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Pulmonary Arterial Hypertension Complicated by Right-Sided Heart Failure during Pregnancy in a Primiparous Woman with Scleroderma

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Abstract

We report the case of a 32-year-old primiparous patient referred to the Cardiology Department for the management of hypertension at 28 weeks' gestation. She has a history of asthma and acute rheumatic fever (ARF) in childhood. At the age of 17, she underwent percutaneous closure of a large atrial septal defect (ASD) of the ostium secundum type. Clinical examination revealed grade 1 hypertension (BP = 157/96 mmHg) and signs of right-sided heart failure. Echocardiography revealed pulmonary arterial hypertension with a pulmonary artery systolic pressure (PASP) of 73 mmHg, dilatation of the right cardiac chambers and the inferior vena cava, and grade 2 mitral regurgitation. Further investigation led to a diagnosis of scleroderma, confirmed by histopathological examination of a skin biopsy. Treatment was initiated with furosemide tablets (40 mg/day), spironolactone (50 mg/day), sildenafil (50 mg/day), and prednisolone (2.5 mg/day). Delivery was scheduled at 34 weeks' gestation by caesarean section, resulting in a live female infant who cried immediately and weighed 2400 g. The postoperative course was uneventful, with both the baby and the mother doing well. Bosentan and methotrexate combined with folic acid were initiated postpartum.

Keywords

Pulmonary Arterial Hypertension, Scleroderma, Right-Sided Heart Failure, Pregnancy

1. Introduction

Pulmonary hypertension (PH) is a pathological condition characterised by elevated pulmonary arterial pressure, leading to progressively worsening dyspnea and right heart failure [1]. The definition of PH is based primarily on haemodynamic assessment via right heart catheterisation. It is defined as a mean pulmonary artery pressure (mPAP) greater than 20 mmHg at rest [2].

PH must be distinguished from pulmonary arterial hypertension (PAH), which is a rare form of PH characterised by specific pulmonary vascular involvement. The clinical classification of PH comprises five main groups: group 1 corresponds to PAH, group 2 corresponds to PH associated with left-sided heart disease, group 3 to PH associated with chronic respiratory diseases and/or hypoxia. Group 4 corresponds to PH associated with chronic pulmonary arterial obstructions. Group 5 comprises multifactorial PH and/or PH of unknown aetiology [1] [3].

Pulmonary arterial hypertension is a rare disease affecting the pre-capillary pulmonary vascular bed. It is one of the serious complications of connective tissue diseases. Approximately 10% of PAH cases are due to connective tissue disease [4]. This involves obstructive remodelling of the pulmonary arterioles coupled with vascular rarefaction, increasing right ventricular afterload and leading to right heart failure. The pathogenesis of PAH is complex and multifactorial [3]. The histopathological lesions in connective tissue associated with PAH are similar to those in idiopathic PAH, and the pathophysiological mechanisms are comparable. However, inflammatory processes appear to be more frequently involved, particularly in lupus or connective tissue associated PAH. As few therapeutic trials have been conducted specifically in connective tissue diseases, management is similar to that of idiopathic PAH, with a poorer prognosis, and relies on bosentan, epoprostenol and other prostacyclin analogues, and sildenafil [4].

Pregnancy in women with PAH is a rare occurrence (1 in 100,000) [5]. It is associated with very high maternal and foetal mortality and morbidity, with a maternal mortality rate of between 30 and 50%. Consequently, experts advise against pregnancy in women with PAH. However, thanks to recent advances in the specific treatment of PAH and multidisciplinary care, the maternal mortality rate in PAH appears to have decreased. The physiological changes associated with normal pregnancy and the postpartum period, such as hypervolaemia and increased cardiac output, can, in patients with PAH, lead to right ventricular decompensation and cause right-sided heart failure [5] [6]. The cardiovascular system of individuals with PAH struggles to adapt to the physiological changes of pregnancy. In particular, the increase in blood volume and cardiac output will reach the limits of the right ventricle's capacity. There is therefore a risk of right heart failure, particularly in cases of severe PAH [6]. Although there is no clear threshold for elevated pulmonary artery pressure and the associated risk, pregnancies with mild PAH and vasoreactive PAH appear to have better maternal and foetal outcomes than those with moderate to severe PAH. Women with severe PAH, non-vasoreactive idiopathic PAH and Eisenmenger syndrome are at the highest risk of ma-

ternal and foetal mortality. Given that the clinical course of PAH during pregnancy remains associated with unpredictable risks and that pregnancy may accelerate the progression of PAH, all women with PAH wishing to become pregnant should be counselled by a multidisciplinary team regarding the very high risk of pregnancy-related adverse events. The risk of foetal and neonatal mortality is high, mainly due to preterm delivery, reduced maternal CO_2 and/or hypoxia. Miscarriages are common. Maternal and foetal outcomes vary depending on the subgroup of PAH. Thanks to improvements in the treatment of PAH and a multidisciplinary approach during pregnancy and the perinatal period, maternal mortality has decreased but remains high, ranging from 11% to 25%.

