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Meadows Syndrome as Manifested by Displaced Ischemia of the Lower Limb

Diarra BI[1*], Doumbia M[1], Toure M, Djiguiba K[4], Keita A[2], Daffe S[1], Coulibaly B[1], Doucoure O[1], Diallo B[1], Traore S[1], Coulibaly M[1], Koita S[1], Diarra MB[2] and Yena S[2]

[1]Cardio-Pediatric Andre Festoc Center, Mother-Child Hospital Luxembourg, Mali

[2]Department of Cardiology, Mother-Child Hospital Luxembourg Bamako, Mali

[3]Faculty of Medicine and Odontostomatology, University of Technical Sciences and Technologies of Bamako, Mali

[4]Department of Nephrogy, Mother-Child Hospital Luxembourg Bamako, Mali

[*]Corresponding author:

Baba Ibrahima Diarra,
Cardio-Pediatric Andre Festoc Center,
Mother-Child Hospital Luxembourg, Mali

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1. Summary

Peripartum cardiomyopathy (PPMC) or Meadows syndrome is recognised as a major cause of pregnancy-related heart failure with high morbidity and mortality. It is a condition of unknown aetiology that manifests itself as heart failure due to systolic dysfunction of the left ventricle during the last month of pregnancy and up to 5 months after delivery. Thromboembolic and ischaemic complications are common. We report a case of peripartum cardiomyopathy in a 19-year-old primigravida patient, primiparous, with no known personal or family history of heart disease, who presented with extensive ischaemia of the left lower limb, bilateral renal and splenic infarction in a context of dyspnoea, abdominal pain and pain of the left lower limb occurring one month after delivery. Cardiac Doppler ultrasonography showed very severe impairment of left ventricular systolic function with a left ventricular ejection fraction of 25% associated with hypokinetic dilated cardiomyopathy. An angioscan of the abdomen and lower limbs showed a renal and splenic infarction and a thrombus in the common femoral artery. The electrocardiogram showed left ventricular hypertrophy and a diffuse repolarisation disorder. His management consisted of a left femoral trans amputation, and medical treatment with a diuretic, a conversion enzyme inhibitor and an anticoagulant. The course was marked by suppuration of the stump, which progressed well with antibiotic therapy and local care.

2. Introduction

Peripartum cardiomyopathy (PPCC) is recognised as a major cause of pregnancy-related heart failure, with high morbidity and mortality [1]. In severe forms (10-15% of cases), thrombo-embolic and ischaemic complications are the hallmark of the disease [2-5]. We report a particular clinical form of ischaemia of the left lower limb and a bilateral renal and splenic infarction which was managed medically and surgically.

3. Observation

This 19-year-old patient was initially admitted to a local hospital for NYHA stage 4 dyspnoea, a dry cough and oedema of the lower limbs 7 days after vaginal delivery. The delivery took place in the context of severe pre-eclampsia, requiring unsuccessful consultations at local facilities. The evolution was marked 3 weeks later by the sudden onset of abdominal pain followed by severe pain in the left lower limb. His general condition deteriorated rapidly, with a fall in blood pressure (95/60 mm Hg), significant tachycardia (134 bpm), tachypnoea (28 cycles/min), temperature of 36.7°C, and saturation of 99% AA.

Cardiac auscultation revealed a regular tachycardia with a galloping sound.

Comparative examination of the lower limbs revealed coldness and cyanosis on the left, with a weak femoral pulse; the popliteal,

posterior tibial and pedal pulses were all absent.

Biological tests: creatinine: 76.68 umol/l, blood glucose: 6.30 mmol/l, blood ionogram, transaminases and prothrombin level were normal. HIV serology was negative, as were those for hepatitis B and C; the emmel test was negative. Haemoglobin was 9.8 g/dl.Chest X-ray showed global cardiomegaly (cardiothoracic index = 0.53) with no infectious pulmonary parenchymal process or radiological pleuropathy.

The electrocardiogram showed regular sinus tachycardia at 134 bpm, left ventricular hypertrophy and a diffuse repolarisation disorder.

