Obstetrical and Pediatric Anesthesia

Two cases of postpartum cardiomyopathy initially misdiagnosed for pulmonary embolism

[Deux cas de cardiomyopathie du postpartum diagnostiqués d'abord comme embolie pulmonaire]

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Purpose: To underline the crucial role of urgent echocardiography in the differential diagnosis of acute respiratory and/or circulatory failure in the postpartum period.

Clinical features: A 24-yr-old woman was admitted to the intensive care unit (ICU) with a preliminary diagnosis of pulmonary embolism (PE) one week after Cesarean section. Neither computerized tomography nor Doppler sonography showed any signs of deep venous thrombosis or PE. In the ICU she required intubation and ventilatory support for acute respiratory and circulatory failure. Bedside echocardiography revealed features of left ventricular failure. A diagnosis of postpartum cardiomyopathy (PPCM) was made, appropriate treatment instituted and the patient soon improved.

A 29-yr-old, previously healthy primipara presented at the Maternity Clinic on the fourth postpartum day complaining of increasing dyspnea and fatigue. Within eight hours she required intubation, ventilatory support and subsequent defibrillation due to cardiac arrest. She was transferred to the ICU with a preliminary diagnosis of PE. She developed pulmonary edema followed by cardiac arrest with successful resuscitation. Bedside echocardiography revealed a left ventricular ejection fraction below 30% with an increased systolic diameter of the left ventricle, restrictive diastolic abnormalities and no signs of pulmonary hypertension. Peripartum cardiomyopathy was diagnosed and supportive treatment for heart failure was instituted.

Conclusion: It is possible to misdiagnose postpartum cardiomyopathy for PE. An error in diagnosis is life-threatening for the patient. Echocardiography is a valuable tool in the differential diagnosis. As a noninvasive procedure, it should be performed at the bedside as soon as possible to institute proper treatment and to avoid potentially fatal errors.

Objectif : *Souligner le rôle crucial d'une échocardiographie d'urgence dans le diagnostic différentiel d'une insuffisance respiratoire et/ou circulatoire aiguës du postpartum.*

Éléments cliniques : *Une femme de 24 ans a été admise à l'unité de soins intensifs (USI) à la suite d'un diagnostic préliminaire d'embolie pulmonaire (EP), une semaine après une césarienne. Ni la tomographie par ordinateur, ni l'échographie Doppler n'ont montré de signes de thrombose veineuse profonde ou d'EP. À l'USI, la patiente a eu besoin d'assistance par intubation et ventilation en raison d'une insuffisance respiratoire et circulatoire aiguës. L'échocardiographie réalisée au chevet de la patiente a révélé une insuffisance ventriculaire gauche. On a alors diagnostiqué une cardiomyopathie du postpartum (CMPP) et commencé un traitement approprié qui a rapidement amélioré l'état de la patiente.*

Une primipare de 29 ans, auparavant en bonne santé, s'est présentée à la Clinique de maternité le quatrième jour après l'accouchement, se plaignant de dyspnée et de fatigue. En moins de huit heures, elle a eu besoin d'intubation, de ventilation et de défibrillation à la suite d'un arrêt cardiaque. Elle a été transférée à l'USI avec un diagnostic préliminaire d'EP. Un œdème pulmonaire s'est développé, suivi d'un arrêt cardiaque et d'une réanimation réussie. L'échographie faite au chevet de la malade a montré une fraction d'éjection ventriculaire gauche au-dessous de 30 % ainsi qu'une augmentation du diamètre systolique du ventricule gauche, des anomalies diastoliques restrictives mais aucun signe d'hypertension pulmonaire. Une cardiomyopathie du péripartum a été diagnostiquée et un traitement de soutien instauré pour l'insuffisance cardiaque.

Conclusion : *Il est possible de confondre la cardiomyopathie du postpartum et l'EP. C'est une erreur de diagnostic qui peut être grave pour la patiente. L'échocardiographie est alors un outil précieux pour*

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le diagnostic différentiel. Comme ce n'est pas une intervention effractive, elle peut être faite au chevet du malade aussi tôt que possible pour permettre l'instauration du traitement requis et éviter des erreurs potentiellement fatales.

