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Case Report

Complete heart block in fetus – A rare combination of maternal autoimmune disorder and fetal structural cardiac disease – A case report and review of literature

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ABSTRACT

Background: Majority of rhythm disturbances detected by fetal echocardiography are self-limiting. Appropriate work up of dysrhythmias is essential to prevent further damage of fetal conducting system and to initiate fetal therapy thereby reducing fetal losses.

Case Report: 25 year old Primi, 31 weeks of gestation, no comorbidities, was referred to our Antenatal OPD as severe fetal bradycardia. Antenatal Fetal Echo revealed third degree heart block with no evidence of hydrops. Maternal serum workup revealed autoimmune etiology. Fetal therapy started and Pregnancy followed up weekly till term gestation to deliver an alive baby by vaginal delivery. Baby was diagnosed to have Large ASD with a large PDA having bidirectional shunt by paediatric cardiologist. Surgical management was done by PDA ligation and permanent pacemaker. Unfortunately, baby succumbed due to respiratory failure and sepsis, as it was high risk complete heart block with multifactorial causation.

Conclusion: Fetal bradyarrhythmia need to be evaluated carefully both for maternal autoimmune disorders and intrinsic cardiac causes of fetus. This case is reported for its rarity of having both etiologies presenting together as third degree or complete heart block.

1. Introduction

Fetal cardiac dysrhythmias are mostly isolated transient ectopic beats with less than 2% of cases contributing to clinically significant arrhythmias. If not identified earlier, it may lead to congestive cardiac failure, non-immune hydrops and fetal/neonatal losses.

2. Case Report

25 year old Gravida 1, with no antecedent risk factors was evaluated for fetal bradyarrhythmia. Fetal Echo - Bradycardia – rate 48bpm, complete heart block with AV dissociation (Atrial rate - 133, Ventricular rate – 49), no hydrops. Cardiac chambers were of appropriate dimensions. There was no family history of young age sudden deaths, connective tissue disorders or heart diseases. Mother had euthyroid status, no history of recent illness / drugs usage. Clinical examination of mother revealed no significant abnormalities.

Autoimmune workup - Serum Anti Ro antibody (SSA) – positive (4.62); ANA – positive. Anti La antibody (SSB) - negative, serum compliment levels C3, C4 - normal, anti DS DNA - negative.

She was started on glucocorticoids (Dexamethosone 4 mg /day oral) for 2 weeks, DMARD (hydroxychloroquine 200 mg/ day oral) and terbutaline 2.5 mg bid till delivery. Fetal echo was done once in 2 weeks. She was allowed for spontaneous normal vaginal delivery at 39 weeks gestation

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as there was no indication for early termination or elective caesarean. Careful electronic fetal heart rate monitoring was done. Baby did not have any signs of neonatal lupus. Neonatal echo identified severe bradycardia (40beats/min) along with large atrial septal defect and patent ductus arteriosus with bidirectional shunt. Left ventricular function was normal. Baby was immediately managed post-delivery in neonatal intensive care unit. Permanent pacemaker implantation and patent ductus arteriosus ligation was done. Baby died postoperatively due to respiratory failure and sepsis.

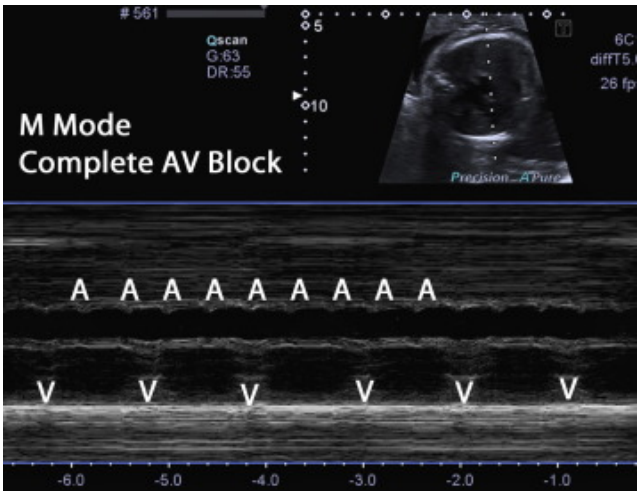

Fig. 1: M mode complete AV block

3. Discussion

Fetal cardiac abnormalities may be of two types: tachyarrythmias and bradyarrythmias, the former twice more common than the latter. The most common cause of tachyarrythmias is extrasystole which may be atrial or ventricular in origin. Types of tachyarrythmias are sinus tachycardia, atrioventricular re-entry supraventricular tachycardia, atrial flutter, rare causes are atrial fibrillation and junctional tachycardia. Treatment for controlling ventricular rate are transplacental digoxin and beta blockers like flecanaide and sotalol. Most common cause for bradyarrhythmia is complete atrioventricular heart block or third degree heart block, other causes being first and second degree heart blocks, QT prolongation syndrome and sinus bradycardia.[1] Tools used to detect fetal rhythm disturbances include auscultation by stethoscope, fetal doppler, fetal ECG and ST analysis, fetal magnetocardiography and fetal echocardiography, the last one being gold standard for diagnosis. Fetal echo is widely available, safe throughout pregnancy and also has the ability to detect structural cardiac abnormalities, syndromic association of fetus, evidence of hydrops and overall fetal wellbeing. M mode and spectral doppler are most commonly used modalities in fetal echo. Tissue doppler analysis is used to construct fetal kinetocardiogram which aids in accurate diagnosis of rhythm abnormalities.

