Chronic thromboembolic pulmonary hypertension: uneventful pregnancy after pulmonary balloon angioplasty treatment. Case report

Hipertensión pulmonar tromboembólica crónica: embarazo sin incidentes después del tratamiento de angioplastia pulmonar con balón. Reporte de un caso

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ABSTRACT

Pulmonary hypertension is associated with a significant risk for both mother and child during pregnancy. While pulmonary endarterectomy is the treatment of choice for patients with chronic thromboembolic pulmonary hypertension, balloon pulmonary angioplasty has emerged as an alternative for patients who are not eligible for surgery. This is the case of a 33-year-old patient with chronic thromboembolic pulmonary hypertension who was considered inoperable and who became pregnant after balloon pulmonary angioplasty treatment. The delivery was achieved without complications under strict multidisciplinary monitoring.

Keywords: *chronic thromboembolic pulmonary hypertension, balloon pulmonary angioplasty, pregnancy outcome.*

RESUMEN

La hipertensión pulmonar durante el embarazo implica un riesgo significativo tanto para la madre como para el recién nacido. Mientras que la endarterectomía pulmonar es el tratamiento de elección en pacientes con hipertensión pulmonar tromboembólica crónica, la angioplastia pulmonar con balón se convirtió en una alternativa válida para los pacientes que no son candidatos a cirugía. Presentamos el caso de una mujer de 33 años con hipertensión pulmonar tromboembólica crónica, no candidata para resolución quirúrgica, que cursó su embarazo después de ser tratada con angioplastia pulmonar con balón, lográndose un parto sin complicaciones bajo un monitoreo estricto multidisciplinario.

Palabras claves: *hipertensión pulmonar tromboembólica crónica, angioplastia pulmonar con balón, resultado del embarazo.*

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INTRODUCTION

Chronic thromboembolic pulmonary hypertension (CTEPH) is a rare complication of acute pulmonary thromboembolism. Its incidence rate is between 0.1% and 9.1% and its long-term mortality rate is high[1].

Although pulmonary endarterectomy (PE) is the treatment of choice in patients who are not eligible for surgery, medical treatment or balloon angioplasty should be both considered since they are both good alternatives[1].

The expert clinical practice guidelines on the management of pulmonary hypertension (PH) recommend avoiding pregnancy[1]. However, when this is the case, both the mother and the fetus have a high mortality rate during pregnancy and delivery[2-4]. This is the case of a patient with CTEPH who became pregnant after receiving treatment with balloon pulmonary angioplasty (BPA).

CLINICAL CASE

This is the case of a 33-year-old woman with a past medical history of multiple fractures in hip, femur, and left kneepad after a motorcycle accident occurred in 2006. The patient's health status became complicated with chronic osteomyelitis that required the insertion of a permanent central venous catheter for prolonged antibiotic therapy. Back in 2017, she was admitted to a different center after presenting with progressive dyspnea. An acute pulmonary thromboembolism was diagnosed and oral anticoagulation with acenocoumarol was started. After completing a 3-month course of treatment, the patient failed to show any symptom improvement (functional class III according to the World Health Organization) and she had to be transferred to our hospital.

The physical examination confirmed the presence of tachypnea and tachycardia. The oxygen saturation level on room air was 90%. The electrocardiogram confirmed the presence of sinus tachycardia with right bundle branch block (RBBB). At the lab, the most significant finding was a proBNP of 4310 pg/mL. The presence of deep venous thrombosis was discarded on the venous Doppler ultrasound. The transthoracic echocardiography performed revealed the presence of right atrial and ventricular dilatation, moderate right ventricular systolic dysfunction, and a right ventricular systolic pressure of 80 mmHg. No signs of cardiac vegetations were seen. The ventilation/perfusion lung scan performed revealed the presence of bilateral mismatch with multiple areas of hypoperfusion at baseline level. These findings were confirmed on a CCTA with contrast that revealed the presence of thromboembolic disease at the level of subsegmental branches with both lower lobe predominance. These were the parameters obtained

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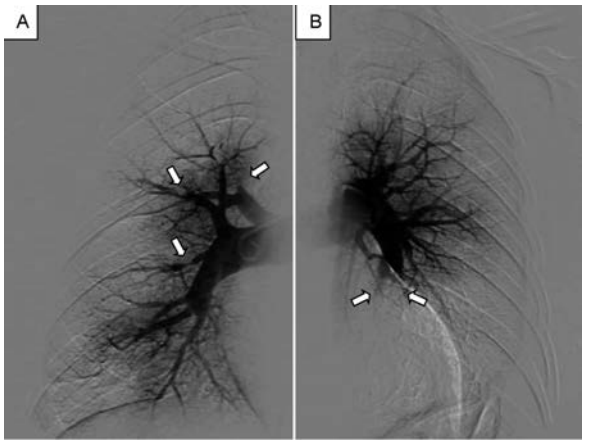

Figure 1. *Bilateral pulmonary angioplasty. White arrows: significant lesions of segmental and subsegmental vessels..*

during right cardiac catheterization: pulmonary artery pressure (PAP), 62/25 (50) mmHg; pulmonary capillary wedge pressure (PCWP), 7 mmHg; volume per minute (VM): 4.1 L/min; pulmonary vascular resistance (PVR), 878 dyn·s·cm^{-5} (10.9 Wood units). The pulmonary angiography showed web-like and annular lesions with subsegmental predominance (**Figure 1**). The case was discussed with the CTEPH heart team and the patient was considered non-eligible for PE because her lesions were at peripheral pulmonary vasculature level. Treatment with riociguat and multiple sessions of BPA were prescribed.

