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Case Report

Postpartum dilated cardiomyopathy and antiphospholipid syndrome: A rare association revealed by a pulmonary embolism (case report)☆,☆☆

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ABSTRACT

Antiphospholipid syndrome is a rare entity that must be systematically evoked in front of recurrent miscarriages associated with venous or arterial thrombosis, its diagnosis is based on a set of clinico-biological arguments. In rare cases, it can be associated with postpartum cardiomyopathy, which is defined by a dysfunction of the left ventricle with an LVEF<45%, which may or may not be associated with a dilation of the left ventricle. This association is rare and poorly described in the literature, which makes management difficult and uncodified. In this context we report the case of a 33-year-old patient with cardiovascular risk factors such as arterial hypertension 2 previous miscarriages and repeated phlebitis, she was admitted to the emergency room for the management of acute dyspnea related to a proximal right pulmonary embolism and in whom the transthoracic echocardiography had objectivated a dilated left ventricle and an alteration of the ejection fraction of the left ventricle, the coronary angiography came back without particularity as part of the etiological work-up, a biological work-up was carried out, which came back in favor of an antiphospholipid syndrome. This case shows diagnostic difficulties and management of this disease.

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Introduction

Peripartum cardiomyopathy is a rare etiology of heart failure. It affects, by definition, young women and results in dilated cardiomyopathy. It presents as heart failure most often immediately after delivery, more rarely during the last month of pregnancy [1].

Antiphospholipid syndrome is defined by hypercoagulability (venous or arterial thrombosis) and pregnancy morbidity (fetal loss, pre-eclampsia, and eclampsia) occurring in the presence of antiphospholipid antibodies, anticardiolipin antibodies and lupus-anticoagulant test [2]. Pulmonary embolism is the most common pulmonary manifestation and may be the first sign of antiphospholipid syndrome [3,4]. the antiphospholipid syndrome must be systematically evoked in front of arterial or venous thrombosis, and recurrent fetal losses [5]. Recently also patients with antiphospholipid have been described in whom the known maternal fetal complications were followed by the appearance in the puerperium of severe, sometimes fatal syndromes involving the lungs and the heart [6,7].

In this context, we report the case of a 33-year-old female patient with a history of 2 miscarriages and repeated phlebitis who was admitted for the management of acute rest dyspnea related to a proximal right pulmonary embolism secondary to an antiphospholipid syndrome associated with postpartum cardiomyopathy.

Our case report was written according to CARE guideline [33].

Case presentation

A 33-year-old female patient presented to the emergency room with acute resting dyspnea. The patient has several cardiovascular risk factors, including poorly controlled arterial hypertension for 3 years, recent childbirth, 2 miscarriages, and a history of thrombophlebitis in the right lower limb in 2014 and 2018, as well as thrombosis of the inferior vena cava in 2019. She has not undergone any previous surgeries.

Upon examination, the patient was found to be conscious, with normal weight and height, but hypertensive. She exhibited signs of respiratory instability, including an elevated respiratory rate and decreased oxygen saturation. The EKG revealed a regular sinus rhythm with features of left ventricular hypertrophy, and the chest X-ray was unremarkable.

Transthoracic echocardiography (TTE) indicated severe left ventricular dysfunction with dilated cardiomyopathy (LVEF of 35%) (Fig. 1). The right ventricle was nondilated, hypertrophied, and dysfunctional. The filling pressures of the left ventricle were normal, and there was an intermediate probability of pulmonary hypertension.

Due to the high clinical probability of pulmonary embolism, a thoracic angioscan was performed immediately revealing a proximal right pulmonary embolism (Fig. 2). Additionally, given the left ventricular dysfunction, coronary angiography was conducted, revealing no particular abnormalities (Figs. 3 and 4).

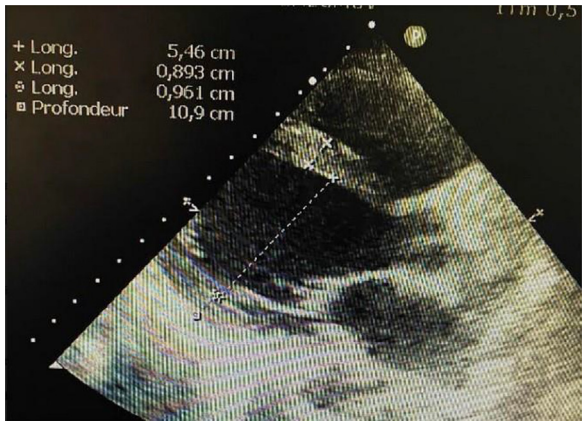

Fig. 1 – Parasternal long axis section showing a dilated LV with nonhypertrophied walls and a telediastolic diameter of the left ventricle of 54 mm.

