Prenat Cardio 2021; 11(1): 70–73
DOI: https://doi.org/10.5114/pcard.2021.108545
Received: 15.03.2021
Accepted: 13.07.2021

Prenatal Cardiology
www.termedia.pl/Journal/Prenatal_Cardiology

Case report

Fetal autoimmune myocarditis and complete heart block – a case report

Julia Murlewska[1], Magdalena Szlagor[2], Maria Respondek-Liberska[1,3]

[1]Department of Prenatal Cardiology, Polish Mother's Memorial Hospital, Research Institute, Lodz, Poland
[2]Students' Prenatal Cardiology Team, Medical University of Lodz, Poland
[3]Department for Fetal Malformation Diagnoses and Prevention, Medical University of Lodz, Poland

Abstract

The patient was referred for fetal echocardiographic examination at 25.3 weeks of pregnancy due to bradycardia. Third-degree atrioventricular block was diagnosed, and hydroxychloroquine, steroid, and antibiotic therapy, as well as plasmapheresis, were started due to confirmation of the presence of antinuclear Ro-52 and Ro-60 maternal antibodies. The occurrence of disseminated focal areas of myocarditis was confirmed.

Key words: fetal fibroelastosis, fetal heart block, fetal myocarditis, prenatal therapy.

Corresponding author:
Magdalena Szlagor
Students' Prenatal Cardiology Team
Medical University of Lodz, Poland
e-mail: szlagor.magdalena@gmail.com

Introduction

Myocarditis in a fetus is a rare disease, and there are no large studies that establish its clear pathomechanisms. It is a disease with an extremely wide range of clinical presentations, from asymptomatic to life-threatening, importantly including sudden death [1-4].

Case study

The patient was referred for a fetal echocardiographic examination at 25.3 weeks of pregnancy due to bradycardia of 55 bpm. Third-degree atrioventricular block was diagnosed, and treatment with hydroxychloroquine and steroids (at a daily dose of 200 mg, and dexamethasone – initially at a dose of 4 mg/day orally then intravenously 3 × 4 mg, and betamethasone – 2 × 12 mg intramuscularly), as an anti-inflammatory and immunosuppressive therapy due to confirmation of the presence of antinuclear Ro-52 and Ro-60 maternal antibodies, was initiated (Table 1). So far, the patient has not been chronically ill; it is her first pregnancy. Echocardiography revealed the following: cardiomegaly – cardiothoracic ratio 0.5 and cardio-vascular profile score 8. Mild mitral and mild-to-moderate tricuspid regurgitation [5] were also seen (Figure 1), with correct ventricular contractility, correct width, and right-to-left foramen ovale valve opening, in which the peak systolic foramen ovale flow was 40 cm/s. The occurrence of disseminated focal areas of myocarditis was confirmed: scattered echogenic spots within the left ventricle, right ventricle, interventricular septum, and non-contiguous echogenic patches throughout the tricuspid valve, mitral valve, left atrium, right atrium and hyperechogenic bowels, and oligohydramnios, with an amniotic fluid index of 6.4 cm (Table 1). The next examination took place after about two weeks (at 26.6 weeks). The patient was diagnosed with a decrease in cardiomegaly – the cardiothoracic ratio was 4, a discrete increase in pericardial fluid (3.5 mm) was observed, as well as the appearance of trace pulmonary regurgitation, and the cardio-vascular profile score

Table 1. Echocardiographic presentation and prenatal therapy of fetal myocarditis with an autoimmune background

GA DOM (weeks)	25.3	26.6	28.2	28.5	29.4	33.4
GA US (weeks)	24.5	26.1	27.5	27.6	28.0	31.0
EFW (g)	693	730	936	936	1193	1723
FHR VR/AR (bpm)	55/150	55/140	56/138	43/150	56/139	57/134
AFI (mm)	64	13	12	12	11	7
Fetal presentation	Breach II	Vertex II				
PE (mm)	3	3.5				4
Placenta (mm)	38.3	33	35	35	37	37
Nuchal cord	Yes					
Hyperechogenic bowels	Yes					
Hyperechogenic stomach	Yes					
UMB PI	1.45		1.56			3.16
HA/CA (CTAR)	0,5	0.4				0.3
PVR PS (cm/s)	50					59
PVL PS (cm/s)	55			50		70
CVPS	8 –1 for heart size –1 for TR	7 –1 for heart size –1 for PE –1 for TR				8 –1 for PE –1 for TR
FO direction	R-L	Bilateralis				R-L
AOV PS (cm/s)	120	125	120	120	130	130
PAV PS (cm/s)	100	100	106	100	120	120
TR	Protosystolic, mild-to-moderate					
MR	Protosystolic, mild					
PR	No	Mild		No		
Medications	HCQ					
	Dexamethasone and betamethasone					
		Cefuroximum Nifuratel Nystatin				
		Salbutamol, isoptin Plasmapheresis – 4 courses				

