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Case Report

Management of Annulo-Ectasitic Disease during Pregnancy

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Abstract

Annulo-ectasitic disease is a group of disorders with similar phenotypic features, combining aneurysm of the aortic duct and dilatation of the aortic annulus. Its incidence is estimated at 4.5 cases per 100,000. It is the 13th leading cause of death in Western countries. Aneurysms of the ascending aorta are divided into two distinct entities according to etiology and surgical management. Pregnancy and peri-partum are two situations where the risk of aortic dissection is higher, with a more rapid progression of aortic diameters. This risk is directly related to the diameter of the aorta at the beginning of pregnancy, compliance with beta-blocker treatment, and the need for concerted cardiological and obstetric monitoring as soon as the desire for pregnancy is expressed and up to the 2nd month of the peripartum period. The aim of this review is to illustrate, through 02 clinical situations, the difficulties in managing parturient patients suffering from the different types of annuloectasitic disease of the aorta.

Keywords: Ascending aortic aneurysm; pregnancy; aortic dissection; Marfan syndrome; bicuspidy; beta-blockers.

INTRODUCTION

Ascending aortic aneurysm is a localized dilatation with loss of parallelism of the walls of the aorta from it's origin to the emergence of the supra-aortic arteries. This condition is rare in pregnant women and is often associated with an adverse outcome [1, 2]. The hormonal and hemodynamic changes associated with pregnancy, particularly those that occur during labour and the post-partum period, increase the risk of progressive dilatation or dissection of the aorta in these women [1]. This risk increases in the presence of hypertension. We report here 02 cases of ascending aortic aneurysm that had a favorable outcome during pregnancy.

Clinical case N°01

Mrs. M.A, 42 years old, first-born, was admitted to our department for treatment of an annulo-ectasitic disease with aneurysmal dilatation of the ascending aorta associated with severe aortic insufficiency discovered at 31 weeks' gestation + 3 days. The diagnosis of Marfan syndrome was based on the marfanoid morphotype, a history of childhood scoliosis treated by corset, glaucoma with myopia, ligament hyperlaxity and joint hypermobility (Fig.1). The patient was hemodynamically stable, in functional class II of the NYHA. Physical examination revealed a 4/6ths diastolic murmur at the aortic site, beginning immediately after B2 and radiating downwards to the xiphoid process. The pulses were bouncing with a widening of the blood pressure differential to 120/40mmHg. The echocardiography showed an initial aorta dilated to 66 mm, a moderately dilated Left ventricle (LV) with an LVEF of 55% with severe Aortic regurgitation (AR) (Fig.2-4). Gynecological examination revealed a viable eutrophic fetus (1400 g) with normal umbilical Doppler. In view of the very high maternal-fetal risk (class IV according to the modified WHO), a medical-surgical staff meeting was held, and the decision was taken to proceed with fetal extraction by caesarean section at 32 weeks' gestation, preceded by a course of corticosteroids, under strict control of blood pressure and heart rate. At 32 weeks' gestation, the planned delivery took place without any major incidents or accidents. On the 2nd post-partum day, the patient was transferred to a cardiac surgery center where she underwent a Bentall procedure with a good operative outcome.

