

Case Report

Diagnostic-therapeutic management of a chronic rheumatic heart disease in pregnancy

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A 37 year old Asian pregnant woman referred to our outpatients facility at the 31st gestational week with a few days history of asthenia, dyspnea by walking and tachycardia in the early morning by waking up. The objective examination revealed elevated heart rate (80beats/min), arrhythmia and abnormal respiratory rate (20 breaths/min). The urgent haemogram showed hypochromic anaemia (Hb 10.0 g/dL), erythrocytopenia ($3.79 \times 10^6/\mu\text{L}$) and normal leukocytes ($8.06 \times 10^3/\mu\text{L}$). The cardiology evaluation with echocardiography found severe atrial fibrillation, mitral stenosis and pulmonary hypertension associated with severe bi-atrial dilatation. In relation to the patient conditions, an intensive heart monitoring was started. The patient was treated for respiratory distress syndrome prophylaxis (Betametasone 12 mg i.m. x2) and Low Molecular Weight Heparin prophylaxis (Seleparine 6000 UI tid). Initially, there was a successful pharmacological cardioversion, but after some days atrial fibrillation reappeared (150 bpm). At the 33 gestational week, after a multidisciplinary discussion, cesarean section was performed. Five days after delivery the patient developed anaemia, fever, leukocytosis and C-reactive protein increase. Active treatment included haemotransfusion, wide spectrum antibiotics and dicumarolic prophylaxis, and forty days after delivery the patient was successfully discharged with good health conditions.

Keywords: Chronic rheumatic heart disease, mitral stenosis, pulmonary hypertension, atrial fibrillation arrhythmia, pregnancy.

Abbreviations list

HR: heart rate

AP: arterial blood pressure.

INTRODUCTION

Cardiac diseases affect the 0.5-1% of pregnancies, being an important cause of death during pregnancy (Ayoub et al., 2002), and their incidence is continuously rising because the current medical treatments allow more females with rheumatic heart disease or congenital heart defects to reach childbearing age.

Pregnancy per se determines significant hemodynamic changes which have a different impact depending on the type and severity of cardiac anomalies (Ayoub et al., 2002). The diseased heart is sometimes unable to meet the normal demands of pregnancy: 25% increase in

increased oxygen requirements, retention of salt and water, weight gain, and alterations in hemodynamics during delivery. This physiologic stress for the maintainment of an adequate circulation often leads to the heart failure. The severity of the pathology depends on patient age, cardiac disease onset, and heart functional capacity by pregnancy start.

Rheumatic fever is the consequence of a previous group A haemolytic streptococcal infection, which may be cured or evolve in a serious deformation of the cardiac valves (more often the mitral valve). About 12 million people are affected by acute rheumatic fever worldwide, and about 400,000 of them develop a rheumatic heart disease, which accounts for the 25-40% of all cardiac

cardiac output, 40% increase in plasma volume, diseases worldwide.

Acquired rheumatic heart disease in some countries (i.e. New Zealand, Tunisia, Africa, Latin America, South Asia) represents the most common heart disease of pregnancy, while in western industrialized countries (characterized by low natality) it has declined and the most common cause of heart disease in pregnancy has become the congenital heart diseases (Wilson, 2010; Westhoff-Bleck et al., 2008). Rheumatic fever sequelae should be considered during pregnancy, especially in countries with new immigration from high risk regions (Saraiva et al., 2009; Chopra & Gulwani 2007).

The general incidence of rheumatic fever and consequent endocarditis during pregnancy is reported as low (0.006%). General maternal mortality rate, after endocarditis complication, is very high (33%), particularly related to heart failure or to embolic events; while foetal mortality can reach 29% (Vizzardi et al., 2009; Lin et al., 2007).

Drug therapy of arrhythmia in pregnancy is limited by side effects on the foetus, and the best management requires a multidisciplinary team of obstetricians, neonatologists, cardiologists, anaesthesiologist and sometimes cardiac surgeons, to optimize maternal and foetal outcomes.

