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(CASE REPORT)

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Challenges in the management of pulmonary hypertension manifested during pregnancy: Case report

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Abstract

Pulmonary hypertension (PH) during pregnancy is a highly complex clinical entity. It is estimated to affect 1.1 out of every 100,000 pregnancies, generating a high maternal mortality which is close to 50% according to the severity of presentation; however, with appropriate treatment, mortality can be reduced by 15-30%. This paper presents the case of a female patient in the second decade of life with severe PH, initially attributed to congenital heart disease, and later associated with systemic lupus erythematosus (SLE). The clinical evolution required escalation of pharmacological therapy to triple therapy, achieving improvement in quality of life and functional capacity. Diagnostic challenges during pregnancy, the need for multidisciplinary approaches, and the importance of adequate risk stratification are discussed. Current guidelines recommend initial triple therapy in patients with high-risk PAH, supported by evidence demonstrating its impact on survival. This case underscores the importance of early intervention, individualized management, and cross-specialty coordination to optimize clinical outcomes in the high-risk setting

Keywords: Pulmonary hypertension; Pregnancy; Pulmonary Arterial Hypertension; Echocardiogram

1. Introduction

Pulmonary hypertension (PH) is a clinical condition characterized by elevated mean pulmonary arterial pressure (mPAP). It is classified into 5 clinical groups, according to pathophysiological mechanisms, clinical presentation and hemodynamic characteristics (1). Group 1 corresponds to pulmonary arterial hypertension (PAH), a specific form of PH characterized by remodeling of the pulmonary arteries, leading to a progressive increase in pulmonary vascular resistance (2,3). PAH is a rare disorder that affects between 15 and 50 persons per million population in the United States and Europe, has a female predominance, and its age of presentation varies between 30 and 60 years. However, it can occur in men and is often associated with worse clinical outcomes (4).

PH may initially manifest as nonspecific symptoms such as exertional dyspnea and fatigue. In undiagnosed pregnant women, the nonspecific symptoms in early stages may be confused with those related to physiological changes during normal gestation, presenting with some degree of dyspnea, fatigue and lower extremity edema, which contributes to a diagnostic delay in this population. PH during pregnancy has a high morbidity and mortality, with associated mortality rates reaching between 25% and 56%, which is one of the relevant points in this case (5). Pregnancy is considered totally contraindicated in patients with this pathology, since it can become lethal or exacerbate the disease. The pregnant patient with this condition constitutes a major challenge, given the severity of the pre-existing cardiopulmonary disease together with the changes inherent to pregnancy. The seventh world symposium on PH

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suggested that high-risk patients be treated with triple therapy; however, there are no current guidelines for standard targeted therapy in pregnancy (1,5).

2. Case

A 36-year-old female, with a history of atrial septal defect (ASD) who, at 23 years of age during the course of her first pregnancy, presented dyspnea on medium exertion associated with fatigue, so a transesophageal echocardiogram was performed with a finding of dilated right atrium (23 cm2), tricuspid insufficiency with moderate to severe regurgitation jet and enlarged right ventricle, considering a high probability of pulmonary hypertension. In consensus with the patient, it was decided to continue pregnancy, and due to the progression of dyspnea, induced delivery by cesarean section was performed at 32 weeks. Subsequently, a right heart catheterization was performed with a finding of severe pulmonary hypertension with negative vasoreactivity test; pulmonary arterial hypertension was found to be related to congenital heart disease, and management with phosphodiesterase 5 inhibitor (sildenafil) and synthetic prostacyclin analogue (iloprost) was initiated.

At 2 years of age, taking into account the maternal history of systemic lupus erythematosus (SLE), the screening for connective tissue disease was continued and the diagnosis of SLE was confirmed and immunomodulatory management was initiated.

Over time, his functional class deteriorated, requiring supplemental oxygen, so vasodilator therapy was modified, suspending sildenafil and initiating guanylate cyclase stimulator (riociguat) together with an endothelin-1 receptor antagonist (ambrisentan).

The evolutionary course is progressive, deteriorating the quality of life and presenting oxygenation deterioration. Hemodynamic stratification was performed with findings of mean pulmonary artery pressure of 75 mmHg with pulmonary vascular resistance of 18 Wood units, pulmonary capillary pressure 6 mmHg. Cardiac resonance was performed and reported ostium secundum atrial septal defect with bidirectional shunt, severe tricuspid insufficiency with dilatation of right chambers, aneurysmal dilatation of pulmonary trunk and branches with severe systolic dysfunction of the right ventricle (RVEF 31%) and normal systolic function of the left ventricle (LVEF 62%). Considering high risk pulmonary arterial hypertension, which leads to rethinking therapy, an interdisciplinary meeting was held and management was adjusted by changing ambrisentan to macitentan, as well as iloprost for treprostinil.

At follow-up, the patient is under treatment with home oxygen at night, triple vasodilator therapy with riociguat, macitentan and subcutaneous treprostinil; presenting improvement in functional class. Good adherence to treatment is highlighted, which has contributed to maintain control over pulmonary hypertension.