Cardiac ultrasound revealed hypokinetic dilated cardiomyopathy with a severely impaired left ventricular ejection fraction, elevated filling pressures and apical intra-left ventricular thrombus (Figure 1).

CT angiography:

At the thoracic level, there was a thrombus in the left ventricle measuring 14x09 mm (Figure 2 and 3).

At the abdominal level, there were multiple foci of bilateral renal infarcts and a splenic infarct (Figure 4and 5).

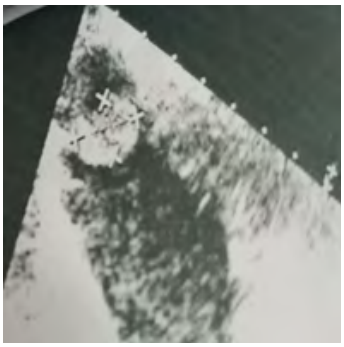

Figure 1: Left ventricular thrombus on cardiac echography

Figure 2: Thoracic angiogram (mediastinal window) Left apical thrombus

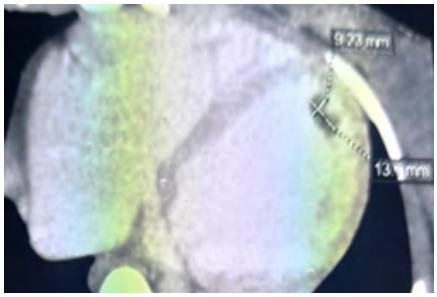

Figure 3: Thoracic angiogram (parenchymal window) right lower lobar pulmonary parenchymal infarction

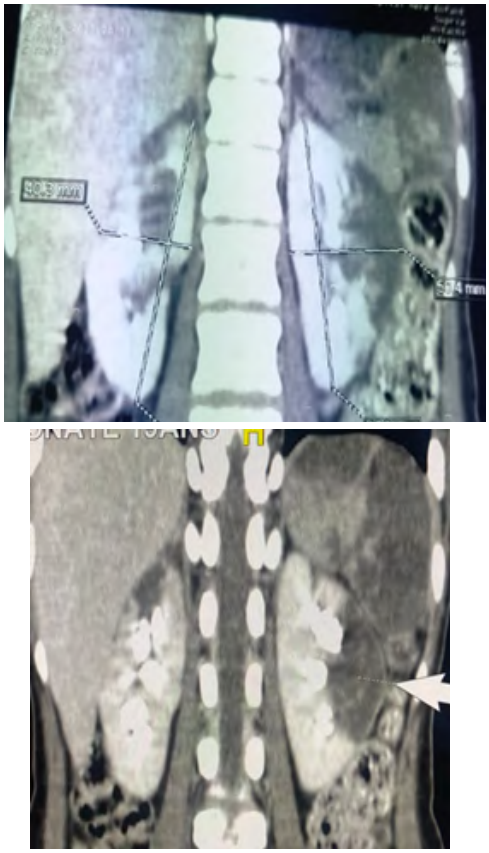

Figure 4 and 5: Angioscan (frontal section) bilateral renal infarction

4. Discussion

Peripartum cardiomyopathy (PCM) is a rare form of dilated cardiomyopathy associated with high maternal morbidity and mortality, accounting for 4% of maternal deaths in the United States [1, 2]. It is defined according to the following four criteria:

1) Development of heart failure during the last month of pregnancy or within five months of delivery;

2) No identifiable cause of heart failure;

3) Absence of recognisable heart disease before the last month of pregnancy;

4) Left ventricular systolic dysfunction demonstrated by echocardiographic criteria [1, 3].

Its incidence is between 1/1500 and 1/4000 live births [4], with wide geographical variation. The mortality rate is estimated at between 20% and 50%, with death occurring as a result of progressive heart failure, arrhythmias and thromboembolism [5].

Several risk factors for peripartum cardiomyopathy have been identified: maternal age > 30 years, multiparity, multiple pregnancy, obesity, arterial hypertension, pre-eclampsia, prolonged tocolysis [3].

