


MIDDLE AORTIC SYNDROME IN AN ELDERLY PATIENT: DESCRIPTION OF A SURGICAL CASE

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ABSTRACT

The middle aortic syndrome is a rare form of presentation of aortic coarctation that represents 0.5-2% of this same entity. It usually presents in the pediatric or young adult population; therefore, the age of this patient is of interest. It mainly affects women with an estimated ratio of 4:1. Renal involvement is the most prevalent, and idiopathic etiology is the most frequent.

This pathology presents clinically with hypertension challenging to control along with intermittent claudication. This article comprises the case of an 81-year-old female patient with a history of hypertension, sedentary lifestyle, smoking, osteoporosis, cholecystectomy in 2010, and appendectomy in childhood. On this occasion, the patient had a stenosis of the infrarenal abdominal aorta and a simultaneous superficial femoral artery lesion and was treated in a hybrid approach with a balloon expandable stent and a suprapatellar femoral-popliteal bypass.

Keywords: *aortic coarctation, intermittent claudication, aorta, hypertension, aortic stenosis*

INTRODUCTION

The term "medial aortic" syndrome was first described in 1947 by Quain and colleagues¹, who observed this pathology in a pediatric age group with an average age of 9 years. The medial aortic syndrome is the name we use to denote infra-diaphragmatic aortic stenosis². This entity represents 0.5 to 2% of all aortic coarctation¹ and mainly affects the pediatric or young adult population with a higher prevalence in women³ (ratio of 4 to 1 for men⁴).

These stenoses occur in different locations that have a relevant influence on the clinical manifestations of the subject depending on the affected arterial territory. In most cases, the involvement of the renal artery is observed, reaching up to 70%⁵ (within this percentage, both renal arteries are involved two-thirds of the time). The most frequently described affected renal areas (69% approximately) are the suprarenal; secondly, the intrarenal location (23% approximately); and, last but not least, there is the infrarenal location (8%), as in the patient described⁵.

When the renal artery is involved, it generally manifests as hypertension that is difficult to control pharmacologically⁶, i.e., it cannot be controlled by using up to 3 antihypertensive drugs at maximum doses. In these cases, the objective would be to reduce blood pressure to below the 95% percentile in the pediatric population or, if there is left ventricular hypertrophy or target organ damage, to reach the 90% percentile²; that

is, maintaining systolic pressures below 120 mmHg and diastolic pressures below 80 mmHg in adults. If left untreated, this hypertension could lead to left ventricular hypertrophy, resulting in heart failure, a common cause of mortality in subjects with this pathology⁷. When the compromised artery is the superior mesenteric artery or the celiac trunk, this may be reflected as symptoms of intestinal ischemia with its suggestive periumbilical post-eating pain and weight loss due to food rejection. Recall that the patient, apart from these signs and symptoms, may also experience intermittent claudication, which will be expressed by abdominal murmurs and the possible absence of femoral pulses⁸.

According to the study of Rawan and collaborators, its etiologies are multiple. However, idiopathic etiology is the most frequent among all varieties, with almost two-thirds of all cases⁵. It is followed by the inflammatory cause evidenced by Takayasu as the most relevant, and within the genetic origins, we can highlight neurofibromatosis type 1, among others. With a lower percentage of the total, other causes such as tuberculosis, syphilis, rubella, radiotherapy, or fibrosis are evidenced, to mention a few⁹ (Table 1).

Diffuse congenital hypoplasias such as those seen in Noonan³, Williams, Alagille, and tuberous sclerosis syndromes, some of them could have their embryological origin due to the overfusion of the two dorsal aortas in the fourth week of development, generating aortic hypoplasia².

Etiology	Percentage (%)
Idiopathic	67
Inflammatory	17
Genetic	15
Others (syphilis, tuberculosis, rubella, radiotherapy, fibrosis, etc.)	3

TABLE 1. Distribution of etiologies⁵.

Knowing the origin of the medial aortic syndrome is essential because the abdominal aorta may not be the only vessel affected. For example, neurofibromatosis type 1 may be associated with aneurysms and cerebral arterio-venous fistulas⁵. In rubella, it may co-occur with aortic valve stenotic pathology. Takayasu's vasculitis mainly involves the aortic arch and descending aorta, among many other associations⁸.

The medial aortic syndrome diagnosis can be made by renal Doppler, CT, or MRI. Renal Doppler is the first line, with a sensitivity of 90% and a specificity

of 68%. Despite its low specificity, this modality is indicated to reduce exposure to lightning and the use of contrast. As an advantage, MRI allows us to estimate pre- and post-stenosis flows, but if we wish to evaluate the lesion and its characteristics, then angiography is the indicated method. We should consider a tomography or an encephalic resonance if we suspect brain lesions.

It is recommended as initial therapy to start with a single antihypertensive agent and reach the maximum dose or adverse effect before adding another drug. In principle, enalapril or losartan

should be started, and diuretics should preferably be used as combination therapy. Surgical treatment is evaluated when elevated pressures persist and several hypertensive drugs cannot control it. If the lesion presents as isolated stenosis, it can be treated with an isolated patch or a bypass that skips it if the coarctation is very narrow¹⁰. The same applies to thoracoabdominal stenosis, which can be treated with a bypass connecting the thoracic and abdominal cavities. Polytetrafluoroethylene (PTFE) patches are preferred over Dacron woven or knitted.

Renal or splanchnic reconstruction will be performed as appropriate. For renal reconstruction, reimplantation away from the diseased aorta is recommended. Homologous arterial grafts are generally preferred, but venous grafts are not recommended due to the risk of aneurysm formation. When the renal artery's stenotic lesions are multiple, or this same kidney is found to be hypotrophic, i.e., 2 to 3 cm smaller than its size and non-functioning, nephrectomy may be indicated¹⁰. Remember that nephrectomy is only performed if the other kidney can supply the function. An alternative to consider is renal autotransplantation which improves hypertension and renal function. Splenic artery repair

can be performed with a prosthesis or implantation of venous material (*Figure 1*).