The aim of our case report is to present the diagnostic-therapeutic management of a chronic rheumatic heart disease in pregnancy complicated by atrial fibrillation and pulmonary hypertension, in order to evidence the importance of heart diseases in childbearing age and the need of a prompt pre-gestational diagnosis.

CASE PRESENTATION

A 37-years-old pregnant woman, coming from a small island of the Pacific Ocean, with 2 previous vaginal deliveries and 1 miscarriage, applied to our obstetrics outpatients facility at the 31th gestational week with a few days history of asthenia, dyspnea by walking and tachycardia in the early morning by waking up. Vital signs at admission revealed AP 90/60 mmHg, HR 80 bpm with arrhythmia, normal body temperature (36.8 C) and respiratory rate of 20 breaths/min. During the visit we collected the following data: 57 kg weight (52 kg pre-gestational weight), 150 cm high, no-smoking, no-drinking, no previous surgical intervention, normal routine laboratory exams and a good foetal biometry up to date.

The patient did not report any symptom of Streptococcal infection (i.e. polyarthritis, skin disorders) in the previous few weeks or during the following hospitalization. She also had no previous cardiologic or echocardiographic evaluation, nor radiographic evaluation of the thorax. She presented varicose veins at both legs, so that the last surgical evaluation suggested heparin prophylaxis for at least 20 days before the expected delivery date.

The urgent hemogram showed hypochromic anemia

(Hb 10.0 g/dL), erythrocytopenia ($3.79 \times 10^6/\mu\text{L}$), and normal leukocytes ($8.06 \times 10^3/\mu\text{L}$). Obstetrical ultrasound examination showed an appropriate for gestational age foetus in vertex presentation. The electrocardiogram revealed a severe atrial fibrillation. The echocardiography confirmed the severe uncontrolled atrial fibrillation, demonstrating also a mitral stenosis and a pulmonary hypertension associated with severe bi-atrial dilatation.

In relation to the patient conditions, an intensive heart monitoring was started. The patient was treated for respiratory distress syndrome prophylaxis (Betametasone 12 mg i.m. x2), and assumed also Atosiban against the painful contractions caused by Betametasone, and Low Molecular Weight Heparin prophylaxis (Seleparine 6000 UI tid). The pharmacological cardioversion in Cardiac Intensive Care Unit (Lanoxin 0.125, Heparin 25.000 UI ev and Metoprololo 50 mg) was initially successful, but after some days atrial fibrillation reappeared (150 bpm).

At the 33th gestational week, after a multidisciplinary discussion with neonatologist, cardiologist and anaesthesiologist, an elective cesarean section was performed, and the patient gave birth to a well being female newborn.

Eight days after delivery, during the hospitalization period, the patient developed anaemia with an important decrease of haemoglobin (Hb 6.9 g/dL, RBC $2.46 \times 10^6/\mu\text{L}$), fever (40 C), leukocytosis (WBC $16.49 \times 10^3/\mu\text{L}$) and increase of C-reactive protein (205 mg/L). Blood cultures and microbiological examination of cesarean-scar tampons were negative. Echodopplercardiography evaluation showed a minimal pericardial effusion around the left atrium.

Following treatment included haemotransfusion and dicumarolic prophylaxis (5 mg/die). Wide spectrum antibiotics (Ceftazidime 2 gr every 8 hours/day then substituted by Meropenem 500mg every 6 hours/day, Teicoplanine 400 mg every 12 hours/day and Fluconazole 200 mg/day) were administered for 19 days according to the infectivologist prescription. Antistreptolisine titer measure was in range. Finally, 40 days after delivery the patient was discharged in good health conditions, and at the 3-month follow-up visit was still in good clinical conditions.

DISCUSSION

Rheumatic disease in pregnancy is very rare in Europe: in fact, acute rheumatic fever with carditis has been uncommon for many years, and chronic rheumatic heart disease, which presents years to decades after the episode of acute rheumatic fever, is becoming uncommon among the native childbearing population.