The standard diagnostic algorithm, in accordance with the 2022 guidelines of the European Society of Cardiology and the European Respiratory Society (ESC/ERS) for the diagnosis and treatment of pulmonary hypertension, should be followed in a pregnant woman suspected of having PAH [7].

2. Observation

We report the case of a 32-year-old female patient with asthma, a nulliparous woman with a history of acute rheumatic fever, who had recently been seen in the Rheumatology Department for an undiagnosed inflammatory rheumatic condition. Her maintenance treatment consisted of 2.5 mg of prednisolone per day. At the age of 17, through the humanitarian aid organisation "La Chaîne de l'Espoir", she had undergone percutaneous closure of a large ostium secundum type atrial septal defect (ASD). The post-operative course was uneventful. The immediate post-operative echocardiogram showed an occlusive prosthesis that did not interfere with the surrounding structures (the pulmonary veins, the coronary sinus, the cavernous veins or the atrioventricular valves). A follow-up echocardiogram two months post-operatively showed a watertight patch, minimal tricuspid regurgitation with a PASP of 21 mmHg; the cardiac chambers were not dilated. Minimal mitral regurgitation was noted with a velocity of 2.9 m/s. She had never consulted cardiology again for the follow-up of the ASD.

She was referred to us for a cardiology consultation regarding the management of hypertension at 28 weeks' gestation. The decision to conceive was made by the patient of her own free will. She presented with dyspnea classified as New York Heart Association (NYHA) class 2 and had oedema of the lower limbs. The clinical examination revealed grade 1 hypertension (BP = 157/96 mmHg); heart sounds were regular with a B2 murmur at the pulmonary focus (HR = 94 bpm); SpO_2 = 98%. There were signs of right-sided heart failure: soft, painless oedema of the lower limbs, tenderness in the right hypochondrium, and hepatojugular reflux. The electrocardiogram was normal.

On echocardiography, we visualised the ASD closure patch, which was leak-free, dilatation of the right cardiac chambers (RV = 28.2 mm, RA = 22 mm) and the inferior vena cava (24.2 mm), TAPSE = 13 mm, PASP = 73 mmHg; LV = 53.9 mm, LVEF = 81.63%, grade 2 mitral regurgitation with Vmax = 4.73 m/s; the mi-

tral valve was slightly thickened but flexible. Dilated LA (22 mm). Stage 2 mitral profile, without diastolic dysfunction (Figure 1).

