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Pregnancy Follow-up and Outcome in a Patient with Takayasu's Arteritis

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We describe here the pregnancy follow-up and outcome in a patient with Takayasu's arteritis, with a detailed account of the complications during gestation and delivery and the impact of the disease on the newborn's health.

Introduction

Takayasu's arteritis (TA) is a chronic inflammatory vascular disease of unknown origin. It often affects the aortic arch and its main branches, although it can affect other segments of the aorta as well as pulmonary and renal arteries^{1,2}.

This arteritis can result in systemic arterial hypertension (SAH), cerebral vascular accident (CVA), aortic failure (AF), renal failure (RF), intermittent claudication (IC) or congestive heart failure (CHF)³.

Women are more often affected than men, at a proportion of 4:1 and its incidence is higher in the second and third decades of life. The diagnosis of TA in pregnant women is rare; however, when it affects pregnant women, hypertension, usually of the renovascular type, can be observed in more than 50% of the cases and determines higher maternal-fetal risk⁴.

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The patient, R.S.S., was a 17-year-old black female who had been hypertensive since 12 years of age. She had an echocardiographic diagnosis of aorta coarctation and was pregnant for third time. The first pregnancy had ended in miscarriage at six weeks of gestation, four years before, and the second pregnancy had resulted in premature birth and fetal death at seven months of gestation, two years before.

The patient was admitted at our Institution on March 2006, complaining of cephalea, dyspnea at moderate physical exertion, palpitations and intermittent claudication. She was at the second trimester of her third pregnancy (24 weeks) and presented high blood pressure levels in the upper

Key words

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limbs – systolic pressure (SAP) = 180 mmHg and diastolic pressure (DAP) = 110 mmHg – and decreased BP levels in the lower limbs – SAP = 110 mmHg and DAP = 60 mmHg.

An aortography was carried out to define the procedure for the treatment of aorta coarctation, which showed a narrowing in the distal 2/3 of the descending thoracic aorta, with parietal irregularities, presence of small- and medium-caliber collateral circulation and important obstruction in the distal extremity of this segment (proximal diameter =13.9 mm x distal diameter =2.5 mm). The angiographic aspect was suggestive of TA and a balloon-catheter dilation was carried out at the area with the highest stenosis (i.e., the smallest luminal area).

At the end of the procedure, there was an increase from 2.5 mm to 7.5 mm in the diameter of the area with the highest stenosis and the maximum aortic gradient decreased from 112 mmHg to 61 mmHg (decrease of 51 mmHg).

A Doppler ultrasonography (USG), performed during the hospital stay, showed the involvement of the proximal third of the left renal artery and a normal uteroplacental flow.

The patient remained hypertensive (SAP=160 mmHg and DAP=90 mmHg) although she received propranolol (60 mg/day), nifedipine (60 mg/day) and prednisone (30 mg/day) in association with alpha-methyldopa (1.5 g/day), which the patient used, with a slight improvement in SAH (150 x 90 mmHg). The patient presented improvement of symptoms and was discharged.

At the 36^{th} week of gestation, the patient was admitted to the hospital again, complaining of cephalea and SAH (170 x 100 mmHg). A new Doppler USG showed symmetrical intrauterine growth retardation, with an estimated fetal weight of 1,935 g (expected fetal weight: 2,813 g for the gestational age) and moderate increase in the uteroplacental flow resistance.

The obstetric medical team decided for the pregnancy resolution when the patient went into labor, due to the arterial hypertension peaks of 230x120 mmHg during uterine contractions, leading to acute fetal distress.

The delivery by Caesarean section was performed, with no maternal or fetal adverse events and maternal BP monitoring, with SAP remaining between 150 and 180 mmHg. The infant's birth weight was 1,990 g, with 1st-minute and 5th-minute Apgar scores of 9 and 9, respectively. The newborn received rooming-in care, remaining in the same hospital room as the mother and both were discharged five days after the delivery. The patient received the same medications throughout the postpartum period, which were gradually decreased at the outpatient clinic 15 days after the delivery.