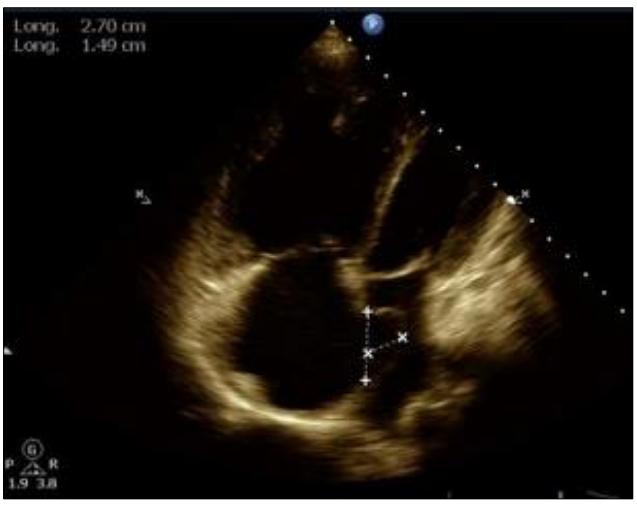

Figure 1 Echocardiographic apical plane of 4 chambers: Dilatation of right chambers, ASD and aneurysmal dilatation of the septum

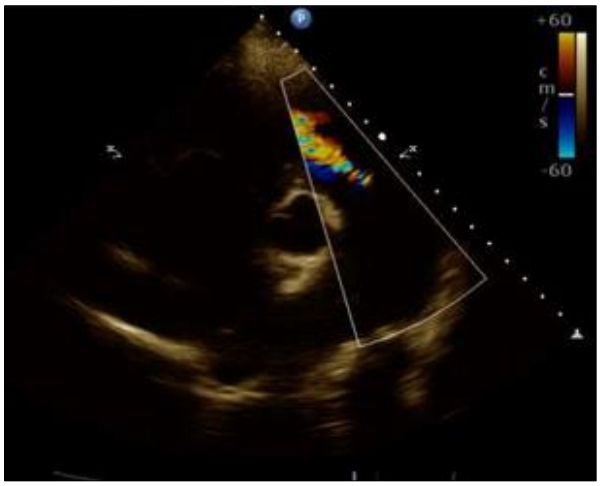

Figure 2 Echocardiographic axis at the level of great vessels: aorta and pulmonary artery, the latter dilated

3. Discussion

PH during pregnancy presents a high risk of morbidity and mortality, which justifies its classification as class IV heart disease according to the WHO, discouraging gestation (1). In this case, our patient is obtained the findings of high probability PH by transesophageal color Doppler echocardiography being suspected for the first time during pregnancy, the patient decided to continue her gestation which led to a delay in its diagnostic and therapeutic approach, causing a progression of the disease. The Pregnancy and Heart Disease Registry

in the USA reports a mortality rate of up to 43% for PAH in pregnancy, depending on the etiology and severity of PAH, while the CAR-PREG II study in Canada reports about 35.6% mortality regardless of PAH etiology. Women diagnosed with PAH are therefore strictly advised to avoid pregnancy because of the high maternal mortality reported above; moreover, many PAH-specific drug therapies are also fetotoxic. The difficulty arises when contraceptive precautions are inadequate for those with preexisting PAH or if PAH is first diagnosed during pregnancy. [6]

Initially attributed to congenital heart disease (ostium secundum ASD), the later finding of SLE revealed a mixed etiology, with the addition of an autoimmune component. SLE, as a connective tissue disease, induces chronic vascular inflammation, which increases pulmonary vascular resistance and contributes to right ventricular overload (7). This association makes classification and management more complex, since different pathophysiological mechanisms may coexist and require a comprehensive multidisciplinary approach (8).

According to the 2022 European Society of Cardiology and European Respiratory Society (ESC/ERS) clinical practice guidelines, in patients with high-risk PAH, as in this case, it is recommended to consider an initial triple combination therapy, which includes an endothelin receptor antagonist, a soluble guanylate cyclase stimulator, and an intravenous or subcutaneous prostacyclin analog. This strategy is based on the results of the French study by Boucly et al. which showed that such a combination is associated with a significantly higher survival rate compared to monotherapy or dual therapy (9,10).

In this context, the patient was finally treated with triple vasodilator therapy (macitentan, riociguat and subcutaneous treprostinil), showing functional and clinical improvement, which coincides with the findings of the aforementioned study. The therapeutic approach also included immunomodulators for SLE, being key to stabilize disease progression and improve quality of life.

On the other hand, the use of multidisciplinary teams (MDT) has been shown to improve maternal outcomes in pregnant women with PH, reducing complications such as heart failure and emergency cesarean sections, and even preventing maternal deaths (10). Low TT et al. Document a reduction in maternal mortality and improved prognosis, from 38% in the late 1990s to 28% in the 2010s and 12% in the 2020s. [11]

Finally, it should be noted that, although echocardiography is a useful diagnostic tool, it can overestimate pulmonary pressures in pregnant women, so right catheterization should not be delayed, as it is essential to confirm the diagnosis (12).

4. Conclusion

Pulmonary hypertension in pregnancy represents a high-risk clinical condition that requires a timely, multidisciplinary and evidence-based approach. This case reflects the challenges derived from late diagnosis and multifactorial etiology, with direct implications in disease progression. The implementation of triple vasodilator therapy, following current guideline recommendations and robust clinical studies, allowed significant clinical stabilization. The role of systemic lupus erythematosus as an aggravating factor underscores the need for a comprehensive approach in patients with connective tissue disease. Overall, this case highlights the importance of individualizing treatment and adopting proactive strategies to improve maternal outcomes in patients with PAH

Compliance with ethical standards

Disclosure of conflict of interest

No conflict of interest to be disclosed.

Statement of ethical approval

If studies involve use of animal/human subject; authors must give appropriate statement of ethical approval. If not applicable then mention 'The present research work does not contain any studies performed on animals/humans subjects by any of the authors'.

Statement of informed consent

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

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