CASE PRESENTATION

What is really kinky: resistant hypertension and kinking of aorta

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Abstract: Kinking of aorta is a rare congenital anomaly described by elongation and tortuosity of the aorta, with a slight narrowing of the lumen. Although it can be very similar to an aortic coarctation, the lack of pressure gradient above and below the kinking gives to this anomaly a benign description. We report on a case of resistant hypertension in a man of 52 years old with multiple risk factors that associated kinking of aorta. We discuss the relation between these two pathologies in a multifactorial context, especially because there are several opinions that suggest there may be an etiologic correlation. However, given the additional risk that this anomaly brings, mainly for dissecting aneurysm, we consider important a close careful follow-up of this type of patient.
Keywords: resistant hypertension, kinking of aorta, pseudocoartaction

Rezumat: Kinking-ul aortei este o anomalie congenitală rară, caracterizată prin alungirea și tortuozitatea aortei, cu o ușoară îngustare a lumenului acesteia. Deși poate fi foarte similară unei coarctații aortice, lipsa gradientului de presiune deasupra și dedesubtul angulării conferă acestei anomalii un caracter benign. Raportăm cazul unui bărbat de 52 de ani cu hipertensiune arterială rezistentă la tratament, cu factori de risc multipli la care s-au asociat și aceat/ angulare/meandrare a aortei. Discutăm despre legătura dintre aceste două patologii survenită într-un context multifactorial, mai ales pentru că există mai multe opinii care sugerează că între acestea ar putea exista o corelație etiologică. Având în vedere riscul suplimentar pe care această anomalie îl aduce, în principal legat de apariția unui anevrism disecant, considerăm importantă supravegherea atentă și de durată a unui asemenea tip de pacient.
Cuvinte cheie: hipertensiune rezistentă, kinking de aortă, pseudocoarctație

INTRODUCTION

Pseudocoarctation or kinking of aorta is a rare congenital anomaly defined by elongation and tortuosity of the aortic arch and thoracic aortic segment with a slight narrowing of the lumen. Since the first use of the term „pseudocoartaction" by Dotter and Steinberg in 1951, over 150 cases of the disease have been reported[1]. Although it can be very similar to an aortic coarctation, there are several differences between these two entities that worth be mentioned. Pseudocoarctation of aorta presents no pressure gradient above and below the kinking, no rib notching, no collateral circulation, no symptoms. One of the objective signs that may be present in these patients is a precordial systolic murmur explained by the turbulance of the blood flow through the buckled aortic segment or from associated congenital defects[2].

The diagnostic is strictly imagistic, in most cases the anomaly being first described on a casual chest X-ray. The radiology sign that suggest the kinking of aorta is the presence of a round opacity of soft-tissue density that exceeds the contour of the heart, an abnormal mediastinal mass that can be easily mistaken for aorta coarctation, aneurysm, tumor or patent ductus arteriosus. In this context, it is vital that the presumptive diagnostic be confirmed with thoracic CT or angiography[3]. Confusions between coarctation and kinking should be avoided, in coarctation a post-stenotic dilation may occur, but there is identifiable an associated transstenotic pressure gradient[4]. Sometimes this

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disease may be associated with other congenital lesions like congenital aortic stenosis, bicuspid aortic valve, coarctation of the aorta, corrected transposition, ventricular septal defect, patent ductus arteriosus, fibroelastosis and aneurysms of the aortic sinuses, all these associations imposing a rigorous evaluation of this kind of patients[1].

On the other hand, resistant hypertension is defined as hypertension that remains uncontrolled under treatment with more than three antihypertensives agents, including a diuretic. In contrast to this true hypertension, „pseudoresistant" hypertension can be caused by poor clinic blood pressure measurement technique, patient non-adherence/ intolerance to prescribed medication, or white coat hypertension[5]. The association between uncontrolled blood pressure and kinking of aorta is not a very common discovery, with few citation in the literature. Other similar cases suggest there may be a connection between the two conditions, whether prognostic or etiologic.