Multiple factors to be considered as causative factors like maternal connective tissue disorders, structural cardiac lesions, hyperthyroidism, infection, recent illness, recreational or medical drug usage, excessive caffeine intake, family history of QT prolongation, congenital deafness, sudden cardiac deaths and tuberous sclerosis.[2]

While evaluating bradyarrhythmias, extrinsic factors like cord compression, drugs and maternal hypothermia should be ruled out. Milder degrees of heart block like Wenckebach type 1 and Mobitz type 2 may progress to complete heart block, a phenomenon where there is complete failure of conduction of atrial impulse to ventricles. Majority of third degree or complete heart block are due to structural cardiac disease or maternal autoimmune disorders. It becomes detectable at around 18 weeks of gestation. Systemic lupus erythematosus, Sjogren's syndrome, rheumatoid arthritis, scleroderma and undifferentiated connective tissue disorders are common autoimmune causes. There may be only serological evidence of autoimmunity with no clinical manifestations. Recurrence risk of complete heart block is 10 to 16%. Prevalence of complete heart block in liveborn babies is 1 in 20000.

Congenital AV block is immune mediated in 50% of the cases. AV block manifests in transplacental transfer of Anti Ro and Anti La antibodies. The fetal risk is maximal between 16-26 weeks of gestation. Ventricular rate <50 bpm, underlying severe congenital heart disease, presence of hydrops / heart failure are important mortality risk factors according to American Heart Association. Clinically detectable autoimmune disease is not present in 70-80% of mothers of affected fetuses.

The conduction system is damaged by maternal antibodies which cross the placenta and react with antigens expressed on the surface of cells in the fetal cardiac conduction system, resulting in immunoglobulin deposition on the cells and fibrosis of the affected cells. The inflammatory reaction causes permanent damage of the fetal cardiac conduction system and developing myocardium due to localized cellular apoptosis. Impaired physiologic clearance of the apoptotic cells results in their accumulation which in turn leads to inflammation and cellular injury.[3]

The second group of fetuses with congenital atrio-ventricular block (CAVB) have structural cardiac disorders. The etiology of nonimmune complete AV Block are complete absence of the AV node, lack of union between the AV node and the distal conduction system, or the presence of a poorly functioning system. The most common structural cardiac abnormalities associated with CAVB are AV septal defects, with or without left atrial isomerism and atrioventricular discordance. TGA, ASD, VSD, TOF, PDA are all associated with conduction abnormalities. In

our case, both autoimmunity and structural defect (PDA and ASD) were associated.

Other rare causes are mutations in cardiac transcription factors, Holt-Oram syndrome and mutations in sodium ion channel causing Brugada syndrome.

Several studies document a very high mortality rate among fetuses and neonates with complete heart block and structural cardiac lesions.[4] The mortality rates range from 7 to 33% in the setting of a structurally normal heart and as high as 86% for those with structural cardiac lesions. Poor outcomes are especially associated with structural cardiac disease, left atrial isomerism, hydrops, ventricular rates below 55 beats/min, atrial rates below 120 beats/min, AV valve regurgitation, dilated cardiomyopathy, endocardial fibroelastosis, prolonged corrected QT interval and delivery at less than 33 weeks' gestation.

Most studies and systematic reviews suggest irreversibility of complete heart block with steroid therapy, though there may be some benefit in reduction of hydrops and onset of later cardiomyopathy. PRIDE study also suggests irreversibility with steroid treatment.[4] The recommended dose of oral dexamethasone is 4-8 mg day for 2 weeks after discussing risks and benefits with the patient. Side effects of steroid therapy include growth restriction, oligohydramnios, ductal constriction and maternal diabetes mellitus.

Beta sympathomimetic drug oral terbutaline 2.5 to 7.5 mg oral QID is suggested if ventricular rate is < 56 bpm. There was significant improvement in mean fetal ventricular rate and live birth rates.[5]

IV immunoglobulin administration to mother and plasmapheresis have been tried in antibody meditated heart block, in small cohort studies.[6]

The prognosis of children with CHB depends on the underlying cause. In a study of 59 cases, rates of live birth and 1-year survival were 88% and 75% in patients with immune-mediated CHB (n = 35). However, those with underlying structural heart defect (n=24) had poor outcomes, with survival rates as low as 56% at birth and 19% at the age of one year. Long term neurodevelopmental delay is also reported in babies who recovered from complete heart block.[7]

4. Conclusion

Early detection of heart block helps us to identify causative factors and to start monitoring the pregnancy. Prognosis is bad if multiple etiologies like autoimmunity and structural cardiac disease are present, as in our case. The present case was referred late at 30 weeks of gestation. Role of fetal therapy is not promising in advanced cases of heart blocks. Recurrence risks should be educated to patients to facilitate optimization of maternal autoimmune conditions before attempting future pregnancies to avoid fetal losses.

5. Source of Funding

None.

6. Conflict of Interest

None.

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