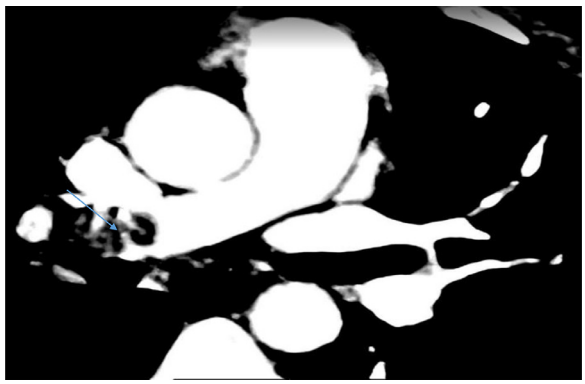

Fig. 2 – Thoracic angioscan showing a proximal right pulmonary embolism (blue arrow).

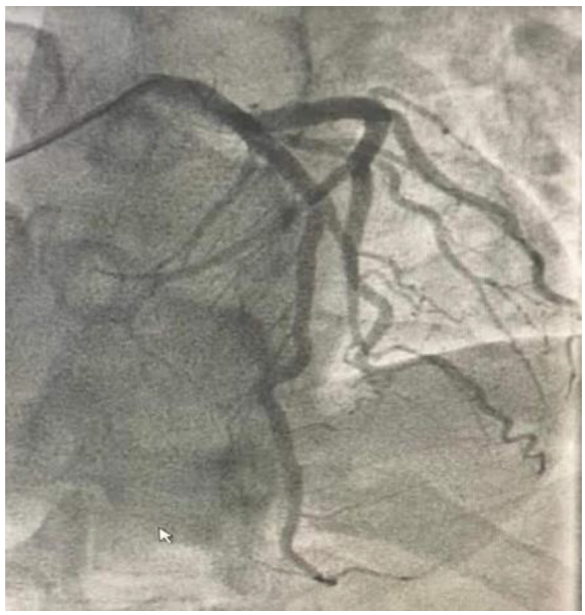

Fig 3 – Coronary angiography performed by radial approach showed a left network without significant lesions.

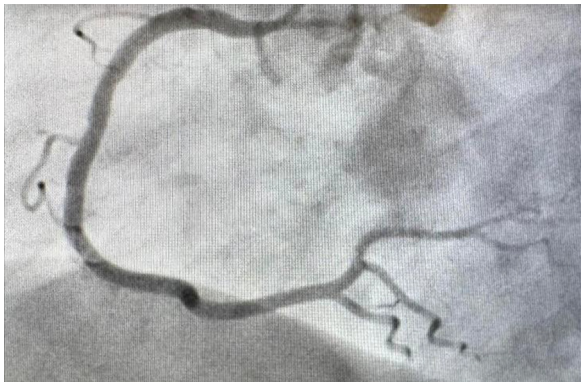

Fig. 4 – Coronary angiography performed by radial approach showing a right network without significant lesion.

This complex case involves multiple cardiovascular issues, including dilated cardiomyopathy, arterial hypertension, a history of thrombophlebitis, and a possible pulmonary embolism. The multidisciplinary management of this patient may involve specialists in cardiology, pulmonology, and potentially hematology, given the history of thrombotic events. Further investigations and a comprehensive treatment plan would likely be necessary to address the various aspects of her cardiovascular health and to determine an appropriate course of action.

On admission, the patient's hemoglobin was within the normal range (12 g/dL), troponin levels were elevated (0.028 pg/mL), indicating cardiac involvement, and the CRP was significantly elevated (45 mg/L), suggesting inflammation. The extended activated partial thromboplastin time (TCA) indicates a coagulation abnormality. Renal function, however, was normal on admission.

As part of the etiological workup, the patient tested positive for circulating anticoagulant antibodies (IgG) and anti-β2GPI antibodies (IgG), indicative of antiphospholipid syndrome. Notably, despite the presence of antiphospholipid antibodies, the patient did not meet the criteria for a diagnosis of systemic lupus erythematosus (SLE).