POSTPARTUM cardiomyopathy (PPCM) is a rare cause of respiratory and/or circulatory failure in late pregnancy and puerperium. Recently it has been advocated that data on the presentation and management of this rare condition be collected internationally. We report two cases of PPCM that were initially misdiagnosed for pulmonary embolism (PE). Transthoracic echocardiography (TTE) allowed proper diagnosis in both cases.

Case 1

A 24-yr-old woman was admitted to the labour ward at the 37th week of a twin pregnancy in preterm labour. Her past medical history was unremarkable; her obstetric history included one spontaneous abortion at the eighth week of gestation. Urinary tract infection was diagnosed and treated with antibiotics twice during the present pregnancy. The labour was terminated with a Cesarean section (ultrasonography revealed one dead fetus and the second presenting as a transverse lie). The immediate postoperative course was uneventful. On the third postoperative night she started to complain of dyspnea that was not relieved by theophilline. She was not pyretic, her heart rate was 96 beats·min^{-1} and blood pressure was 155/90 mmHg. On auscultation bilateral rales and crepitations were heard below the level of scapulas. On the next morning the chest *x-ray* (CXR) taken in the supine position showed almost uniform opacity of the right lung, a normal left lung and an enlarged heart. Pneumonia and PE were considered and the patient was given antibiotics, an infusion of heparin intravenously, as well as digoxin and theophillin. For the next four days she did not improve, complained of cough and reported light, slightly bloody sputum. Her body temperature remained below 38°C throughout. The next CXR showed progression of the previously described pathologic changes to the left lung. The consulting pneumologist suggested PE, directed therapy accordingly (increased dose of heparin, oxygen via a face mask) and suggested further investigations. In the radiology department the patient progressed to respiratory failure and, after an urgent anesthetic consultation, she was admitted to the intensive care unit (ICU). Computerized tomography (CT) of the chest revealed bilateral hydrothorax and vast opacities of both lungs, characteristic of pulmonary edema. *IV* contrast solution did not reveal any changes