CASE PRESENTATION

A 52-year-old male patient, with multiple cardiovascular risk factor (smoking, alcohol abuse, obesity, sedentary), diagnosed with grade 3 hypertension and angina pectoris at the age of 44, arrived to Emergency Room complaining nocturnal paroxysmal dyspnea, orthopnea, effort dyspnea and occipital headache, all symptoms being associated with high blood pressure. Patient history reveals a previous addmision in the cardiology clinic a year earlier with similar symptoms. After it was ruled out a secondary cause of hypertension, he do not declare a family history of hypertension and the recomandations for hypertension management included an association of five antihypertensive drugs: Candersartan 16 mg x 2 daily, Amlodipine 5 mg x2 daily, Carvedilol 6,25 mg x2 daily, Rilmenidine 1 mg daily, Indapamide 1.5 mg daily.

At the actual presentation, the clinical exam revealed a grade 2 obesity (BMI 38.87 kg/m^2), blood pressure values of 200/100 mmHg supine and 180/100 mmHg standing, lower limbs edema. The cardiac exam was normal, without any pathological murmurs.

Laboratory tests showed dyslipidemia (cholesterol=220 mg/dl, LDL-C=157 mg/dl, HDL-C=37 mg/dl, TG=153 mg/dl), hyperuricemia (uric acid=81mg/dl), normal renal and liver function, normal glycemia. The resting electrocardiography (ECG) showed sinus rhythm=60/min, ÂQRS=+15°, left ventricular hypertrophy with repolarisation abnormality[6].

The echocardiography confirmed the left ventricular hypertrophy (interventricular septum thickness 14 mm, LV posterior wall thickness 13 mm), the chamber diameters were enlarged (LA 52/54 mm, RA 44/43 mm, RV 42 mm, LV 60 mm), it was present a mild hypokinesia of the lateral wall and the basal portion of the septume, mild mitral regurgitation (grade I), moderate aortic regurgitation (grade II) and tricuspid aortic valve, aortic atheromathosis, normal dyastolic relaxation (E/A >1), LVEF= 48%.

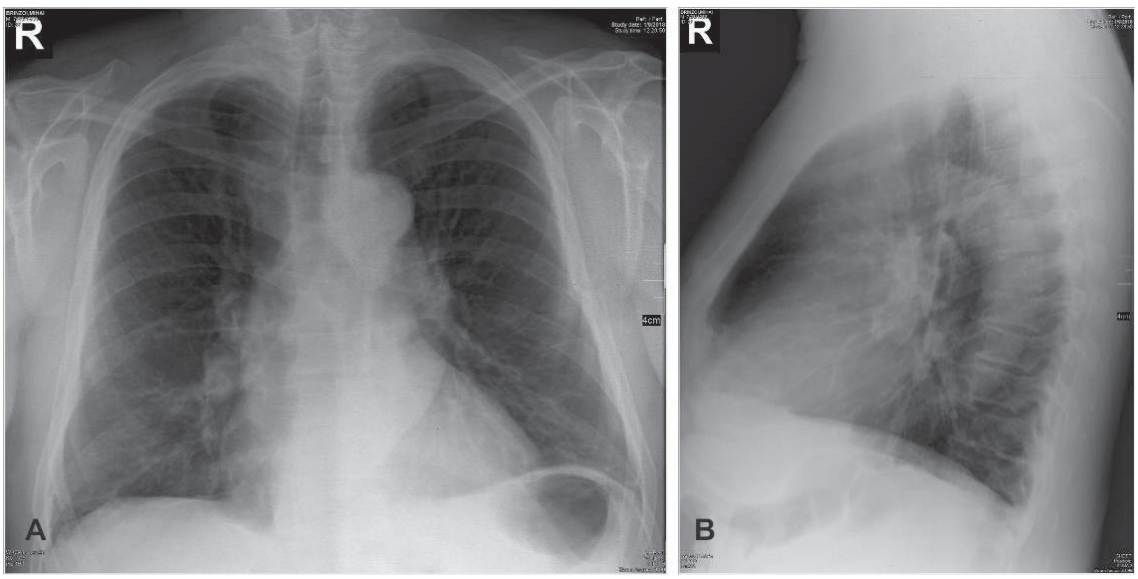

Figure 1. A) Chest X-Ray: frontal view. **B)** Chest X-Ray: profile view.