Although in European women chronic rheumatic heart disease is an uncommon pathology, there is an increasing number of Extra-european women among the

local childbearing population: in particular, the deliveries of immigrant mothers in the last ten years raised in our hospital from 4% to 22% without a significant increment in the total amount of deliveries. And also the case reported by this paper regards an immigrant woman coming from a high-risk region for chronic rheumatic heart disease.

There are, however, no published studies which compare cesarean section and spontaneous delivery in women affected by rheumatic heart disease, and European guidelines provide no useful information on such topic. We found a few recent reports about rheumatic heart disease during pregnancy, and some isolated cases described in the literature.

The more characteristic lesion of rheumatic heart disease is mitral stenosis, followed by the combination of mitral stenosis with aortic regurgitation. The mitral valve may become both stenotic and incompetent, and the valve may calcify.

Pregnancy drastically stresses the circulation in women with severe mitral stenosis. The increased blood volume, heart rate, and cardiac output raise left atrial pressure to a level that causes severe pulmonary congestion, leading to progressive exertional dyspnea, orthopnea, paroxysmal nocturnal dyspnea, and pulmonary edema. Women who have not been receiving antenatal care often present initially with severe pulmonary edema during pregnancy. In long standing cases, severe right heart failure develops. Infective endocarditis, pulmonary embolism, and massive hemoptysis may also occur.

Pregnant women with heart disease are at higher risk for cardiovascular complications during pregnancy and also have an higher incidence of neonatal complications (Siu et al., 2002). In particular, maternal mortality rate related to mitral stenosis with atrial fibrillation is about 14-17% (Ueland, 1978), being maternal death risk highest in the third trimester and in the puerperium (Szekely et al., 1973).

The significant hemodynamic changes that accompany pregnancy, however, make the diagnosis of certain forms of cardiovascular disease difficult. During normal pregnancies, women frequently experience dyspnea, orthopnea, easy fatigability, dizzy spells, and occasionally, even syncope. On physical examination, dependent edema, rales in the lower lung fields, visible neck veins, and cardiomegaly are commonly found. Systolic murmurs occur in more than 95% of pregnant women, and internal mammary flow murmurs and venous hums are common.

On the other hand, some findings indicate heart disease in pregnancy and should advise confirmatory diagnostic tests of possible significant cardiovascular abnormality. The symptoms include severe dyspnea, syncope with exertion, hemoptysis, paroxysmal nocturnal dyspnea, and chest pain related to exertion. Physical signs of organic heart disease include cyanosis, clubbing, diastolic murmurs, sustained cardiac arrhythmias, and

loud harsh systolic murmurs.

It is of paramount importance preconception counselling in women with heart disease, because of the high morbidity and mortality of those pathologies during pregnancy. And in case of cardiac condition that can be completely or partially resolved, the patient should be strongly advised to undergo the necessary treatment before pregnancy and allow some months to elapse before becoming pregnant. And in particular mitral stenosis and regurgitation could be ameliorated by therapy.

In our case during post-delivery hospitalization the leukocytosis allowed us to hypothesize the beginning infection, but we were at the same time frightened of an endocarditic complication. We strictly monitored the patient for shock, severe heart failure and embolic events.

Our patient had two previous deliveries and both pregnancies were unaffected by complications, on this basis we can hypothesize that during previous pregnancies she had milder pathology unable to compromise cardiocirculatory system during pregnancy and only at third pregnancy during the third trimester we observed the complications of her mitral stenosis.

Close attention should be paid to any pregnant woman with unexplained fever, cardiac murmur and new or pre-existing heart disease. And management should be initiated as soon as possible to control maternal clinical conditions and prevent worse complications.

General practitioners and obstetricians should be aware about heart diseases to be found in the preconceptional period, in order to ameliorate the prognosis of these patients, and in particular chronic rheumatic heart disease should be always considered in patients from coming high-risk regions.

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