CASE REPORT

Autoantibody-associated congenital heart block: a rare cause of persistent fetal bradycardia

Aruna Nigam, Ayesha Ahmad

Department of Obstetrics and Gynaecology, Hamdard Institute of Medical Sciences and Research, New Delhi, India

Correspondence to

Dr Aruna Nigam,
prakasharuna@hotmail.com

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SUMMARY

Persistent fetal bradycardia in early pregnancy is a rare finding and indicates towards congenital heart block. This is commonly associated with positive maternal anti-Ro/anti-La antibodies. A case of an asymptomatic primigravida with persistent fetal bradycardia on routine antenatal ultrasound is reported with special emphasis on its management options.

BACKGROUND

Isolated congenital heart block (CHB) is defined as an irreversible atrioventricular (AV) block, unaccompanied by fetal cardiac, structural or anatomic abnormalities. The diagnosis can be made in the prenatal, intranatal or postnatal period by the presence of severe fetal bradycardia. It is associated with anti-Ro/anti-La antibodies in the mother. The reported incidence of isolated CHB is 1 in 17 000 live births.¹ The disease usually starts manifesting at between 16 and 24 weeks of gestation because of increased transfer of maternal IgG antibodies into the fetal circulation. The target antigens are sequestered intracellularly and are found in fetal cardiac tissue including that of the conduction system. These autoantibodies cause inflammation, haemorrhage and necrosis in myocardial cells specific to the fetal conduction system leading to conduction defects.

CASE PRESENTATION

A 27-year-old primigravida presented for her first routine antenatal visit at 17 weeks. The conception was spontaneous, within 10 months of cohabitation. There was no history of joint pains, low-grade fever or rashes. History of consanguinity and family history of hypertension and hyperthyroidism were positive. General and systemic examination was unremarkable. Obstetric examination revealed a uterus sized 16–18 week, corresponding to the period of gestation.

INVESTIGATIONS

The patient's level 2 ultrasonography reported persistent fetal bradycardia, with a fetal heart rate of 48–52 bpm. There was no structural cardiac defect and no evidence of hydrops. Fetal echocardiography confirmed the 2:1 AV block (figure 1 and video 1).

The patient was investigated for evidence of autoimmune disease. Her haemoglobin was 8.9 g/dL, total leukocyte count 14 900, erythrocyte sedimentation rate 110, anti-La/SSB antibodies, anti-Ro/SSA antibodies and antinuclear antibodies were positive.

Antiphospholipid antibody, antidouble-stranded DNA and rheumatoid factor were negative.

DIAGNOSIS

Autoantibody-associated CHB in the fetus.

TREATMENT

The patient was put on oral dexamethasone therapy.

OUTCOME AND FOLLOW-UP

The patient developed complete heart block in fetus within 2 weeks of starting the therapy. In view of long-term morbidity and mortality, the patient opted for termination of pregnancy as she was only 19 weeks pregnant.

DISCUSSION

Isolated fetal CHB in the absence of structural cardiac anomalies is mainly associated with positive maternal anti-Ro/anti-La antibodies. The mother is asymptomatic in 50% of cases. The risk of recurrence of CHB in subsequent pregnancy is around 19%.²

Clinical manifestations of CHB in the fetus depends on the ventricular rate and in cases of very low ventricular rate, fetal hydrops and fetal death may occur. The mortality rate of neonates born with CHB is 19–31% in the presence of structurally normal hearts, and in the presence of hydrops fetalis, it is as high as 80%.³ In addition to low ventricular rate, premature birth, low birth weight, significant structural heart disease, ventricular dysfunction and associated cardiomyopathy, hydrops fetalis also suggests a poor prognosis.

There is no known effective therapy. Prenatal interventions consist of drugs given to mothers in order to diminish the maternal autoimmune response and/or the fetal cardiac inflammatory

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Figure 1 Fetal echo showing fetal bradycardia in wave form.

Video 1 Fetal echo showing fetal bradycardia.

injury, and, in some cases, to increase the fetal heart rate. Corticosteroids have been used, especially dexamethasone and β -methasone, since they are not metabolised by the placenta and are thus available to the fetus in an active form. However, treatment is probably unable to revert third-degree heart block once it is established, as in our case. Long-term side effects of therapy to mother and baby are not known. In one case series, plasmapheresis, intravenous immunoglobulin and glucocorticoids together were used in positive anti-Ro/SSA antibody patients, with successful outcome in two of the three babies.⁴ Fetal echocardiography is recommended every 2 weeks starting from the 16th week of gestation to follow the progression of the disease.

Learning points

- ▶ Fetal bradycardia in early pregnancy must be investigated for autoimmune disease even in asymptomatic cases.
- ▶ Congenital heart block in presence of fetal hydrops is a bad prognostic sign.
- ▶ No definitive therapy is known but dexamethasone, plasmapheresis and intravenous immunoglobulin have been tried. Immediate pacemaker implantation after delivery improves the prognosis.

Competing interests None declared.

Patient consent Obtained.

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