The risk factors observed in our patient were arterial hypertension and pre-eclampsia.

The arterial time of the lower limbs showed extensive acute occlusion of the common femoral artery as far as the tripod on the left,

associated with acute occlusion of the right posterior tibial artery. There was also partial occlusion of the proximal 1/3 of the deep femoral artery and the junction of the popliteal artery and the right anterior tibial artery.

Management consisted of a trans-femoral amputation (precisely at the upper 1/3 of the thigh) and medical treatment based on diuretics, conversion enzyme inhibitors and anticoagulants. The evolution was marked by suppuration of the amputation stump, which improved well with local care and triple antibiotic therapy based on Gentamicin, Metronidazole and Amoxicillin clavulanic acid. She had her stump fitted 3 months after her amputation.

Peripartum cardiomyopathy initially presents with signs and symptoms of heart failure and rarely with thromboembolic complications [3, 6, 8].

Our patient's heart failure occurred in the immediate postpartum setting and no other aetiology had been identified.

Her symptoms were typical of a dry cough, dyspnoea, orthopnoea and eodema of the lower limbs immediately post-partum. The cardiac ultrasound had made a positive diagnosis by measuring the systolic function, with an ejection fraction estimated at 25%. It also noted the presence of an apical thrombus in the left ventricle (Figure 1), which easily explains the thromboembolic complications in the renal, splenic and femoral arteries, with the occurrence of extensive ischaemia of the left lower limb, a renal infarction and a splenic infarction.

This thromboembolism in peripartum cardiomyopathy is thought to be due to a combination of several conditions, including the hypercoagulable state of pregnancy linked to hormonal changes persisting for several weeks after delivery, dilatation of the cardiac chambers and inflammatory enthothelial lesions [7]. In addition, the fall in cardiac output observed during hypokinetic cardiomyopathy, including peripartum cardiomyopathy, is more conducive to all types of thrombosis, including intracardiac thrombus, arterial embolism and delayed limb ischaemia [11, 12]. In our patient, this was illustrated by a pulmonary and renal infarction and protracted ischaemia of the lower limb.

This justifies the initiation of full-dose anticoagulant therapy in the presence of severe left ventricular dysfunction [9].

Several complications may be observed during peripartum cardiomyopathy, including heart failure including cardiogenic shock, cardiac arrhythmias and thromboembolic complications as in our patient. These thromboembolic complications have an incidence ranging from 6.6% to 50% in the United States according to the prospective study by Avila et al [9, 10].

Our patient's electrocardiogram showed only sinus tachycardia and a diffuse repolarisation disorder with no arrhythmia predisposing to thromboembolism.

Some authors have reported the case of a 37-year-old multiparous woman with peripartum cardiomyopathy who developed ischaemia of the lower limbs and presented with a rhythm disorder of the atrial fibrillation type [8].

The management of thromboembolic complications associated with peripartum cardiomyopathy is similar to that of other forms associated with other aetiologies [5]. However, our patient presented with advanced ischaemia of the left lower limb, for which trans-femoral amputation was the only option, together with curative anticoagulation.

The standard treatment for heart failure indicated in peripartum cardiomyopathy had already been undertaken, aimed at reducing preload and afterload and improving myocardial contraction [5]. It was combined with bed rest and fluid restriction.

Ischaemia of the lower limb and renal and splenic infarction in our patient were suspected on the basis of a number of clinical (acute abdominal pain, sharp pain in the left lower limb, absence of pulses, coldness and cyanosis) and paraclinical (thrombus of the left common femoral artery on angioscanner) factors.

Early recognition of these complications can improve management.

5. Conclusion

Peripartum cardiomyopathy is a special pathological entity, due to its target population (women of childbearing age who are genitally active) and its context.

Their management requires close collaboration between cardiologists, gynaecologists, paediatricians, anaesthetists and vascular surgeons or traumatologists. Prevention can be envisaged if certain factors are effectively managed in this target population during antenatal consultations.

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