an aortic coarctation after the emergence of left subclavian artery. She then underwent a cineangiocoronariography for anatomy assessment. The diameters were 6 and 14mm for the coarcted and descending aorta, respectively (Figure 1).

She decided to undergo a balloon angioplasty, motivated by the perspective of a shorter hospitalization time and better recovery. She underwent the procedure in September 2015 with the insertion of an endoprosthesis (Figure 1). After the procedure, she remained hospitalized for 1 day and resting for 5days after discharge. After 10 days, she was able to work and, after 3weeks, she was fully recovered and able to exert physical activity. Her blood pressure was controlled with Atenolol 50mg/day for 3months and she took Aspirin 100mg/day for 6months following the procedure. After one-year, she underwent an angio CT, which showed adequate function of the endoprosthesis, and an echocardiogram which showed normal cardiac function, an adequate aortic gradient of 12mmHg and no valvar nor myocardial abnormalities.

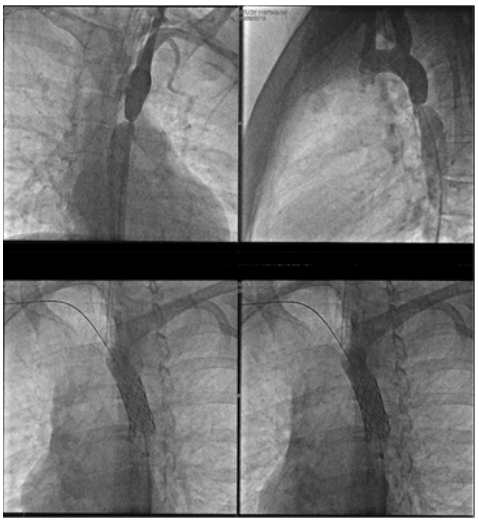

Figure 1 Cardiac catheterization during procedure and results.

Currently, she is 33 years-old, works as an ENT doctor, practice physical activity regularly and remains asymptomatic without medications. She is under life-long medical surveillance with annual echocardiographic and clinical evaluation.

Discussion

In this manuscript, we described a case of an adult female diagnosed with coarctation of the aorta 10years after the beginning

of her symptoms. More importantly, this patient presented as a young female with resistant hypertension. A short investigation was performed with echocardiogram and biochemical analysis, and a diagnosis of primary hypertension was given. It was only after 10years of disease progression and occurrence of new symptoms, such as worsening shortness of breath and lower limbs edema, that a new medical evaluation was provided, when an Angio CT was performed giving the definitive diagnosis of coarctation of the aorta (CoA). A few aspects of this case are worth mentioning.

Firstly, it is worth stressing that this patient remained asymptomatic until she was 20years-old, when she presented with an elevated blood pressure and heart murmur at a checkup visit. Her case is in agreement to a previous case-series performed by our group, in which we found that 40% of patients were asymptomatic and 60% were hypertensive at diagnosis. Note worthily, she had palpable lower limbs pulse. Although absence of lower limbs is commonly related to CoA in different reviews, our case-series reported that only half of patients had this finding at diagnosis.[1]

Importantly, at her first visit an echocardiogram was request and the CoA diagnosis was not concluded. Of note, transthoracic echocardiography (TTE) has a diagnostic accuracy of 68%, which may be increased to 90% when a color or continuous wave spectrum doppler is performed.[2] According to the ACC/AHA 2008 Guidelines, if the diagnosis of CoA is suspected or confirmed in the TTE exam, CT angiography (CTA) is mandatory,[3] as it not only has a 100% diagnostic accuracy, but also has the ability to measure the diameter and length of aortic coarctation. Furthermore, CTA is currently the first choice for follow up and surveillance of patients treated with stent implantation, as it allows early detection of stent fracture and displacement.[4] In the reported case, while the TTE did not confirm the diagnosis at the first visit, the requisition of an CTA was crucial for the diagnosis in a second moment. Also, the CTA was correctly adopted as the surveillance method after stent implantation.

As previously mentioned, the life-expectancy of unoperated patients is as low as 35years-old. Hence, prompt correction of aortic coarctation is mandatory, in order to improve long-term survival and attenuate the incidence of severe complications, such as aortic aneurysm, accelerated coronary artery disease and heart failure. Classically, treatment of CoA is performed by a left thoracotomy and resection of the coarctation segment, followed by an end-to-end anastomosis. Regardless its low surgical mortality (<1%), this technique requires a longer post-operative recovery and around 0,3% of patients become paraplegic due to spinal cord ischemia during the procedure.[5] Accordingly, equally safe techniques have been developed over the past decades as an alternative to the treatment of CoA. In this, the use of endovascular stenting has proved to be an effective and safe alternative treatment for older children and adults, allowing an effective correction of the aortic coarctation with comparable success rates.[5] A growing body of evidence have supported the use of stenting as a save method, with complications such as neointimal proliferation and aortic dissection only rarely occurring in well-designed trials.[5] Indeed, in the reported case, a female adult submitted to endovascular stenting showed full recovery, adequate blood pressure control and no complications in a long-term follow up after stenting.

Ethical statement

Authors had no conflict of interest in the aforementioned case report.

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None.

Conflicts of interest

The authors declare there are no conflicts of interest related to the article.

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