Given the complex clinical presentation and the need for differential diagnosis, potential causes of acute dyspnea were considered, including asthma decompensation, pneumothorax, tamponade, and acute lung edema.

In the cardiac intensive care unit, the patient was treated with unfractionated heparin, an anticoagulant, to address the acute pulmonary embolism. This was administered at a dose of 50 IU/kg intravenously followed by 500 IU/kg/d for 5 days. Subsequently, antivitamin K therapy, analgesics, a proton pump inhibitor, and beta-blockers were initiated. The patient was also started on sacubitril/valsartan (50 mg*2/d) and spironolactone (50 mg/d) for heart failure management.

This comprehensive treatment approach addresses both the acute pulmonary embolism and the underlying cardiac dysfunction, aiming to stabilize the patient and manage the multifaceted aspects of her condition. Continued monitoring and further adjustments to the treatment plan may be necessary based on the patient's response and ongoing assessments. The involvement of a multidisciplinary team is crucial for optimal care in such complex cases.

It's reassuring to learn that the patient remained stable throughout her hospitalization and that her dyspnea resolved by the fourth day of admission. The 7-day stay in the cardiology intensive care unit, followed by a transfer to clinical cardiology, suggests a carefully monitored and managed recovery process.

The fact that the patient was discharged with a follow-up appointment scheduled for 1 month indicates a continued commitment to monitoring her progress and ensuring ongoing care. This follow-up appointment is essential for assessing the patient's recovery, adjusting medications as needed, and addressing any lingering concerns or potential complications.

Discussion

In 2010, the European Society of Cardiology Heart Failure Association Working Group defined postpartum cardiomyopathy as an idiopathic cardiomyopathy with heart failure secondary to left ventricular systolic dysfunction late in pregnancy or in the months after delivery, when no other cause of heart failure is found" [8]. Diagnostic criteria indicate that the LVEF is <45% and that there may or may not be ventricular dilatation [8,9]. Global estimates of the incidence of postpartum cardiomyopathy vary by regions, with reports as high as 1 in 100 deliveries in Nigeria [10]. and 1 in 300 deliveries in Haiti [11], to as low as 1 in 20,000 deliveries in Japan [12]. In the United States, reported incidence ranges from 1 in 1000 to 1 in 4000 [13–15], Many cases may moreover be unrecognized; thus, the true incidence is unknown. The etiology of Postpartum cardiomyopathy is not fully understood and is likely multifactorial. Suggested but not proven mechanisms for the development of Postpartum cardiomyopathy have included nutritional deficiencies [16,17], viral myocarditis [18,19], and autoimmune processes [20,21]. Hemodynamic stress of pregnancy has been postulated as a potential etiology. However, the maximal cardiovascular changes occur in the second trimester [22], when most women with preexisting cardiac disease develop symptomatic heart failure [23]. In contrast, the majority of women with Postpartum cardiomyopathy develop symptoms during late pregnancy or after the delivery [24]. Antiphospholipid syndrome (APS) is defined by the association of clinical manifestations (arterial or venous thrombosis and obstetrical events) and biological manifestations (repeated presence of antiphospholipid antibodies) [25]. This syndrome may be isolated, and therefore considered primary, or associated with other autoimmune diseases, and in particular with systemic lupus erythematosus. Revised criteria for the definition of APS were published in 2006, based on the principle of the combination of one of the clinical criteria and one of the biological criteria [26]. In addition to the manifestations of venous or arterial thrombosis, fetal loss, and other obstetrical complications, the cardiovascular system is indeed one of the target organs of APS. Indeed, the cardiac manifestations of APS are very diverse and can range from asymptomatic valvular damage to life-threatening myocardial infarction. Among these manifestations, we will

Table 1 – Classification criteria for the antiphospholipid syndrome (Sydney criteria) [28].

Clinical criteria	Venous thrombosis (e.g., deep vein thrombosis, pulmonary embolism, unusual site venous thromboembolism)
	Arterial thrombosis (e.g., coronary artery disease, transient cerebral ischaemia or stroke, peripheral artery disease)
	Obstetric complications: • Three or more unexplained consecutive spontaneous abortions <10th week of gestation. • One or more unexplained deaths of a morphologically normal fetus ≥10th week of gestation. • One or more premature births of a morphologically normal neonate <34th week of gestation due to eclampsia, severe pre-eclampsia, or placental insufficiency
Laboratory criteria	Lupus anticoagulant, detected according to international guidelines
	Anti-cardiolipin antibodies, IgG, or IgM isotype, at high titre (>99th percentile of normal controls)
	Anti-β2 glycoprotein-I antibodies, IgG, or IgM isotype, at high titre (>99th percentile of normal controls)

mention valvular anomalies (valvular thickening and vegetations), arterial occlusive diseases (atherosclerosis and myocardial infarction), pulmonary hypertension, ventricular dysfunctions, and intracardiac thrombi associated with Antiphospholipid syndrome [27]. It should be noted that pulmonary embolism is the most common pulmonary manifestation and may be the first sign of APS [3,4].

