

## Association of aortic coarctation to a cerebral aneurysm in 53-year-old male with coronaropathy: What should be prioritized?

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### Abstract

Coarctation of the aorta (CoA) is a common congenital defect, which although normally detected and surgically repaired in childhood. Rarely present in adults. Unless significant hypertension exists, adult patients are usually asymptomatic. We report a case of a 53-year-old Man with cardiovascular risk factors, presented with CCS class II angina and Wernicke's aphasia. Further investigation of his aphasia was needed for the evaluation of his hemorrhagic risk under antithrombotic therapy and anticoagulant. These last revealed coarctation of the aorta (CoA) associated to cerebral aneurysm (CA). The patient underwent an angioplasty of his coronary artery disease, then he was presented to surgery for the CA and endovascular repair for the CoA. The issue raised by the case was whether to treat first the CoA or the CA. Although uncommon in adults, aortic coarctation should be included in the differential diagnosis of resistant high blood pressure, since it is associated with significant complications and mortality when untreated as aneurysms.

**Keywords:** Aortic coarctation; Cerebral aneurysm; Coronaropathy; Resistant high blood pressure; Treatment

### 1. Introduction

Coarctation of the aorta (CoA) is a narrowing of the aorta, most commonly diagnosed in childhood. Rarely presented in adults and most are generally asymptomatic. A common presentation of coarctation is systemic arterial hypertension. The narrowing of the aorta raises the upper body blood pressure, causing upper extremity hypertension. Unrepaired coarctation leads to premature coronary artery disease, ventricular dysfunction, aortic aneurysm/dissection, and cerebral vascular disease by the third or fourth decade of life. [1-3].

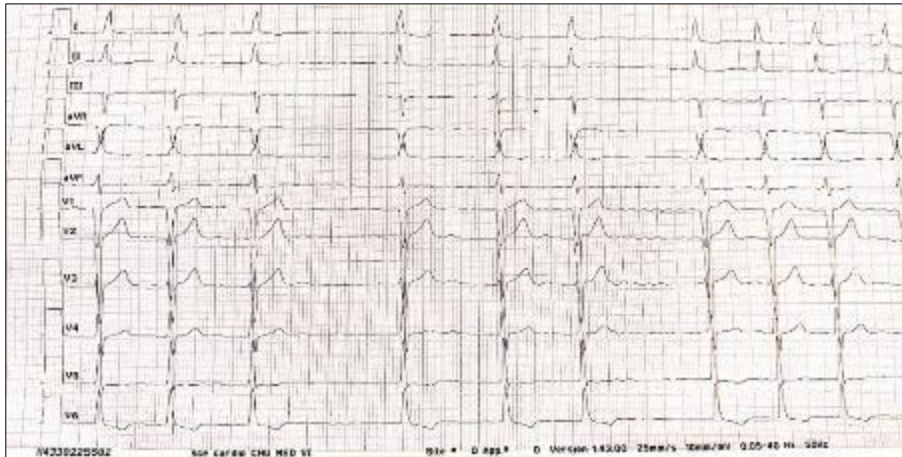
### 2. Case Presentation

A 53-year-old man, presented to the hospital for angina occurring during ordinary activities evolving for about 15 days. His medical history included uncontrolled hypertension, type II diabetes, and abdominal obesity. He was followed for atrial fibrillation for 7 years treated with oral anticoagulation.

Physical examination revealed highly elevated blood pressure at 176/90 mmHg without significant difference between the right and left arms, pulse was 90 beats/min, respiratory rate 16 breaths/min, temperature 37.4°C, and oxygen saturation was 98% on room air. In both lower limbs, peripheral pulses were poorly perceived, blood pressure was remarkably lower compared to upper limbs with decreased Systolic Pressure Index (SPI). In examination of high neurological functions, he presented deterioration of comprehension and memory in favor of Wernicke's aphasia.

Electrocardiogram showed atrial fibrillation and pre-existing left bundle branch block.