Eight sessions of BPA were conducted in the cath lab. A 7-Fr Flexor guiding sheath (Cook France S.A.R.L, Charenton-le-Pont, France) was used via left or right femoral vein in each procedure. A 0.014 in guidewire (Runthrough NS PTCA Guide Wire, Terumo, Japan) or Choice PT Floppy guidewire (Boston Scientific, Marlborough, Massachusetts, United States) mounted over a JR4 7-Fr guide catheter (Boston Scientific Marlborough, Massachusetts, United States) was advanced. The lesions were crossed uneventfully. Successive dilatations were performed with Emerge balloons (BostonScientific, Marlborough, Massachusetts, United States) of between 2.0 mm and 4.0 mm in diameter. Measurements were decided based on the size of the blood vessel in a 1:1 ratio (**Figure 2**). Inflations lasted from 30 to 60 seconds. After 8 sessions without serious complications or symptoms during the procedure, the hemodynamic parameters improved significantly: PAP decreased to 50/20 (30) mmHg and PVR dropped to 417 dyn·s·cm^{-5} (5.2 Wood units). The oxygen saturation levels were 95%. Although treatment with BPA had not been completed the patient became pregnant and it had to be suspended. The risks associated with the pregnancy were explained to the patient who, nevertheless, decided to go on with it. Acenocoumarol was combined with subcutaneous enoxaparin twice a day and riociguat was withdrawn. The patient was closely monitored by a multidisciplinary team. Full-term pregnancy came without complications and a C-section was scheduled for the 37th week of pregnancy. Prior to surgery a Swan Ganz catheter was inserted for strict hemodynamic monitoring purposes (mean PAP, 37 mmHg; VM, 4.3 L/min; PVR, 465 dyn·s·cm^{-5}) and under epidural anesthesia the baby was born without complications. The newborn baby weighed 2600 grams and scored 9/10 in the Apgar scale. Both the mother and the baby were discharged

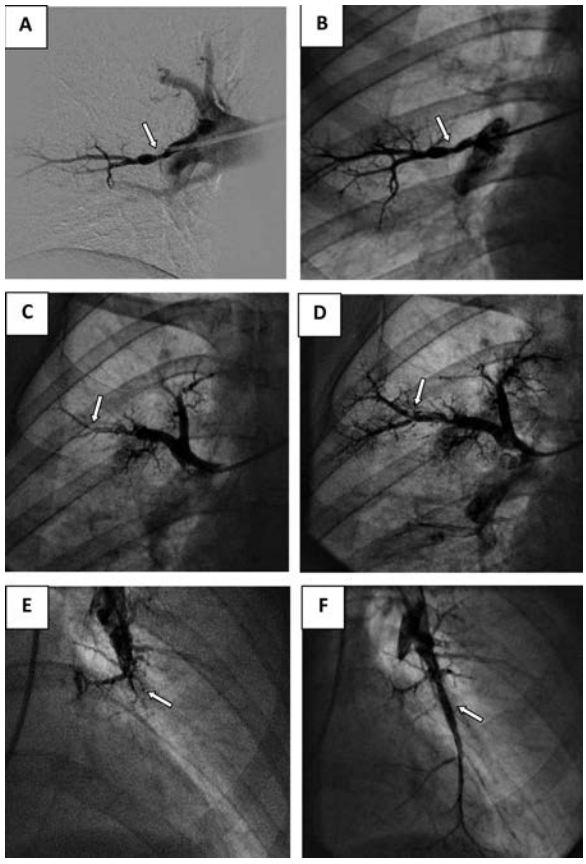

Figure 2. *Pulmonary angiography during balloon pulmonary angioplasty. (A) Annular lesion in the A5 right segment before and (B) after the BPA. (C) Web-type lesion in the A4 right segment before and (D) after de BPA. (E) Subocclusive lesion in the A9 left segment before and (F) after the BPA..*

from the hospital after 5 days showing good clinical conditions at the 30-day follow-up.

DISCUSSION

During pregnancy, the body of women undergo physiological changes; at the end of pregnancy blood volume can increase over 50%[5].

Through changes in systemic vascular resistance and heart rate, the cardiac output can increase up to 50% compared to non-pregnant women[5]. Patients with PH have an increased PVR due tovascular remodeling. Therefore, these patients may not tolerate increased heart rates and blood volume overloads. Our patient may have tolerated well the physiological hemodynamic changes thanks to previous improvement after treatmentwitha BPA.

The increased mean PAP during right cardiac catheterization prior to delivery may be associated with reduced pulmonary artery compliance as reported by Magon et al.[6]

Epidural anesthesia was used. Local anesthesia is the preferred one because general anesthesia is often associated with an up to a 4 times higher maternal mortality rate due to reduced cardiac contractility and increased PVR and PAP[7]. Discussion on the optimal type of delivery is still controversial. Although vaginal delivery is associated with fewer significant changes in blood volume, fewer complications of coagulation or hemorrhage, and a lower risk of infection[7], the C-section was decided to avoid a second prolonged stage of labor and a possible and incontrollable vaginal blee-

ding[5]. Very few cases of pregnant women with CTEPH have been reported in the medical literature. Both Ikeda et al.[8] and Kopec et al.[9] reported on cases of patients with CTEPH treated with BPA who became pregnant. In both cases, treatment with BPA was considered completed. This was shown in the final mean PAP obtained. Our patient became pregnant before all the sessions scheduled for her would ever be completed. This may explain the high mean PAP seen before pregnancy was ever confirmed.

CONCLUSION

Pregnancy in patients with PH is a very high-risk situation both for the mother and the baby. After the BPA our patient improved significantly on both the hemodynamic and clinical levels. We believe that these interventions facilitated an uneventful pregnancy and delivery. Close monitoring by the multidisciplinary team should be the standard of care in high-risk patients like these.

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