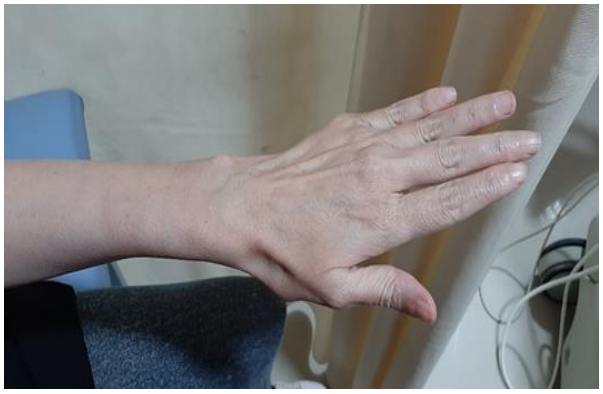

Figure 1: Articular hypermobility and ligament hyperlaxity in Marfan syndrome

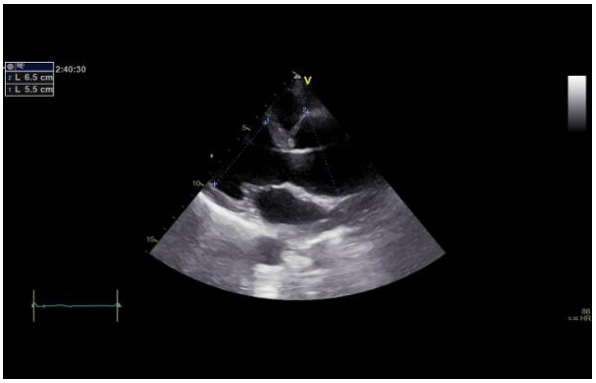

Figure 2: Initial aortic aneurysm on 2D TTE

Clinical case N°02

Mrs S.H 32 years old, nullipare G1P0 with a history of Marfan's disease, complicated by an aneurysm of the initial aorta with an important aortic regurgitation (AR) for which the patient underwent an aortic valve replacement by mechanical prosthesis St Jude N°23 with placement of an aortic tube in position supra coronary N°28 in 2020, without follow-up since. In March 2022, the patient was consulted for chest pain that had been evolving for more than a month in a progressive pregnancy of 33 SA+1 day. Clinical examination: BP: 130/80 mmHg, HR: 75 bpm (on bisoprolol+ Nicardipine), no anisotension. The Electrocardiogram (ECG) was without particularity. Echocardiography, complemented by transesophageal echocardiography, revealed an initial aorta dilated to 56 mm at sinus level with an intimal flap and a double-channel appearance (Fig. 3) extending to the descending aorta and the carotid arteries, associated with significant Aortic regurgitation (Fig 4). The diagnosis of De Bakey type I subacute aortic dissection (pain for one-month, hemodynamic tolerance) was confirmed by angioscan.

After a multidisciplinary discussion involving the cardiologist, gynecologist and pediatrician, the decision was made to perform a vaginal delivery with tubal ligation, which was carried out without maternal or fetal incident.) The patient was transferred to a cardiac surgery center where she underwent a Bentall operation with a good clinical outcome.

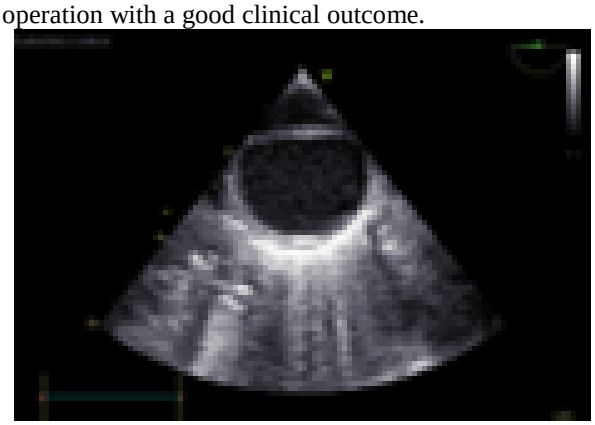

Figure 3: TEE image shows dilatation of the aorta, an intimal flap and a double-channel appearance of the aorta

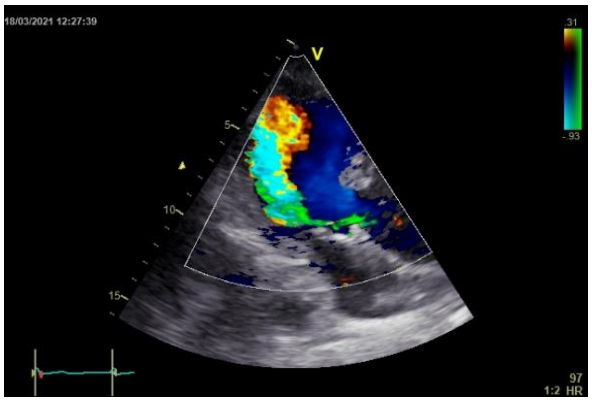

Figure 4: Colour Doppler showing significant AR

DISCUSSION

Pregnancy is a high-risk period for all patients with aortic disease, which is rare in pregnancy but associated with very high mortality [3]. Most deaths occur in women not previously identified as having aorta disease. Pregnancy in women with this condition presents a challenge for both clinician and patient due to the increased incidence of maternal-fetal complications, and therefore deserves special attention. Hemodynamic and hormonal changes during pregnancy increase susceptibility to dissection. Dissection most often occurs in the last trimester of pregnancy (50%) or early post-partum (33%) [4]. Hypertension, advanced maternal age, connective tissue diseases and aortic malformations are the main risk factors: Marfan syndrome, Ehlers Danlos type IV syndrome, aortic coarctation and bicuspidia [4-6]. Marfan syndrome is an autosomal dominant genetic disorder that causes a reduction in the strength of supporting tissues. Pregnancy increases this risk [7, 8]. In both cases, Marfan syndrome was the cause of the aneurysm. The effect of pregnancy on aortic dilatation is unclear [4]. The overall risk of a woman with Marfan syndrome having a pregnancy-associated aortic dissection is approximately 3%. Aortic size is a major determinant of risk, but even women with an aortic root <40mm have a