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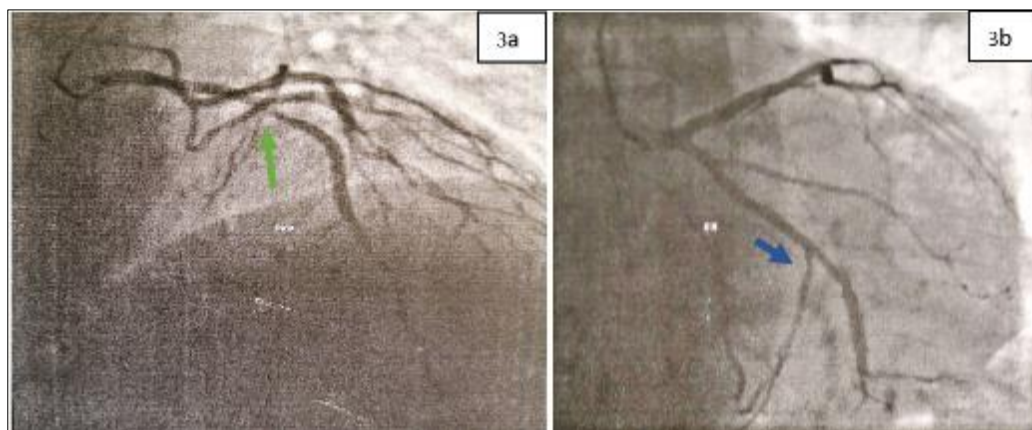
**Figure 1** 12-lead Electrocardiogram showing atrial fibrillation and left bundle branch block

Transthoracic Echocardiography revealed a concentric left ventricle hypertrophy, preserved ejection fraction, no significant mitro-aortic valve disease especially no bicuspid aortic valve and Initial undilated aorta.



**Figure 2** The parasternal long axis view showing the left ventricle hypertrophy (2a).The initial aorta non-dilated (2b)

Due to persistent angina, the patient underwent the coronary angiography that showed a sever stenosis of the left anterior descending artery (LAD) and a plaque of the obtuse marginal (OM) artery. We performed a successful primary angioplasty of LAD artery with active stent implantation.

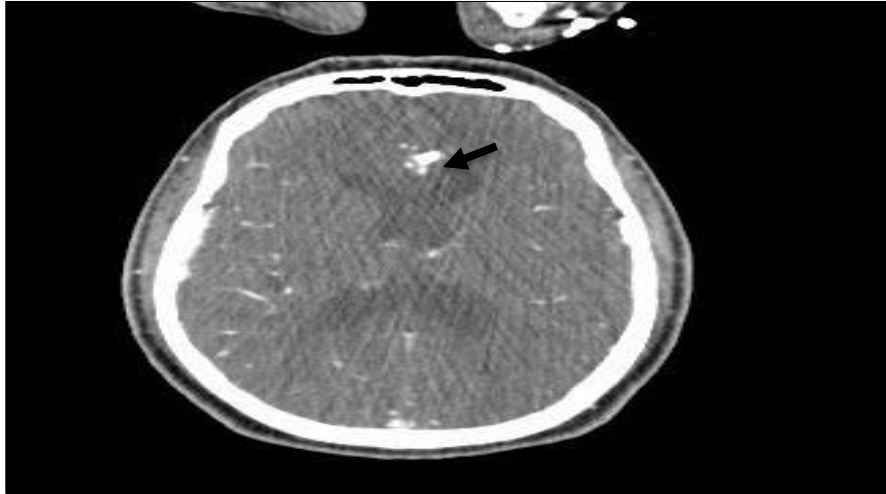


**Figure 3** Left coronary angiogram:

(3a) Left anterior oblique 30°/cranial 30 View revealing a stenosis estimated at 85% of LAD (green arrow)

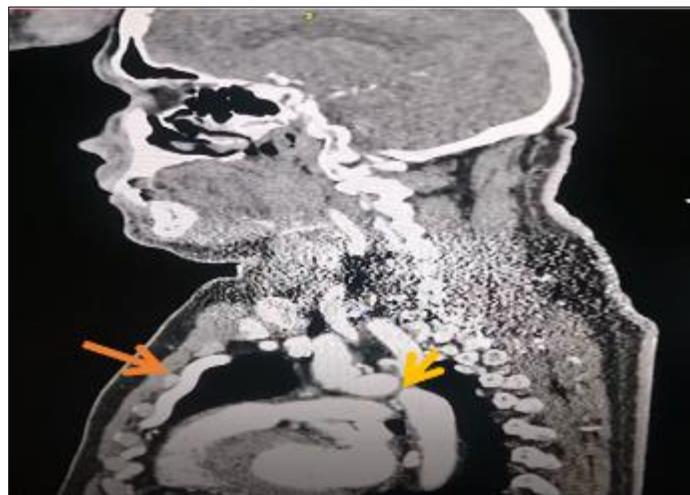
(3b) Left anterior oblique 15°/caudal 25View revealing a plaque estimated at 40% of the OM artery (blue arrow)

Brain Computed Tomography (CT) scan was carried out to look for an organic cause of wernicke's aphasia and to assess the risk of bleeding with the need to put antiplatelet therapy in combination with anticoagulation, and the results showed an ischemic stroke in the middle cerebral artery territory and a saccular aneurysm measuring 9.1× 6.8mm of the left anterior cerebral artery.