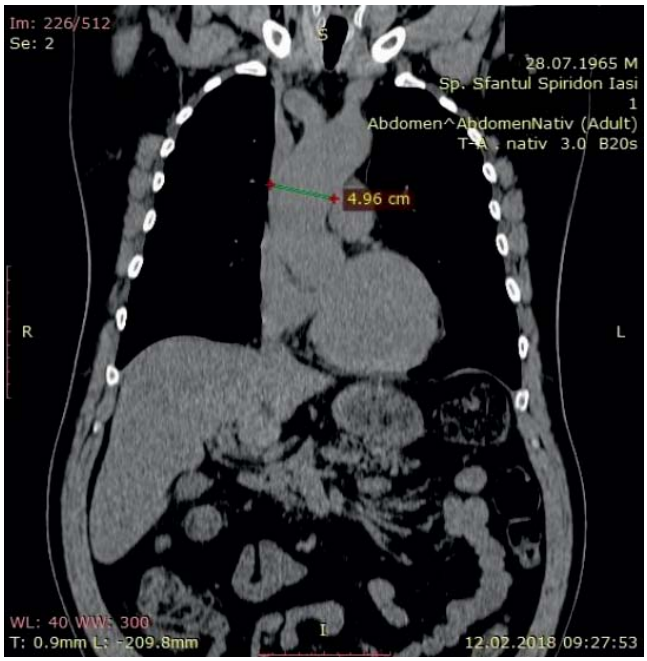

Figure 2. CT scan: aneurysmal dilatation of the ascending aorta.

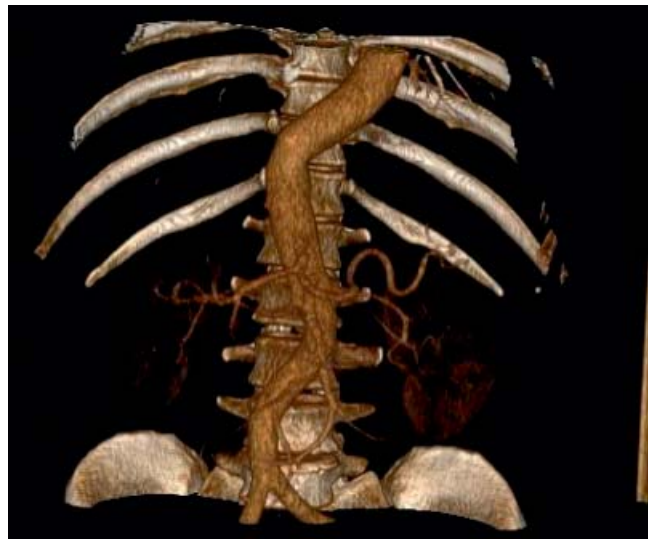

Figure 3. CT scan 3D reconstruction: kinking of descending aorta.

The chest X-ray revealed an enlarged cardiac area with cardiothoracic ratio of 0.52, calcified proeminent aortic arch, kinking of thoracic aorta (Figure 1a, 1b). The thoracic CT described aneurysmal dilatation of the ascending aorta (caliber of 49 mm) and kinking of the thoracic aorta in the inferior segment and of the abdominal infrarenal aorta, without sudden decalibration (Figure 2, Figure 3).

Abdominal echography was normal, with symmetrical and normal dimensions of the kidneys, without renal artery stenosis. The fundoscopy described hypertensive angiopathy grade II. The ankle-brachial index indicated an elevated vascular rigidity (with a value of 1.33 bilateral).

Doppler ultrasound of carotid arteries emphasize in the left common carotid artery an atheromatous plaque with a stenosis below 10%, without other alterations.

The treatment in Emergency Room consisted in intravenous Furosemid and sublingual Captopril, with the recommendation to continue the therapeutic regimen prior to admission, his five antihypertensives association. After three days of treatment, it was performed an ABPM that showed high blood pressure values throughout the day and a non-dipper profile. The main recorded values were: mean BP=170/100 mmHg, mean BPday=170/105 mmHg, mean BPnight=168/98 mmHg, mean HR=65 bpm (Figure 4).

These ABPM results supports the diagnosis of resistant hypertension, that imposed changes of the therapeutic scheme by replacing Amlodipine with Nifedipine-retard, Rilmenidine with Clonidine and by add an additional Carvedilol capsule in the other two. Five days after treatment optimization it was recorded a new ABPM that showed a better blood pressure control (Figure 5). It was also performed a cardiac stress test with the cycle ergometer. The test revealed an symptomatic tension response with the rise of the SBP up to 220 mmHg, which caused the test to be interrupted at 58% from it's course. There were not any symptoms or ischemic changes during the test. Increased blood pressure during exercise and the presence of ascending aortic dilation, requires emphasizing the avoidance of excessive / large physical effort.