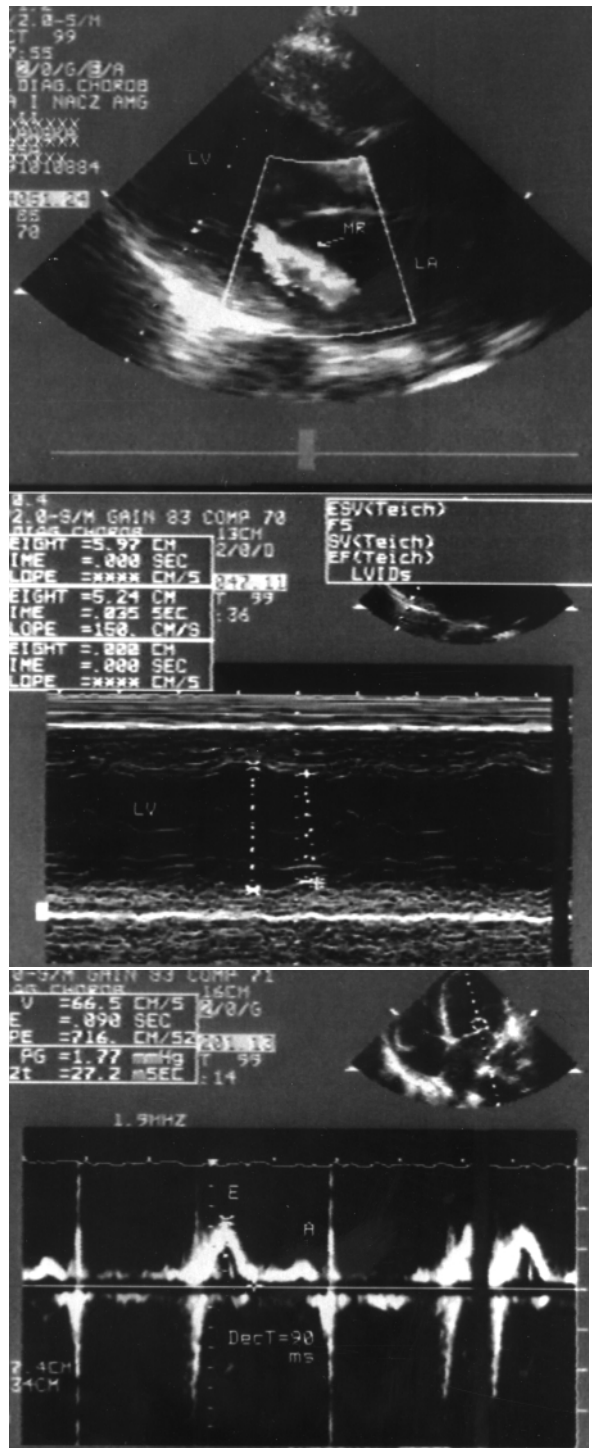

FIGURE 1 Case 1: Echocardiographic examination before treatment. A, Parasternal long axis view, dilated left ventricle (LV) and left atrium (LA). Jet of mitral regurgitation (arrow) detected in colour Doppler technique. B, M-mode: Abnormal diastolic and systolic diameter of the left ventricle with low ejection fraction (EF, 35%). C, CW Doppler curve: restrictive type of the mitral inflow pattern with short deceleration of E wave and E/A >2.

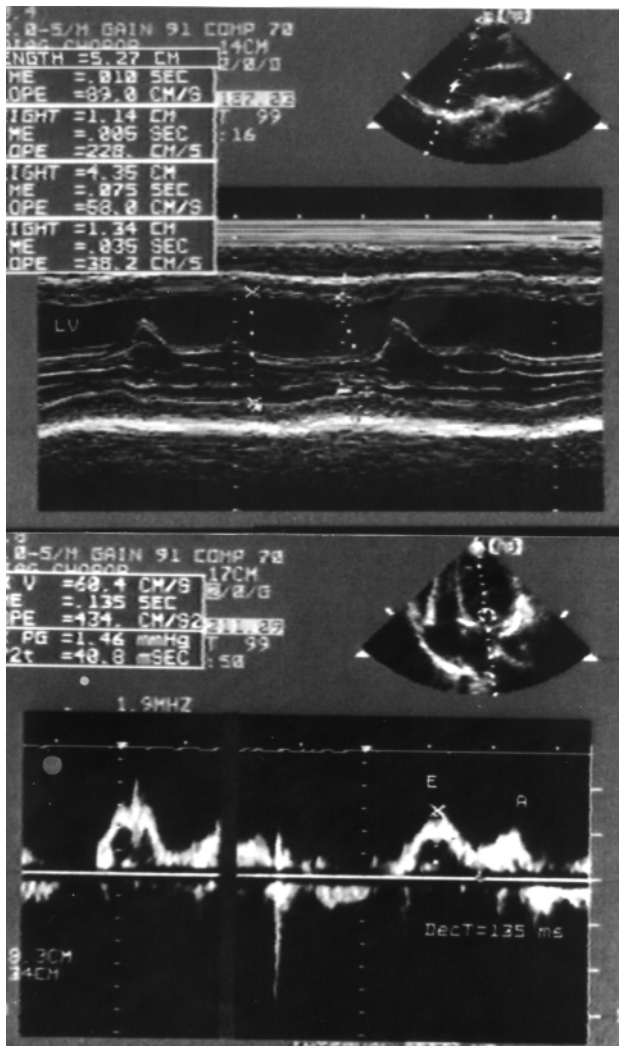

FIGURE 2 Case 1: Echocardiographic examination four days later. A, M-mode normal diastolic diameter of the left ventricle and improved ejection fraction (EF) – 47%. B, Mitral inflow pattern - longer deceleration time of E wave and E/A <2.

suggestive of emboli within the pulmonary arteries. Doppler sonography of the deep veins of the lower limbs and pelvis did not show any signs of deep venous thrombosis (DVT).