1% risk of dissection [4]. Chest pain is the most consistent finding in the literature. The diagnosis of aortic dissection should be considered in all patients presenting with chest pain during pregnancy [4, 5]. The imaging tests that should be ordered in cases of suspected aneurysm of the ascending aorta in a pregnant woman, or in cases of dissection, are: transthoracic echocardiography, transoesophageal echocardiography, angioscanner +/- MRI [4-6, 8].

Dissection during pregnancy can be treated either surgically or medically:

- Stanford A aortic dissection during pregnancy is a surgical emergency [4-6]. Experienced cardiothoracic physicians, cardiologists, obstetricians and cardio-anaesthetists must act rapidly to deliver the fetus (if viable). Conservative treatment is associated with a mortality rate of up to 80% in the first week after diagnosis [4, 5]. Before 28 weeks' gestation, aortic repair is performed without fetal extraction. The cardiovascular surgical mortality rate is between 2 and 6% for the mother and between 20 and 30% for the fetus. After 32 weeks' gestation, caesarean section is the first option, followed by aortic repair. Surgical repair consists of aortic replacement [5].
- In the case of type B dissections, medical treatment in an intensive care unit result in a 1-month survival rate of around 90% [5]. Endovascular treatment is taking on its rightful place in the management of type B dissections not stabilised by medical treatment alone. Promising medium-term results have been reported [4, 5]. However, the results of endovascular repair of the thoracic aorta during pregnancy have only been described in a few cases, and it is not recommended in cases of genetic aortopathy [4].

Subsequent pregnancies: Follow-up should take account of the main risks involved, namely aortic rupture, recurrence of dissection and refractory hypertension. Some studies recommend repeat imaging at 1, 3, 6 and 12 months [4]. Patients with Marfan syndrome or a related syndrome require early management, as soon as they wish to become pregnant [6, 9]. There are no more complications during pregnancy in patients with Marfan syndrome without cardiovascular repercussions, and with an aortic diameter of less than 4 cm. However, it is not only the aortic diameter that is important, but also the rate of dilation [5, 8, 9]. In patients with vascular Elhers Danlos syndrome, background treatment with celiprolol should be instituted [5].

The European Society of Cardiology (ESC 2018) guidelines on the management of aortic disease in pregnancy suggest the following [4]:

- When a woman with known aortic dilatation, aortic dissection or a history of aortic dissection or a genetic predisposition for aortic dissection becomes pregnant, strict BP control is recommended (I, C).
- Pregnancy is not recommended in women with aortic dissection or a history of aortic dissection (III, C).
- Whenever possible, the use of ergometrine is not recommended in women with aortic disease (III, C).
- When the diameter of the ascending aorta is > 45 mm, a caesarean section should be considered (IIa, C).
- In the case of aortic dissection or a history of aortic dissection, a caesarean section should be considered (IIa, C).
- Prophylactic surgery should be considered during pregnancy if the diameter of the aorta is > 45 mm and increasing rapidly (IIa, C).
- When the fetus is viable, delivery before any necessary cardiac surgery should be considered (IIa, C).
- Pregnancy is not recommended in women with severe aortic dilatation (hereditary thoracic aortic disease such as Marfan's syndrome: > 45 mm; aortic bicuspidism: > 50 mm or > 27 mm/m² body surface area, Turner's syndrome: > 25 mm/m² body surface area) (III, C).

CONCLUSION

Annulo-ectasitic disease groups together certain aetiologies responsible for aneurysmal dilation of the aorta, particularly in its initial part, such as Marfan's syndrome. Pregnancy and these physiological changes can increase the progression of the aortic diameter, leading to fragility of the wall, which can be responsible for aortic dissection. This clinical situation endangers the Maternal-fetal prognosis, and its management requires a multidisciplinary team to decide on the type and timing of intervention for the mother and fetus, based on international recommendations, according to the age of pregnancy, the viability of the fetus and the presence or absence of complications.

Disclosure: No conflicts of interest to declare.

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