The diagnosis of antiphospholipid syndrome requires the presence of at least 1 of the 2 clinical criteria and at least one of the biological criteria as illustrated in the table below (Table 1).

It is widely accepted that antiphospholipid syndrome is frequently associated with pregnancy complications such as early onset pre-eclampsia, thrombocytopenia, thromboembolic events, early and late fetal loss, IUGR and corea gravidarum [29,30]. Pregnancy and the postpartum period may accelerate the appearance of the clinical signs of the syndrome. Actually, in the last years, 5 cases have also been described in which the presence of antiphospholipid syndrome was followed by serious puerperal manifestations with prevalent pleuropulmunary and cardiac involvement [6,7]. it should be noted that for women with peripartum cardiomyopathy who have delivered and are not breastfeeding, acute and chronic HF should be managed using standard GDMT (Guideline-directed medical therapy). Given the benefits of breastfeeding, women who are clinically stable should not be discouraged from breastfeeding as long as it is compatible with their heart failure medications. In breastfeeding women, beta-blockers, enalapril, and spironolactone are compatible with breastfeeding. ARBs, neprilysin inhibitors, and Ivabradine do not have enough information and should be avoided during pregnancy and lactation. Captopril and enalapril were found in clinically insignificant amounts in the breast milk and are deemed to be compatible with breastfeeding according to the American Academy of Pediatrics [31]. In the ESC-EORP registry, 30% of deaths at 6 months were due to sudden cardiac death, which may suggest that defibrillators could play an important role. Therefore, it is reasonable to wait for 6 months of optimal medical treatment before considering the timing of defibrillator implantation or cardiac resynchronization therapy [32]. It is important to know that the assessment of cardiovascular risk and early atherosclerosis is essential in patients with a primary form of APS, to propose a preventive therapeutic management. The therapeutic discussion must be done on a case-by-case basis and is most often based on prolonged effective anticoagulation, with an INR target of between 3 and 3.5 [27].

In our case, the patient was 33 years old, with cardiovascular risk factors such as hypertension for 3 years, poorly controlled with dietary hygiene measures, and with a history of 2 miscarriages and 2 thrombophlebitis of the right lower limb in 2014 and 2018 and an inferior vena cava thrombosis in 2019, She was admitted to the cardiology intensive care unit for the management of a proximal right pulmonary embolism related to an antiphospholipid syndrome associated with postpartum cardiomyopathy. The patient was stabilized and then declared discharged under treatment of heart failure associated with curative anticoagulation based on antivitamins K 1cp/d with a control appointment in 1 month.

Conclusion

The diagnosis of postpartum cardiomyopathy must be evoked in the presence of any cardiac symptomatology occurring in the days or months following delivery.

The cardiac manifestations of APS are multiple, including pulmonary embolism, which can be the first manifestation of APS.

The association between postpartum cardiomyopathy is rare and its management is multidisciplinary (cardiologist, obstetrician, pediatrician, emergency doctor), the treatment is initially based on curative dose anticoagulation associated with a treatment of heart failure.

Author contributions

Zakaria El Marraki: Study concept, Data collection, Data analysis, Writing the paper. Karim Mounaouir: Data collection, Rokaya Fellat: Supervision and data validation. Nadia Fellat: Supervision and data validation.

Registration of research studies

This is not an original research project involving human participants in an interventional or observational study but a case report. This registration was not required.

Rail registry number

This is not an original research project involving human participants in an interventional or an observational study but a case report. This registration was not required.

Provenance and peer review

Not commissioned, externally peer-reviewed.

Ethical approval

The ethical committee approval was not required given the article type (case report). However, the written consent to publish the clinical data of the patients was given and is available to check by the handling editor if needed.

Patient consent

Written informed consent was obtained from the patient for publication of this case report. CARE guidelines were applied for reporting this case report' findings.

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