On admission to the ICU the patient was conscious, severely dyspneic and tachypneic. During the next hour she became restless, arterial blood gases showed a pO_2 of 6.05 kPa (46 mmHg) and a pCO_2 of 3.42 kPa (26 mmHg) and she required intubation and mechanical ventilation.

At this stage echocardiography revealed left ventricular (LV) enlargement with severe systolic dysfunction, an ejection fraction (EF) of 25%, mild mitral regurgitation, a restrictive mitral inflow pattern (deceleration time of E wave 90 msec) and borderline pulmonary hypertension calculated from a small tricuspid jet of regurgitation (Figure 1 A–C). PPCM was diagnosed on the basis of echocardiography. The patient received digoxin, captopril, furosemide, aldactone and beta-blockers. Captopril was temporarily changed to nitroprusside and ketanserin to improve peripheral perfusion. Heparin was continued in lowered doses to reduce the possibility of thrombembolic complications. After three consecutive days the patient's gas exchange improved, ventilatory support was stopped and her trachea was extubated. At this moment echocardiography showed a normal diastolic LV diameter with moderate improvement of systolic LV function (EF 45%) and longer deceleration time of E wave (120 msec; Figure 2 A–C). On the sixth day after admission the patient was discharged from the ICU to the maternity clinic. TTE performed a week later showed a normal LV diameter with slightly impaired EF (50%) with no signs of mitral regurgitation and normal pulmonary artery pressures.

Case 2

A 29-yr-old previously healthy primigravida, primipara was discharged home three days after an uncomplicated spontaneous delivery in good physical condition. Two days later she was readmitted for observation to the maternity ward with a history of gradually increasing dyspnea. She had been receiving fenoterol from 32 to 38 weeks of pregnancy.

Within five hours from admission she progressed to respiratory insufficiency with a pO_2 of 5.76 kPa (43.8 mmHg) and a pCO_2 of 2.9 kPa (22.1 mmHg). She was intubated and full ventilatory support was initiated with FIO_2 0.5 and PEEP +8 cmH_2O. The chest radiograph was suggestive of congestion. She was given digoxin, furosemide and a bolus of *iv* heparin and seemed to be temporarily stabilized. Before transferring the patient to the ICU, cardiac arrest by ventricular fibrillation occurred. Treatment with defibrillation (x 2), adrenaline 3 mg, atropine 1 mg and lidocaine 100 mg *iv* restored sinus rhythm.

Soon after admission to the ICU another cardiac arrest occurred and the patient was again resuscitated successfully. Thoracotomy for pulmonary embolectomy was considered as a therapeutic option. The patient was too unstable to be transported for a CT scan. Bedside echocardiography revealed a LV systolic diameter above normal, a severely impaired systolic function with EF 22% and no signs of pulmonary hypertension. Supportive treatment for left ventricular

failure (LVF) was started consisting of captopril, digoxin, dopamine as required, furosemide, spironolactone and nitroglycerin.

Weaning from respiratory support was complicated by pulmonary edema due to LVF after the first trial of extubation. Mechanical ventilation was finally discontinued on the fifth day after admission. Four days later, the EF was 45% and the mitral inflow pattern remained abnormal. Echocardiographic examination three weeks later showed almost normal diameters of the left ventricle with an EF of 56%.

Discussion

Although a relatively rare complication of late pregnancy and puerperium – occurring in one out of 3000–4000 live births - PPCM requires attention for several reasons.[1] The disease touches young, previously healthy women at one of the most important moments of their lives. The mortality is high, estimated as 4% of maternal deaths in the U.S. each year,[2] and the prognosis for survivors remains at least uncertain. If the LV function does not return to normal, quality of life deteriorates seriously. Heart failure sometimes progresses to the point that cardiac transplantation remains the only option. Even if the hemodynamic status improves, future pregnancies are questionable and best avoided,[3] as the contractile reserve is lowered[4] and the recurrence of PPCM has been reported.[1] The disease also has a strong association with thromboembolic incidents[5] and can be complicated by multiorgan failure.[6,7]