DISCUSSION

We present the case of a 52 years old man with severe, refractory hypertension (BP max 220/120 mmHg) diagnosed at a relatively young age (44 years old), associating multiple cardiovascular risk factors (obesity, smoking, dyslipidemia), presenting with symptoms of dyspnoea and headache. We may also consider the association of primary hypertension (but the patient do not declare a family history of hypertension), with „congenital aortic kinking" and evolution in the presence of aortic remodelling hypertension including atheromatosis (radiologically evidenced) and vascular stiffness (Doppler exam - ankle-arm index). Since the first diagnosis of hypertension, the patient followed several treatment regimens, the optimal BP control being obtained only after the association of five anti-

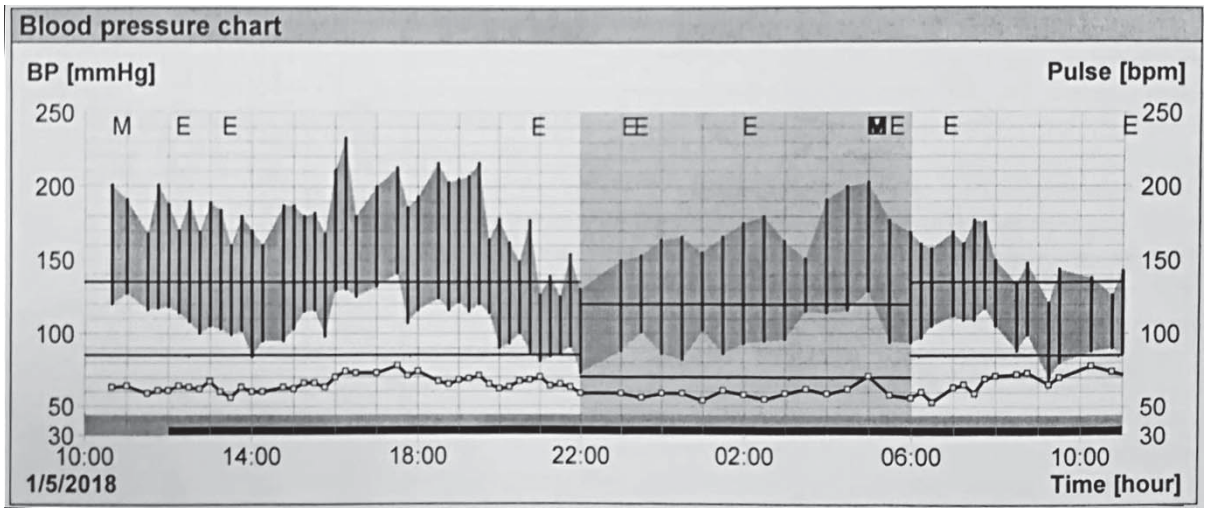

Figure 4. ABPM 3 days after hospital admission.

hypertensives. Given the high BP values resistant to the medication, it was performed an etiologic evaluation that ruled out a secondary hypertension. Despite the attempt to control the high BP with more than three anti-hypertensive drugs (angiotensin receptor blocker, calcium channel blocker, two diuretics, beta-blocker, rilmenidine), the patient maintained the high BP values (170/100 mmg) and a non-dipper profile (BP at day 170/105 mmHg, BP at night 168/98 mmHg). Investigating once again the possibility of a secondary hypertension, on a thoracic radiography we discovered the presence of aortic kinking, confirmed on the CT scan, that raised some new concerns. Literature says that only in a minority of instances resistant hypertension is due to secondary hypertension and that in the absence of a secondary cause the condition is most likely multifactorial wit some proposed mechanisms including genetic factors, aberrant sympathetic nervous system activation and altered renal sodium and water handling due to changes in the renin-angiotensin-aldosterone system etc[7].

Most of the cases cited in literature do not present an association between this aortic anomaly and hypertension, given the lack of pressure gradient (which may be present instead in aortic coarctation)[8]. However we found some similar cases to our situation that raised the same hypothesis[9]. There is also a hypothesis which argues that the pressure gradient may become apparent once the patient with aortic kinking is subjected to physical exercise. We found reported in literature a case of a 6 years old boy with cardiac murmur and tingling sensation of the lower extremiti-