GA DOM – gestational age, date based on menstruation, GA US – gestational age, based on ultrasound, EFW – estimated fetal weight, FHR VR/AR – fetal heart rate ventricular rate/atrial rate, AFI – amniotic fluid index, PE – pericardial effusion, UMB PI – umbilical artery pulsatility index, HA/CA – heart area/chest area, PVR PS – pulmonary veins right peak systolic velocity, PVL PS – pulmonary artery left peak systolic velocity, CVPS – cardiovascular profile score, FO – foramen ovale, AOV PS – aortic valve peak systolic velocity, PAV PS – pulmonary valve peak systolic velocity, TR – tricuspid regurgitation, MR – mitral regurgitation, PR – pulmonary regurgitation

was 7. An improvement in the amount of amniotic fluid was also obtained; the amniotic fluid index was 13 cm. Additional antibiotic therapy was proposed: biofuroxime, intravenous injections with dexamethasone, intramuscular betamethasone, and therapeutic plasmapheresis (in total, four plasmapheresis cycles were performed). During prenatal steroid therapy, significantly higher flow velocities through the fetal pulmonary veins were observed, with a peak systolic flow of 70 cm/s and a two-way nature of the flow through the foramen ovale opening. The patient received continuous rheumatological-cardiological-obstetric surveillance during pregnancy, and it was possible to start treatment with salbutamol due to the absence of cardiac contraindications (Table 1). In 28.5 weeks, improvement in the pulmonary valve regurgitation was achieved after antibiotic therapy. The growth of the fetus was observed within the normal range up to 33.4 weeks, and then we observed a slowdown in the growth of the fetus. The amount of amniotic fluid index also gradually decreased to 7 cm and the cardiothoracic ratio progressively improved/decreased to 0.3, although we observed an increase in pericardial effusion of 4 mm, and abnormalities in peripheral/extracardiac flows appeared: a significant increase in umbilical pulsatility index (to 3.16) and ductus venous reversal flow, but still with a cardio-vascular profile score of 8 with no cardiac failure obtained. During echocardiographic monitoring, the fetus had a constant tendency for nuchal cord while maintaining the same fetal position (Table 1). Myocarditis evolved to form the following: non-contiguous echogenic patches in the mitral valve,

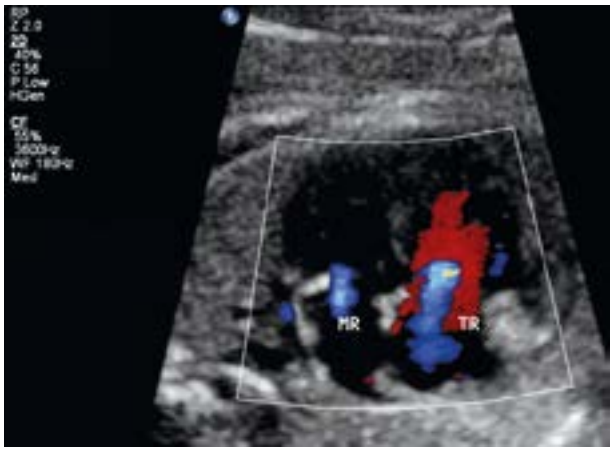

Figure 1. Mild mitral and mild-to-moderate tricuspid regurgitation, based on the length of the jet into the RA's classification: mild (length of jet < 1/3 of the distance to the opposite atrial wall); mild-to-moderate (length of jet between 1/3 and 2/3 of the distance to the opposite atrial wall) [5]

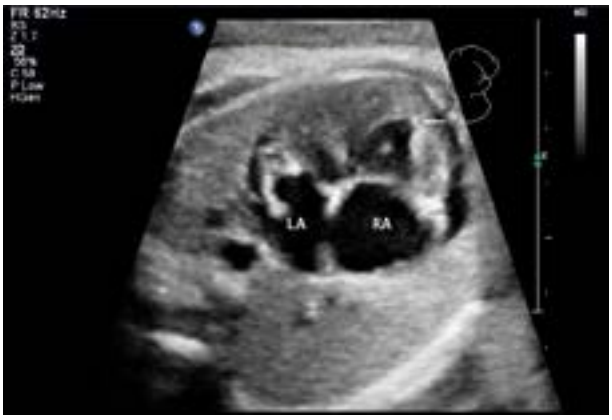

Figure 2. Presentation of myocarditis as non-contiguous echogenic patches mainly in the mitral valve and tricuspid valve with right ventricular hypertrophy and pericardial effusion at 33.4 weeks of pregnancy

Table 2. Evaluation of autoimmune myocarditis severity based on the division proposed by McElhinney et al. [6]

Echocardiographic grading of myocarditis/EFE	Severity of myocarditis/EFE	
	25.3 weeks	29.3 weeks
1 – Mild	Scattered echogenic spots within the LV, RV, IVS	Scattered echogenic spots within the RV, TV, MV
2 – Moderate	Non-contiguous echogenic patches throughout the TV, MV, LA, RA	Non-contiguous echogenic patches in the MV, TV, LA, RA
3 – Severe	Not seen	Not seen