Although some factors seem to correlate with the disease, e.g., silent myocarditis, autoimmune or idiopathic processes, their contribution to the etiology and pathogenesis of the disease remains hypothetical. An article published recently by Pearson *et al.* briefly summarises the actual state of knowledge.[1]

Potential risk factors for this condition are not well defined. Those described (multiparity, multifetal pregnancy, advanced maternal age, pregnancy induced hypertension and tocolytic therapy) are too widely distributed in the maternal population to allow screening. Classically, PPCM has been diagnosed when heart failure occurs within a month before or five months after delivery, without previous evidence of cardiac dysfunction and with no other identifiable cause. It is only recently that echocardiographic findings were incorporated into the diagnostic criteria of PPCM,[1] although echocardiography had already been strongly recommended before.[8–10]

Establishing an accurate diagnosis of PPCM rapidly is not always easy. First complaints are non-specific and misleading. Increasing fatigue is a feature of late pregnancy and puerperium. Dyspnea is associated with many other common disease states ranging from pneumonia and bronchial asthma to PE.

Uniform opacity of the right lung, treated initially as massive pneumonia in case 1 might also have been a unilateral pulmonary edema. Three similar cases of unilateral pulmonary edema have been described recently as caused by severe mitral regurgitation directed towards right pulmonary vein.[11] Echocardiographic findings in our first patient also included mitral regurgitation secondary to dilatation of the left ventricle.

It has already been reported in the literature that PE was diagnosed instead of PPCM.[8,12] It is widely stressed that the natural hypercoagulability of pregnancy can predispose to thrombembolic complications.[13] As massive PE is the leading cause of maternal death[14] it is the most feared diagnosis and other diseases like PPCM are easily forgotten. However, distinguishing between the two is vital for the patient. In both, progression of symptoms is rapid and can lead to death in a short time from the initial presentation. Management of PPCM is quite different from that of PE. Treatment of PPCM is conservative and concentrates on supporting LV function, leaving time for the symptoms to resolve spontaneously. PE requires more aggresive management. High doses of *iv* heparin remain the treatment of choice for DVT with or without PE. Although they are relatively safe for the fetus (heparin does not cross the placental barrier) the mother is at risk of bleeding complications, especially after delivery. Massive PE can be treated only with urgent surgery and the chances of survival are low.

Sometimes the condition of the patient does not allow for procedures like pulmonary angiography, ventilation/perfusion scintigraphy or spiral CT. Echocardiography has already been described as a useful diagnostic procedure to differentiate between PE and PPCM. Both our cases support this recommendation.

Echocardiographic findings in PE include right ventricular dilatation and a circular shape of the right ventricle in the short axis with paradoxical interventricular septum motion and pulmonary hypertension calculated from tricuspid regurgitation. Sometimes thrombus is visualized by TTE[15] and more frequently by transoesophageal echocardiography.[16,17]

The echocardiographic manifestations of dilated cardiomyopathy are LV dilatation with an end diastolic diameter >2.7 cm/m^2, a low EF (<30%) and mitral regurgitation due to mitral annulus dilatation.[10] Mild pulmonary hypertension and an abnormal - restrictive - mitral inflow pattern may be found using Doppler method.

Because of its rarity, PPCM is considered late in the differential diagnosis, as happened in both of cases

described. Some authors stress the fact that early diagnosis and treatment improves the prognosis of PPCM.[9,18]

Echocardiography can also provide prognostic information. Patients who deteriorate have higher LV end- diastolic diameters as compared to those who improve.[19,20] The response to a dobutamine challenge test assessed echocardiographically is also diminished, even when routine parameters return to baseline.[4]

Conclusion

Initial presentation of PPCM can resemble many other emergency conditions, including PE. In the cases of acute postpartum respiratory and/or circulatory failure, TTE should be considered a first-line diagnostic tool. TTE is noninvasive, can be performed at the bedside and is extremely helpful in assessing cardiac function and establishing the appropriate diagnosis in this context.