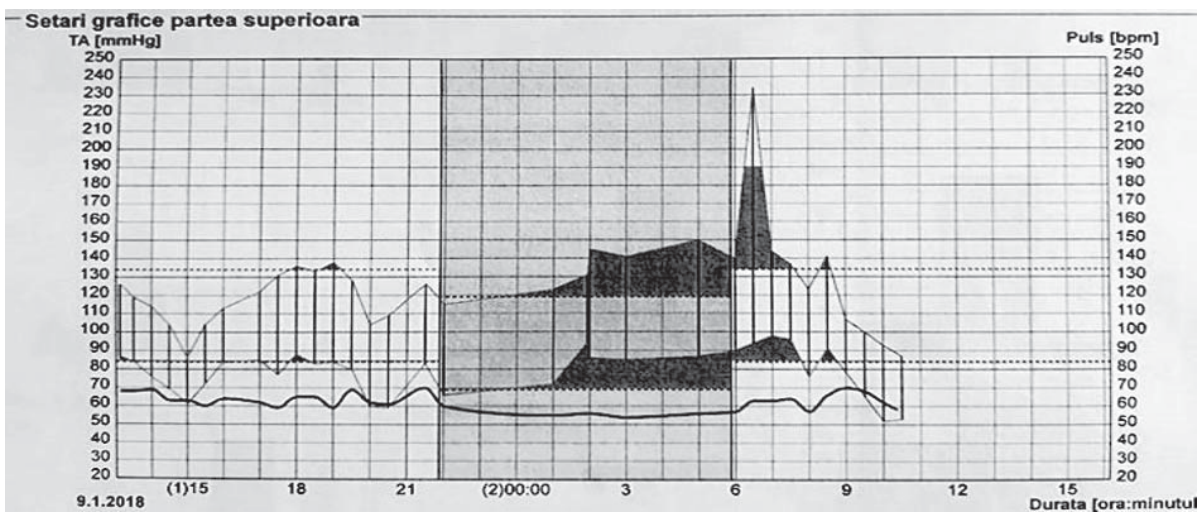

Figure 5. ABPM after treatment optimization.

es on exercise, diagnosed with kinking of aorta. Even though the BP was normal, and there was no significant pressure gradient at rest, the authors concluded that the patient's symptoms were caused by a dynamic aortic narrowing during exercise. The CT scan revealed that the isthmic portion of the descending thoracic aorta was ventral (not adjacent as normal) to the spine, being surrounded by aerated lung, abnormality due to elongated and kinked aortic arch[10]. The unstable pressure gradient in exercise was revealed in another case of pseudocoarctation that progressed to a dissecting aneurysm of the ascending aorta and arch. The article suggested that kinking of aorta may cause hypertension of the upper body (with development of the dissecting aneurysm) during exercise, even though there is no pressure gradient at rest[11]. These observations raise two concerns: the possible causal relationship between aortic kinking and hypertension and the high risk of these patients for dissecting aneurysm, an extremely severe complication. Referring to our case, we observed that the BP values were constantly high, frequently in the morning, no exercise dependent. At the same time, the patient described chest pain suggestive for effort angina pectoris, which may be considered a symptom of pressure gradient presence. In this context, we considered necessary the conduction of an effort test, objectifying the possibility of a pressure gradient. The test showed an exaggerated response with the rise of systolic blood pressure up to 220 mmHg, which imposed discontinuation of the stress test.

One of the most severe complication, relatively frequent is aneurysm developed distal or proximal to the narrowing[12]. Although this abnormaly was considered benign for a long time, most of the studies are showing that kinking of aorta has a high risk for aneurysm formation and dissecting aneurym. There are numerous cases cited in literature that describe this evolution of pseudocoarctation, situations that impose surgical treatment. Although there may not be any etiological relationship between BP and kinking of aorta, the great risk for dissecting aneurysm in the context of an uncontrolled hypertension impose a strict monitorization and treatment. Complications cited for the progressive aortic pseudocoarctation include, in addition to aneurysm dissection, a compressive effect on the mediastinum organs[13]. Our patient presented a refractory hypertension and multiple cardiovascular risks that may explain on one hand the elongated and buckled aorta, raising on the other hand important concerns about a possible severe complications, the need for BP management being imperious.

CONCLUSION

In the case presented, it is difficult to say what the etiological sequence was: kinking of aorta preceded refractory hypertension or the severe hypertension leaded to aortic kinking. Although the aorta kinking is frequently considered a benign patology, being easly overlooked, it is clear, however, in light of literature data, that a patient with this pathological load has a high additional risk that requires strict supervision and treatment.

Conflict of interest: none declared.

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