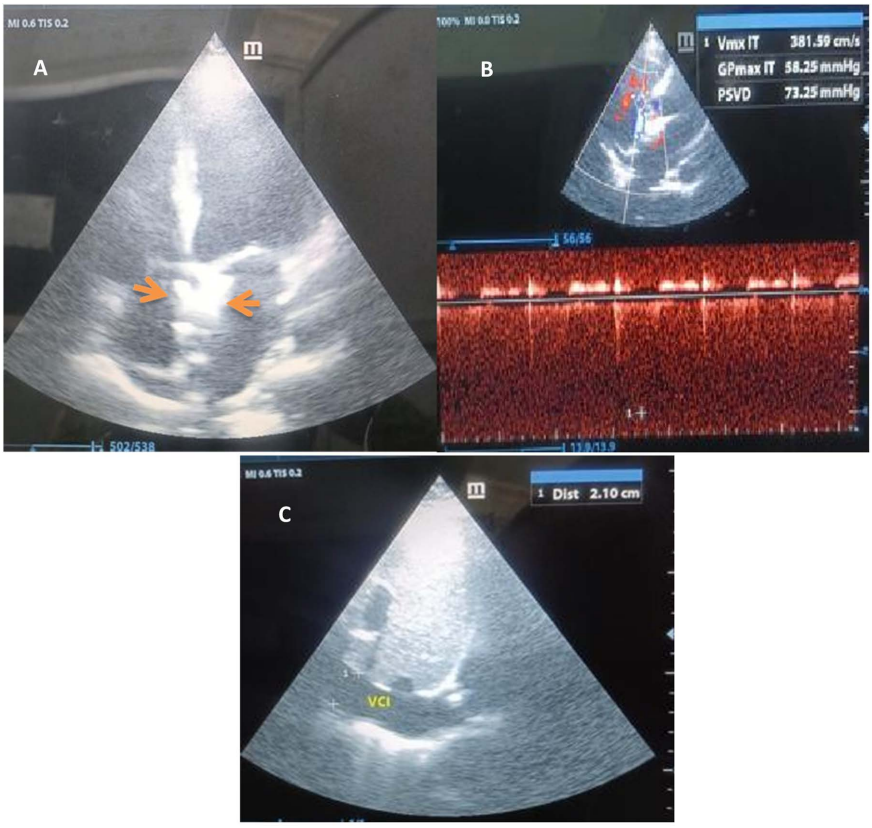

Figure 1. A: Apical view of the four chambers showing the septal closure patch (orange arrows) in the atria; B: Tricuspid regurgitation flow with a PASP of 73 mmHg; C: Dilatation of the inferior vena cava.

Biochemical findings: NT-proBNP = 129.0 ng/L, D-dimer = 2.5 ng/mL, normal complete blood count with haemoglobin level at 13.1 mg/L, serum creatinine = 10.8 mg/L, blood glucose = 0.81 g/L, serum potassium = 4.1 mEq/L; CRP = 24 g/l. PT = 84.2%.

The patient was informed of the diagnosis and the associated maternal and foetal risks.

We cautiously administered Furosemide 40 mg (1 tablet daily), Spironolactone 50 mg (1 tablet daily) and Sildenafil 50 mg (1 tablet daily), and initiated regular weekly maternal-foetal monitoring, both clinical and ultrasound-based, involving the Cardiologist and Obstetrician. Maternal monitoring parameters concerned, on the clinical level, dyspnea, edema of the lower limbs, blood pressure, heart rate. On echocardiography, the estimation of PASP from tricuspid regurgitant flow, the diameter of the RV and IVC, and left ventricular myocardial performance. For the fetus, we monitored fetal heart sounds, the amount of amniotic fluid, and fetal growth parameters (head circumference, biparietal diameter...).

By the first week, the dyspnea had resolved, as had the oedema of the lower

limbs. Blood pressure had normalised. The PASP, meanwhile, had gradually decreased and stabilised at around 45 mmHg, whilst the inferior vena cava and right ventricle remained dilated. We maintained the treatment at the same dose.

Furthermore, following consultation with the internal medicine physician, autoantibody testing had returned results consistent with scleroderma: anti-Scl-70 antibodies were positive. A skin biopsy confirmed the diagnosis, revealing a fibrous and dense dermis, containing thick collagen fibres packed closely together, with sparse blood vessels.

The delivery was scheduled for the 34th week of gestation by caesarean section. The procedure was preceded by a one-week hospital stay during which prophylactic anticoagulation was added to her treatment. Several specialist teams were involved: Cardiologists, Anesthesiologists, Neonatologists, and Obstetricians and Gynaecologists. The cesarean section, performed under general anesthesia according to the preference of the team, resulted in the delivery of a live baby girl, who cried immediately and weighed 2400 g. No resuscitation measures were required for either the mother or the newborn. The post-operative course was uneventful. Breastfeeding was contraindicated. A follow-up echocardiogram at the end of her hospital stay noted an elevation in PASP estimated at 79 mmHg, although the patient was asymptomatic. We recommended continuing her maintenance treatment (sildenafil, furosemide, spironolactone and prednisolone), adding bosentan 62.5 mg (one tablet twice daily). Methotrexate 10 mg per week combined with folic acid 5 mg was initiated in the postpartum period. To date, three months postpartum, the patient has been seen four times. On the first occasion, she was doing well; the PASP had almost normalised, estimated at 25 mmHg, and the IVC was not dilated. We had reduced the sildenafil dose to 1 tablet every 3 days. One month later, the echocardiographic assessment revealed an increase in pulmonary pressures estimated at 86 mmHg and dilation of the IVC. In view of this increase in pressure in the right heart chambers, we resumed sildenafil at a daily dose. At the last two check-ups, the patient remained asymptomatic, with pulmonary pressure stable at around 30 mmHg. This pressure was estimated each time in the apical view of the four chambers focused on the right heart cavities.