EFE – endocardial fibroelastosis, LV – left ventricle, RV – right ventricle, IVS – intraventricular septum, TV – tricuspid valve , MV – mitral valve, LA – left atrium, RA – right atrium

tricuspid valve, left atrium, and right atrium, as well as atrial and right ventricular enlargement and right ventricle hypertrophy (Figure 2, Table 2). The planned subsequent echocardiography at an interval of two weeks was not performed due to life-threatening symptoms in the fetus, and therefore the patient gave birth prematurely by Caesarean section at 36 weeks of pregnancy. A daughter was born with weight 2120 g, Ap 10/10, pH 7.31, base excess 1.3, normal oxygen saturation (98%), and elevated cardiac markers: B-type natriuretic peptide 950 pg/ml and troponin 453 ng/ml. Using a central venous access device, dopamine, and fraxiparin, steroid therapy was initiated shortly after birth with a weight-adjusted dose, due to the confirmation of third-grade atrioventricular block with heart rate 60 bpm, positive for Ro-52 antibodies with a titre of 1 : 320. The feeding was completely parenteral. On the first day of life, two pericardial electrodes were implanted for external stimulation, obtaining effective stimulation with a heart rate of 130 bpm. On the sixth day of life, right pleural emphysema was observed, drainage of the pleural cavity was carried out, and in the following days there were difficulties in healing the wound after the drainage and growing CRP, with positive stool culture of *Salmonella*, *Shigella*, *E. coli*, *Yersinia*, and *Campylobacter*. Antibiotic therapy with amoxicillin and vancomycin was used. On day 17, normocytic anaemia led to the implementation of erythropoietin treatment and transfusion of a 0 Rh D blood product. On day 29, due to increasing stimulation disorders, after initially switching to a reserve electrode, it was finally decided that pacemaker implantation was urgently required. The hospitalization lasted a total of 7 weeks (including the prenatal period), and the child, in a generally good condition, was transferred to a cardiology clinic for further care.

Discussion

In the presented case, the heterogeneous echogenicity and variability of this image made us suspect myocarditis. Murlewska et al. [7] collected very important data about management in maternal autoantibody-mediated clinical fetal myocardial disease. Due to the increased risk of pregnancy complications among women with connective tissue disease, early assessment and detailed counselling should be implemented. If there is suspicion of any fetal myocardial disease, the authors point to the importance of the role of echocardiography. Furthermore, this examination is also an indicator of the degree of cardiac failure that should be taken into consideration during obstetric management. Fetal echo examination is indicated weekly from 16 weeks of gestation upwards in cases of involvement of positive maternal anti-Ro/SSA or anti-La/SSB antibodies. The authors quote one study that suggests that hydroxychloroquine could be used in the prevention of neonatal cardiac involvement in pregnant women who have had a fetal myocardial disease in a previous pregnancy [7].

Conflict of interest

The authors declare no conflict of interest.

REFERENCES

1. von Kaisenberg CS, Jonat W. Fetal parvovirus B19 infection. Ultrasound Obstet Gynecol 2001; 18: 280-288.

2. Fesslova V, Mongiovì M, Pipitone S, Brankovic J, Villa L. Features and outcomes in utero and after birth of fetuses with myocardial disease. Int J Pediatr 2010; 2010: 628451.
3. Watson WJ, Awadallah S, Jaqua MJ. Intrauterine infection with coxsackievirus: is it a cause of congenital cardiac malformations? Infect Dis Obstet Gynecol 1995; 3: 79-81.
4. Hichijo A, Morine A. A case of fetal parvovirus B19 myocarditis that caused terminal heart failure. Case Rep Obstet Gynecol 2014; 2014: 463571.
5. Messing B, Porat S, Imbar T, Valsky DV, Anteby EY, Yagel S. Mild tricuspid regurgitation: a benign fetal finding at various stages of pregnancy. Ultrasound Obstet Gynecol 2005; 26: 606-610.
6. McElhinney DB, Vogel M, Benson CB, Marshall AC, Wilkins-Haug LE, Silva V, et al. Assessment of left ventricular endocardial fibroelastosis in fetuses with aortic stenosis and evolving hypoplastic left heart syndrome. Am J Cardiol 2010; 106: 1792-1797.
7. Murlewska J, Preis K, Słodki M, Strzelecka I, Smolewska E, Respondek-Liberska M. Management in maternal autoantibody-mediated clinical foetal myocardial disease. Prenat Cardio 2019; 9: 5-11.

Division of work:
Julia Murlewska (ORCID: 0000-0002-8266-4585): research concept and design, collection and assembly of data, data analysis and interpretation, writing the article
Magdalena Szlagor (ORCID: 0000-0001-5432-3612): collection and assembly of data, writing the article
Maria Respondek-Liberska (ORCID: 0000-0003-0238-2172): critical revision of the article, final approval of the article