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Case Report Open Access

Fatal Acute Necrotizing Eosinophilic Myocarditis during Peripartum: Case Report

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Abstract

Peripartum heart disease is a rare group of disorders developing during the last trimester of pregnancy through the first 6 months of postpartum. The etiology and pathogenesis of Peripartum Cardiomyopathy (PPCM) is unknown but viral and autoimmune causes may contribute. Myocardial involvement includes myocarditis, coronary artery dissection and peripartum cardiomyopathy. Risk factors for PPCM include advanced maternal age, multiparity, African race, twin birth, gestational hypertension and long-term tocolysis. We report the case of a 29-year-old pregnant, that after the delivery at the 37th week of gestational age, complained weakness and abdominal pain. The young woman showed since the third day postpartum worsening chest pain associated with malaise, non-specific symptoms, and increase in inflammatory markers without peripheral eosinophil increase. Physicians, suspecting a gastric disorder, treated her with antiacids and proton pump inhibitors, and advised gastroenterology consulting. The patient entered in emergency room in critical conditions, with severe left ventricular systolic dysfunction and an ejection fraction of 25%. She died 23 day after delivery.

Learning Objective

Acute eosinophilic myocarditis is a medical emergency that is usually fatal and the diagnosis is difficult before a patient's death. Frequently the cause of the disease remains unknown, but cardiac involvement occurs in more than 50% of patients with idiopathic hypereosiniphilic syndrome.

Case Report

A 29-year-old woman was admitted to critical care unit in respiratory and cardiac failure, just three weeks after giving birth at the 37th week of gestational age. Patient clinical history was silent for allergy or autoimmune diseases. The third day after delivery she complained of chest pain but echocardiogram was negative. During hospitalization physicians treated her with antiacids and gastric inhibitors and then she was discharged with prescription of proton pump inhibitors and suggestion of gastroenterological visit. The following three weeks were characterized by growing front and back chest pain associated with general discomfort, but neither with specific symptoms or peripheral eosinophils increasing. Only inflammatory indexes (velocity of erythrocyte sedimentation, VES, and creatine phosphokinase, CK) were slightly increased (highest value of CPK was 353 U/l and the highest of VES was 120 mm/h). Subsequently, because of the progressive clinical worsening, she was finally sent to Emergency Room in critical conditions: dyspnoea, confusion, fever, and tachycardia. Echocardiogram showed severe left ventricular systolic dysfunction and an ejection fraction of 25%; chest radiography and TC displayed pleural effusion with general oedema. The young woman died after 23 days from delivery and seven hours of cardio-respiratory failure, and no medical approach was effective. External body examination was completely negative. Autopsy revealed bilateral pleural effusion, increased volume of the lungs and hepatomegaly. Heart weighed 300 gr. It was normal in size, shape, and consistency, but myocardium and papillary muscles showed malacic areas (Figure 1).

Figure 1: Section of heart show edmalacic areas and yellowish areas intermingled with pale red ones at myocardium and papillary muscles level.

On cut surface white-grey areas were intermingled with pale red ones involving the full thickness of ventricular chambers walls. Mural thrombi were absent. Examination of the valves and coronary arteries was unremarkable. Microscopic examination showed a diffuse myocardial infiltrate with the association of cardiac necrosis and initial myocardial fibrosis. The infiltrate was composed largely by normal eosinophils, but there were some lymphocytes and monocyte-macrophage cells (Figure 2). In focal areas there were micro abscesses

consisting of eosinophils, polymorphonuclear cells, and occasional giant cells. There were no granulomata and no arteritis. Other necropsy findings included the presence of septal capillaries and interstitial vessels oedema associated to small eosinophils aggregates in a peribronchial and intraalveolar location of the lung. The liver histology showed scattered eosinophils at sinuisodal level. On the base of the histological findings a diagnosis of peripartum eosinophilic myocarditis was made. Systemic and pulmonary diseases were excluded.

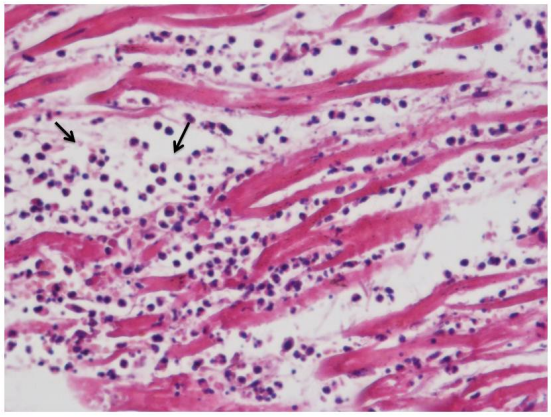

Figure 2: Microscopic photographs of section of ventricular myocardium (magnification 40X.) Histological sections showed a diffuse inflammatory infiltration made predominantly by eosinophils.