With the agreement of the Hospital Management, the patient gave her consent for this interesting case to be published.

3. Discussion

Of all cardiac events occurring during pregnancy, left ventricular cardiomyopathies are the most frequently reported, but right-sided heart failure remains a rare occurrence. The onset of right-sided heart failure during pregnancy is insidious and the signs are misleading, as they can be confused with the physiological changes that occur during a normal pregnancy.

Connective tissue diseases in general, and scleroderma in particular, are often complicated by pulmonary arterial hypertension [8]. Echocardiography is the gold standard screening test but cannot confirm the diagnosis or determine the

mechanism [1]. Right heart catheterisation should only be considered in cases of diagnostic uncertainty regarding the identification of de novo PAH during pregnancy and to aid in making important therapeutic decisions [7]. It should be noted that the echographic assessment of pulmonary arterial pressures is well correlated with the evaluation by right heart catheterization during pregnancy [9]. In the case of our patient, it seemed evident to us that the pulmonary hypertension is related to the systemic disease, scleroderma, especially since the echocardiography did not reveal any residual atrial septal defect leakage, the closure patch was watertight, there was no increase in left ventricular filling pressures to suggest post-capillary pulmonary hypertension, and the negative D-dimers ruled out a thromboembolic cause. Given this certainty, we postponed performing right heart catheterization, which is also invasive and costly. In a pregnant patient with PAH, the increase in blood volume and cardiac output that occurs during pregnancy can exacerbate PAH and lead to right ventricular decompensation [6]. We combined Spironolactone with furosemide in this case of right heart failure and hypertension during pregnancy for the pharmacological benefits it offers both for the mother and the fetus. While furosemide is a powerful diuretic capable of inducing both hypovolemia detrimental to the fetus and hypokalemia that can endanger the mother's life, the administration of spironolactone, which has a less potent diuretic effect than furosemide, moderately enhances the diuretic effect and establishes a certain balance between potassium excretion and reabsorption. Spironolactone is a potassium-sparing diuretic, an aldosterone antagonist. It also has an antihypertensive property [10].

Three critical periods have been identified during pregnancy in a person with PAH: the third trimester, labour and the postpartum period [5] [6].

The postpartum period, and particularly the first week, is very crucial because it is well known that most pregnant patients with PAH decompensate acutely during this period due to hemodynamic changes [11].

Several complex phenomena could explain the increase in PASP in our patient after delivery: the release of the obstruction of the vena cava by the gravid uterus, which leads to shifts of a large blood volume into the systemic and pulmonary circulation, variations in intrathoracic pressure which affect preload and afterload. Also, the rapid drop in estrogen and progesterone levels, on the one hand, and the hemorrhages that occur in the postpartum period, on the other hand, induce vasoconstriction [12].

Endothelin receptor antagonists (bosentan) are an effective class of medication but are contraindicated during pregnancy due to the risk of teratogenicity [5]. The significant decrease in PASP with the introduction of bosentan confirms the effectiveness of endothelin receptor antagonists in the treatment of pulmonary arterial hypertension associated with scleroderma [13].

In most cases, sildenafil is combined with a prostacyclin analogue depending on the severity of the disease [7]. When we had inappropriately reduced the dose of sildenafil in the treatment of our patient after childbirth, the PASP had signifi-

cantly increased, leading to an increase in pressures in the right heart chambers and the IVC. But the restoration of a daily dose had almost normalized the pressures. This episode highlights, on the one hand, the importance of sildenafil in the treatment of pulmonary hypertension due to its vasodilatory effect, and on the other hand, it would testify to the "vasoactive" nature of the patient [14].

Prophylactic anticoagulation is necessary during the perinatal period [15].

In our patient's case, the use of diuretics at relatively moderate doses struck the right balance to provide clinical relief for the patient without jeopardising the foetus's life. The 34th week of gestation is the time when most medical teams choose to perform an elective cesarean section, as it allows for avoiding the hemodynamic constraints related to pain during labor and the pushing efforts [9] [16].

4. Conclusion

Pulmonary arterial hypertension complicated by right-sided heart failure during pregnancy is a rare occurrence. Scleroderma is one of the common aetiologies. It is exacerbated by the physiological changes occurring during gestation. The maternal-foetal prognosis remains guarded in the peripartum period, but improvements in treatment and multidisciplinary management have significantly reduced maternal mortality.

Conflicts of Interest

The authors declare no conflicts of interest regarding the publication of this paper.

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