References

1 *Pearson GD, Veille J-C, Rahimtoola S, et al.* Peripartum cardiomyopathy. JAMA 2000; 283: 1183–8.
2 *Veille J-C, Zaccaro D.* Peripartum cardiomyopathy: summary of an international survey on peripartum cardiomyopathy. Am J Obstet Gynecol 1999; 181: 315–9.
3 *Witlin AG, Mabie WC, Sibai BM.* Peripartum cardiomyopathy: an ominous diagnosis. Am J Obstet Gynecol 1997; 176: 182–8.
4 *Lampert MB, Weinert L, Hibbard J, Korcarz C, Lindheimer M, Lang RM.* Contractile reserve in patients with peripartum cardiomyopathy and recovered left ventricular function. Am J Obstet Gynecol 1997; 176: 189–95.
5 *Brown CS, Bertolet BD.* Peripartum cardiomyopathy: a comprehensive review. Am J Obstet Gynecol 1998; 178: 409–14.
6 *Yahagi N, Kumon K, Nakatani T, et al.* Peripartum cardiomyopathy and tachycardia followed by multiple organ failure. Anesth Analg 1994; 79: 581–2.
7 *Kluger MT, Bersten AD.* Multi-organ failure in peripartum cardiomyopathy. Anaesth Intensive Care 1991; 19: 450–3.
8 *Chan L, Hill D.* ED echocardiography for peripartum cardiomyopathy. Am J Emerg Med. 1999; 17: 578–80.
9 *Heider AL, Kuller JA, Strauss RA, Wells SR.* Peripartum cardiomyopathy: a review of the literature. Obst Gynecol Surv 1999; 54: 526–31.
10 *Hibbard JU, Lindheimer M, Lang RM.* A modified definition for peripartum cardiomyopathy and prognosis based on echocardiography. Obstet Gynecol 1999; 94: 311–6.
11 *Lesieur O, Lorillard R, Ha Thi H, Dudeffant P, Ledain L.* Unilateral pulmonary oedema complicating mitral regurgitation: diagnosis and demonstration by transoesophageal echocardiography. Intensive Care Med 2000; 26: 466–70.
12 *Aroney C, Khafagi F, Boyle C, Bett N.* Peripartum cardiomyopathy: echocardiographic features in five cases. Am J Obstet Gynecol 1986; 155: 103–6.
13 *Woodhams BJ, Candotti G, Shaw R, Kernoff PB.* Changes in coagulation and fibrynolysis during pregnancy: evidence of activation of coagulation preceding spontaneous abortion. Thromb Res 1989; 55: 99–107.
14 *Toglia MR, Weg JG.* Venous thromboembolism during pregnancy. N Engl J Med 1996; 335: 108–14.
15 *Dudziak M, Wojtowicz A, Mierzejewski L, Emerich J, Rynkiewicz A.* Pulmonary artery thrombus visualized in transthoracic echocardiography in women in the 7th month of pregnancy. Polish Heart Journal 2000; 52: 246–7.
16 *Torbicki A.* Echocardiography in pulmonary embolism. *In*: Mopurgo M (Ed.). Pulmonary Embolism. Vol. 75 Lung biology in health and disease. New York: Marcel Dekker, 1994: 153–78.
17 *Rosenberg JM, Lefor AT, Kenien G, Marvasti M, Obeid A.* Echocardiographic diagnosis and surgical treatment of postpartum pulmonary embolism. Ann Thorac Surg 1990; 49: 667–9.
18 *Leonard RB, Schwartz E, Allen DA, Alson RL.* Peripartum cardiomyopathy: a case report. J Emerg Med 1992; 10: 157–61.
19 *Ravikishore AG, Kaul UA, Sethi KK, Khalilullah M.* Peripartum cardiomyopathy: prognostic variables at initial evaluation. Int J Cardiol 1991; 32: 377-80.
20 *Witlin AG, Mabie WC, Sibai BM.* Peripartum cardiomyopathy: a longitudinal echocardiographic study. Am J Obstet Gynecol 1997; 177: 1129–32.