This article aims to describe a surgical case that occurred in June 2022 at the Hospital General de Agudos Juan A. Fernández. The advanced age of the patient makes this case very interesting since there is not much bibliography describing the exposed problem. Usually, cases of pediatric patients or patients younger than 25 years old presenting with significant symptomatology are reviewed.

METHODS

We review the case of an 81-year-old female patient with multiple clinical comorbidities, including hypertension, a sedentary lifestyle, smoking, and osteoporosis. Her surgical history included an appendectomy in childhood and a laparoscopic cholecystectomy in 2010. Imaging showed an infrarenal abdominal aortic stenosis with a concomitant stenotic lesion in the right femoral artery, both lesions diagnosed by arterial angiography (*Figure 2*). On physical examination of both lower limbs, she had both femoral pulses present with detection of left popliteal pulse, and the rest of the examination was complete with positive flow Doppler signals for the other landmarks (*Figure 3*).



FIGURE 1. 3D angiotomographic reconstruction image prior to surgery.

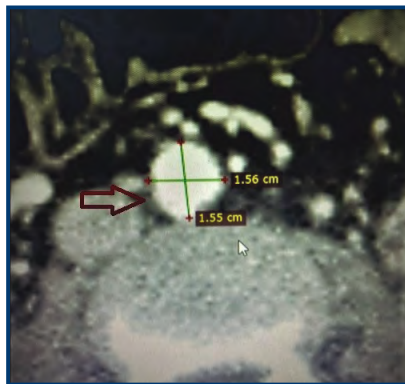


FIGURE 2. Angiotomographic image showing diameters prior to surgery.

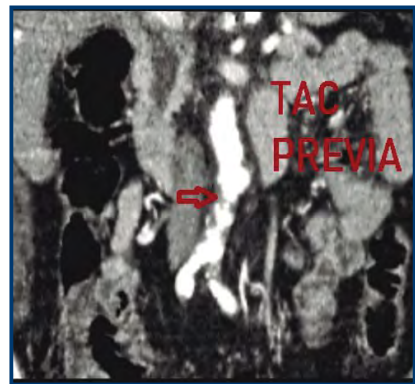


FIGURE 3. Angiotomographic image coronal view prior to surgery.

RESULTS

For this case, it was decided to hold a meeting in June 2022, where it was concluded that the hybrid approach would be the best option given the patient's age and comorbidities. A 43 mm AndraStent™ XL balloon expandable straight stent was placed through an 8F suitable femoral introducer for its resolution. The proximal anastomosis was performed using the femoral approach and exposure by constructing a suprapatellar femoral-popliteal bypass with an 8 mm PTFE prosthesis.

Immediate postoperative examination showed the presence of all pulses with favorable changes in color

and temperature in both limbs. As part of the hospital protocol, typical of the institution, the patient spent the first postoperative day in a closed unit. After 72 hours, given the favorable evolution, hospital discharge was determined to continue ambulatory follow-up by outpatient clinics (*Figure 4*).

If we look at the statistics of open surgery, we see positive results concerning the treatment of hypertension in approximately 9 out of 10 cases. This type of surgery has low morbimortality in patients with good general conditions. Despite presenting good results, the reoperation rate for new stenosis is almost 10% 5-12 years after the first procedure⁵.

The endovascular approach generates a more significant number of morbidities over time, such as dissections, leaks, aneurysms, or thrombosis of the stent placed. Therefore, it requires reinterventions and may cause restenosis. Studies mention that 55% of patients are free of interventions in one year and 33% in 5 years¹¹. Endoluminal interventions have better

results in young adults with fibromuscular dysplasia. Therefore, one should select suitable patients for endovascular procedures, especially those cases where small vessels and fibrotic are evident. Stenting after angioplasty should be considered because of the risk of neointimal hyperplasia or stenosis after the initial procedure.

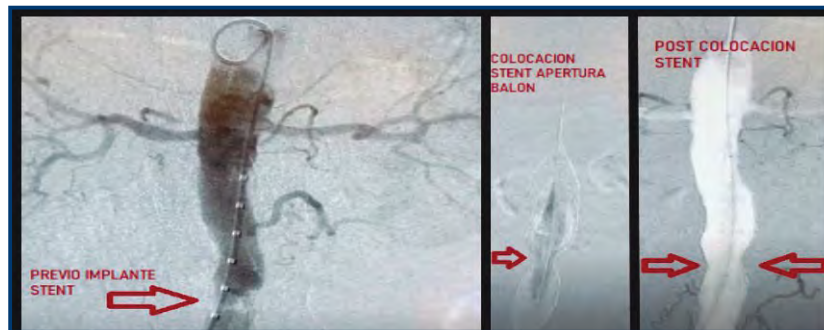


FIGURE 4. Series of angiographic images of endovascular resolution.

CONCLUSION

In conclusion, for all the data presented and the favorable outcome demonstrated by the patient, a hybrid approach is the best way to resolve this type of pathology with the same lesions. On this occasion, the patient's characteristics, age, comorbidities, and availability of resources were evaluated. The hybrid approach would be the best option because it improves surgical times, and the patient's postoperative period is less aggressive. Outpatient follow-up is performed with imaging follow-up using contrasted tomography at one month and then every six months or yearly, depending on renal function.

DISCUSSION

We want to raise the question and consider which is better: an endovascular or an open approach.

Declarations

The authors declare no conflict of interest.

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