Discussion

Cardiac involvement in eosinophilia has been well studied. The known causes of eosinophilia are distinguished in specific and idiopathic. Among the latest it is useful to mention eosinophilia of unknown aetiology, which has also been reported as hypereosinophilic syndrome (HES) [1].

The fulminant type of acute early necrotizing eosinophilic myocarditis has abrupt onset and occurs in the absence of a notable extra cardiac disease. It has very rarely been reported in literature, and the prognosis is usually very poor as it is rapidly fatal. Heart failure with unknown aetiology which occurs in the last month of pregnancy or within five months after delivery has been recognized as peripartum cardiomyopathy. Since this disease is quite uncommon, its aetiology has not yet been clarified [2]. Eosinophils are present in a significant subset of peripartum cadiomyopathy cases (both with and without myocarditis) as well as in peripartum spontaneous coronary dissections. Since eosinophils are often absent, they represent a reactive, secondary phenomenon, rather being etiologically connected [3]. There are many theories that have been proposed to elaborate the hypothetical mechanism for the pathogenesis of this clinical entity. These hypotheses regard interactions of peripartum physiology with inflammatory, infective, autoimmune, metabolic, hormonal, biochemical stimuli and genetic factors [1]. EM is rarely clinically recognized and is often first discovered at postmortem examination. Studies have reported EM in 0.5% of unselected autopsy series, and in more than 20% of explanted hearts from heart transplant recipients [4]. The cause is unknown, but various causes have been suggested. It occurs in the range of 1/300 to 1/15000 pregnancies, with increasing incidences associated with advanced age, multiple pregnancies and twins. Symptoms and signs accompanying EM are nonspecific and include fever, skin rashes, sinus tachycardia, conduction delays and ST-T wave abnormalities,peripheral blood eosinophilia, with dyspnoea, fatigue and peripheral oedema – symptoms which are identical to early congestive cardiac failure – but these are not necessarily seen in all cases of EM, and thus, the diagnosis is often not clinically suspected [4]. From 20 to 40% of these patients demonstrated to have cardiac dysrhythmias [2]. The characteristic histopathology of EM is a mixed inflammatory cell infiltrate containing a variable amount of eosinophils within the myocardium. Myocardial infiltrations present different geographic distributions, such as perivascular or interstitial. As well as myocardium, this cardiomyopathy may involve epicardium and endocardium [4]. The localization of eosinophils in the cleavage plane and the existence of tissue damaging eosinophil granule components has been used as evidence to implicate eosinophils in the process of coronary dissection [3]. There is no apparent relationship between the entity of the eosinophilic infiltrates in EM and clinical symptoms.Myocyte necrosis, fibrosis, granuloma formation and fibrinoid necrosis of collagen are also observed as additional features in this disease [4]. There are many hypothesis about EM pathogenesis. The eosinophils presence in postpartum cardiac disease may be a systemic reflection of events that are taking place in uterine involution. Eosinophils migration and diapedesis occur, and localization in coronary arteries and myocardium may be a phase of a normal process or may represent an idiosyncratic pathological response. Subsequent tissue damage may be due to inappropriate eosinophilic degranulation, release of collagenase or major basic protein following restoration of steroid hormones after uterine involution, or may result from circulating collagenases sensitized by the eosinophils that are measurable postpartum and act on tissues [3]. Acute necrotising eosinophilic myocarditis is arare idiopathic disorder that present with fulminant or acute heart failure, which is frequently associated with ventricular arrhythmias or heart block. Prognosis is strictly linked to ventricular function recovery because those patients with severe myocarditis-induced heart failure have less survival chances if normal cardiac function is not restored [4-7]. Few EM cases are reported in literature and most of them is based only on autopsy diagnosis. Significantly, the predominant pathological process was limited to the heart, and the degree of peripheral eosinophilia did not necessarily correlate with the degree of cardiac damage [8].

Conclusions

In the contest of peripartum cardiomyopathy, eosinophilic myocarditis diagnosis is not simple because of overlaps in clinical presentation, non-specific symptoms and pathological findings. According to these premises, very often clinicians fail in recognizing EM that is then first discovered only at postmortem examination [5]. A timely and correct diagnostic approach in these patients include an echocardiogram study (with evidence of low ejection fraction and decreased left ventricular systolic function) and an endomyocardial biopsy (confirming eosinophilic infiltrates as a major inflammatory component) and should save patient life through management of cardiopulmonary failure. EM can be treated with steroid therapy to reduce the inflammatory process and beta-blockers and ACE inhibitors to support heart failure and recovery heart function.Medical

effort is necessary to avoid these unexpected and unexplained deaths immediately preceding and following delivery.

Conflict of Interest

The authors certify that they have no affiliations with or involvement in any organization or entity with any financial interest or non-financial interest in the subject matter or materials discussed in this manuscript.

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