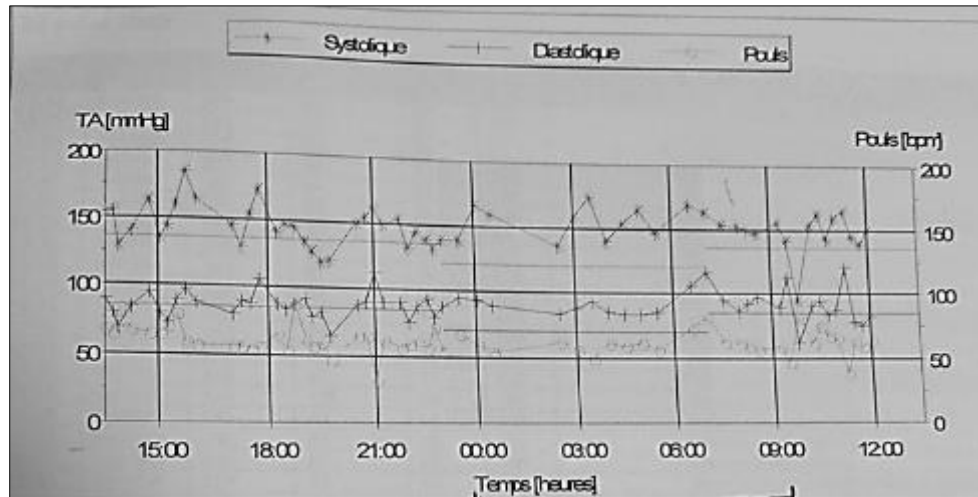
**Figure 4** Axial brain CT scan showing saccular aneurysm of the left anterior cerebral artery (black arrow).

To determine the exact location, size, and shape of the aneurysm, cerebral angiography was realized revealing a blockage in the aortic arch. Therefore, the computed tomography angiography of thoracic aorta and supra-aortic vessels was performed in order to research a malformation or stenosis, which showed a coarctation of the post ductal thoracic aorta associated with significant internal thoracic and peri-spinal collateral circulation. In addition to this a reduced supra- and sub-renal abdominal aorta associated with a large collateral circulation.



**Figure 5** Coarctation of the aorta just at the origin of the left subclavian artery (Yellow arrow )and internal mammary artery dilatation (orange arrow)

24-Hour Ambulatory Blood Pressure Monitoring (ABPM) revealed a systolo-diastolic hypertension diurnal (146/88 mmHg) and nocturnal (158/89mmHg), a reverse-dipper pattern, an important diurnal and nocturnal tensional charge and significant blood pressure variability. setting the diagnosis of severe hypertension with poor prognostic factors that it's consistent with pattern of secondary hypertension in aorta coarctation.



**Figure 6** Graph of blood pressure

Following angioplasty and after discussion of ischemic and hemorrhagic risk, A therapy based on rivaroxaban 15mg, clopidogrel 75mg and Aspirin 75mg was started for one week then rivaroxaban 15mg and clopidogrel 75mg for six months. In addition to this the optimization of anti-ischemic medical treatment and control of cardiovascular risk factors. Then, the patient was treated initially with monotherapy for hypertension. Because of poor blood pressure control, we considered appropriate to switch to a triple combination with ramipril 5 mg and amlodipine 10 mg once a day and bisoprolol 5mg, with normalization of upper arm blood pressure and without deterioration of renal function. Moreover, the patient was referred to neurosurgical department for management of the cerebral aneurysm. Therefore, a mechanical intervention for coarctation of aorta by surgical correction or stent implantation, was proposed after 3 months of angioplasty. However, the patient refused these therapeutic modalities with regular medical follow-up. At the time of this writing, the patient was doing well.

### 2.1. Challenging questions in this case

Two interesting discussions in this patient:

- In diagnosis: what is the challenge of diagnosing coarctation of aorta and cerebral aneurysm in adult patient with coronary artery disease?
- In therapeutic management: what should be prioritized the correction of the Coarctation of the aorta or the surgery of the cerebral aneurysm?