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The patient is currently still hypertensive and uses captopril (75 mg/day) and hydrochlorothiazide (25 mg/day).

Discussion

Takayasu's arteritis has a worldwide distribution, with the highest incidences being found in Japan, East and South Asia and India¹. Its etiology is still poorly understood and genetic factors as well as autoimmunity with the presence of histocompatibility antigens, particularly HLA A9, A10, B5, BW40, BW52, DQW1, DR2, DR4, DR7, DW3 and DW12, are the main postulated causes. The presence of these antigens is associated with a worse prognosis².

Infections have also been implicated in the pathogenesis of the disease, especially tuberculosis, which as a high prevalence in this population (20 to 25%) and can function as an immunological trigger or as a chronic antigenic stimulation of the disease².

The clinical manifestations range from asymptomatic patients with pulse alterations only, to a large neurological involvement such as hemorrhagic cerebral vascular accident and are due to the obstructive process of the aortic arch and its main branches⁵.

Ishikawa et al⁶, in a study of 33 pregnancies and 50 more cases from the literature, described complications in the pregnancy-puerperium cycle in 61.4% of the cases and 4.8% of maternal death; the latter were more frequent in the 3rd trimester and delivery (65%) and in the postpartum period (37%). The main complications were CHF, SAH and stroke, with 4 of them being the hemorrhagic type.

According to the Brazilian consensus on cardiopathy and pregnancy⁷, the fetal risks include intrauterine growth retardation (IUGR) in 18% and intrauterine fetal death in 2-5%. The patient had had a miscarriage and a fetal death, which is in accordance with the literature and emphasizes the necessary care for these patients.

Hung et al⁸ reported a successful pregnancy in a 23-yearold woman submitted to a bypass surgery. The drugs used by the patient were similar to the ones used in our institution and the delivery was postponed until the 33rd week of gestation. In this case, the patient also underwent a caesarean section, due to the severity of the condition. No cases of percutaneous treatment during pregnancy were found in the literature.

The percutaneous aortoplasty was chosen due to the small risk of radiation for the fetus in the second trimester, with adequate abdominal protection, as well as the decreased mortality when compared to the conventional surgery $(30\% \times 5\%)^9$.

The decision to perform a percutaneous aortoplasty was made by the hemodynamic team, due to the severity of the stenosis and the important clinical manifestation of the disease. There was an important improvement in the stenosis and distal flow, resulting in the attenuation of the SAH, probably maintained by the renovascular involvement. However, it allowed, together with the anti-hypertensive medication, the maintenance of the pregnancy up to the 36th week of gestation.

The medications used were alpha-methyldopa, which is harmless in pregnancy (Class A), propranolol, which has a dose-dependent effect and is safe up to 60 mg/day; higher doses can cause IUGR and premature birth (Class B) and nifedipine, which up to 60mg/day is also considered Class B⁷. As the patient presented increased blood sedimentation rate (100 mm), probably reflecting the disease activity, she was given corticosteroids at a dose of 30 mg/day. Pregnancy does not interfere with the evolution of Takayasu's arteritis^{1-5, 8, 10}.

The delivery by Caesarean section was a decision made by the obstetric team, considering the high blood pressure levels, which at the start of labor reached $230 \times 120 \text{ mmHg}$.

Conclusion

Although it is a potentially severe pathology during pregnancy, with a maternal mortality of 4-5%² (against 130/100,000 in the general population), a successful pregnancy in TA is possible if followed adequately, aiming at the control of SAH, which is often observed in these patients. Although the management is eminently clinical, the decision to intervene and the ideal moment to do it can determine a better evolution and higher safety for the pregnancy. In the present case, we believe that the intervention that allowed the pregnancy to reach the 36th week was crucial, considering that the two previous gestations had been unsuccessful.

Potential Conflict of Interest

No potential conflict of interest relevant to this article was reported.

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Study Association

This study is not associated with any graduation program.

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