## 3. Discussion

Though most cases involving coarctation of the aorta are detected in children, some patients are diagnosed and undergo repair procedures during adulthood. Signs and symptoms depend on the severity of CoA, which it is mostly detected in the work-up of arterial hypertension [4]. Thus, the coarctation of the aorta is not common or easy to be found in medium-aged population. The diagnosis become more challenging in presence of coronary artery disease with extensive atherosclerosis. In our case, a diabetic patient presented with polyarterial pathology, a coronary artery disease and a cerebral ischemic stroke associating a coarctation of the aorta complicated by a cerebral aneurysm. For this association, Previously, studies had shown that premature coronary artery disease (CAD) is common in patients with COA because they have higher rates of endothelial dysfunction, circulating levels of pro-inflammatory cytokines, and vascular stiffness [5]. Yet, no significant causal link has been found between coarctation and the development of coronaropathy. Then, this risk is attributable to traditional atherosclerotic cardiovascular disease risk similar to the general population independently in the presence or not of the COA [6]. Going back to our patient, he consulted for chest pain. Then, a complete examination was carried out as part of the screening for silent atheromatous disease. In this context, the discovery of low peripheral pulses with low blood pressure in lower limbs were initially attributed to asymptomatic Peripheral arterial disease (PAD). Another point which masked the diagnosis that his antihypertensive treatment was insufficient until his admission. In this case, it was necessary to make measurements under optimal treatment before confirming uncontrolled hypertension. Furthermore, Ambulatory blood pressure measurement (MAPA) was able to specify the diurnal and nocturnal blood pressure profile which was in favor of secondary hypertension. Finally, all pieces

of the puzzle fit together, the coarctation was the cause of his uncontrolled hypertension and of this difference in pulse and blood pressure found in the patient.

Then, It is widely known that unrepaired coarctation of aorta is a significant risk factor for intracranial aneurysm development and this association has been related to arterial hypertension [7]. In this case, patients are thus at risk for both ischemic and hemorrhagic stroke. However, Recommendations to screen for the presence of CA are variable. American Heart Association/American College of Cardiology guidelines recommend Intracranial Aneurysms screening, but appropriate age and interval of screening and its effectiveness remain a critical knowledge gap [8]. In contrast, routine imaging to assess for intracerebral aneurysms in asymptomatic patients with CoA was not recommended in the 2020 ESC Guidelines. It is indicated in the case of symptoms and/or clinical manifestations of aneurysms/rupture [4].

The incidence of CA rupture in a patient with a CoA is high compared to the general population with an isolated CA [12]. Prognosis in aneurysm rupture is dependent on the size of aneurysm, the presence of previous subarachnoid hemorrhage and diagnosis time. A recent multicentric study showed that patients are considered to have a low risk of rupture that have no history of subarachnoid hemorrhage and have aneurysms located in the anterior circulatory area with the diameter lower than 7mm. [13]Owing to present case's age, size and location of the aneurysm, our patient was categorized in the risky group in terms of rupture. Regardless of symptoms, all patients with aortic coarctation who have a non-invasive pressure difference greater than 20 mmHg between the upper and lower extremities or who have hypertension in the upper extremity or significant left ventricular hypertrophy should be treated. The presence of a saccular aneurysm is an indication for treatment on its own, even if sufficient distal collateral circulation exists. Surgical repair in adulthood of CoA is characterized by an increased mortality risk due to degenerative changes in aortic wall, coronary artery disease and end-organ damage due to long-standing hypertension [14].

There is controversy about the proper timing of the surgical treatment of an unruptured intracranial aneurysm or an aorta coarctation. The incidence of intracranial aneurysm rupture as a cause of death in patients with aorta coarctation is higher compared with the estimates of general population. However, in the context of this combination, the CA becomes symptomatic in the majority of patients (93%) while the diagnosis of CoA was made only in a minority of patients (11.6%) [15]. In addition, the literature review found only two deaths secondary to a complication of CoA (aortic dissection) [16,17]. The risk of rupture also exists after surgical repair of aorta coarctation, in the absence of hypertension [11]. These findings suggest that the CA should be operated on first, given the risk of rupture in the presence of an CoA [18].

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#### 4. Conclusion

To conclude, the diagnosis of aortic coarctation must be included in an adult patient with resistant high blood pressure. The clinical signs of a difference in pulse and blood pressure between the upper and lower extremities can be recognized clinically, although atheromatous etiology is the most frequent, especially in the presence of cardiovascular risk factors. Therefore, a meticulous cardiovascular examination is recommended in all patients with hypertension, including palpation of all pulses and blood pressure measurements of both the upper and lower extremities. However, brain imaging in patients with coarctation of the aorta seems reasonable for the detection of a cerebral aneurysm, especially in adults with long uncontrolled hypertension. These patients must be taken care of quickly by specialists given the risk of fatal complications.

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#### Compliance with ethical standards

##### *Disclosure of conflict of interest*

No conflict of interest to be disclosed.

##### *Statement of informed consent*

Informed consent was obtained from all individual participants included in the study.

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