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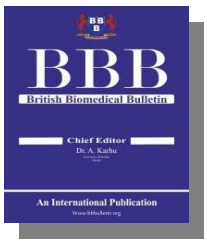

Original

In-utero Presentation of Complete Atrio-ventricular Block - Not Always Neonatal Lupus!

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A R T I C L E I N F O

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A B S T R A C T

Complete atrio-ventricular block (CAVB) presenting in-utero, as widely known, is secondary to neonatal lupus unless proved otherwise. We hereby report a neonate with CAVB associated with heart defects presenting in-utero, without any evidence of neonatal lupus. Hence, we emphasize the association of heart defects in CAVB as the leading factor in diagnosing the etiology.

Introduction

Hydrops fetalis, an end stage process for number of fetal diseases, results in tissue edema and effusion of multiple body cavities.[1] we are reporting a neonate with in utero presentation of complete atrio-ventricular block, presenting with non-immune fetal hydrops.

Case Report

A preterm male baby of gestational age 32 weeks was delivered by labor naturalis to a primigravida mother. History taking reveals a significant antenatal history with fetal bradycardia (heart rate -62/min) in the antenatal ultra sonogram done at 28 weeks of pregnancy (all antenatal visits and ultra sonograms were at a different hospital).

On thorough clinical examination, the baby was noted to have weak cry, bradycardia (HR-60/min) which did not vary with phases of respiration, crying or painful stimulus. Baby had severe respiratory distress, pitting edema over the chest (Figure 1a), abdomen, limbs and diffuse scalp edema (Figure 1b).

Considering the fact that 95% of the fetuses who present with complete atrio-ventricular block (CAVB) in-utero are secondary to neonatal lupus erythematosus (NLE)[1-3], a clinical diagnosis of NLE was made. Mother was examined and no signs of connective tissue disorders were noted clinically. Her ANA, dsDNA, anti-Ro/SSA antibodies were negative. Baby showed no rash, normal liver function tests and normal blood hemogram which made the diagnosis of NLE unlikely. There was no blood group incompatibility between mother and baby with any evidence of hemolysis. Hence, clinical diagnosis was revised to non immune fetal hydrops secondary to complete atrio-ventricular block (CAVB).

Chest roentgenogram of the baby revealed cardiomegaly. Cardio-thoracic ratio was greater than 60 % (Figure 2). Electro-cardiogram (ECG) revealed bradycardia with regular rhythm but absent P waves, suggestive of congenital heart block with an atrial flutter(Figure 3).

Echocardiogram (ECHO) revealed patent ductus arteriosus with gradient 11mm, small atrial septal defect left to right shunt, with good biventricular function. The ECHO findings were not suggestive of NLE as NLE is usually associated with structurally normally heart[4]. A final diagnosis of complex CAVB associated with structural heart defects which presented as non-immune fetal hydrops was made.

The baby was on continuous ECG monitoring. Baby was then treated symptomatically with antiarrythmic agents. As symptomatic complete CAVB with heart defects is an indication for pacing, the same was planned but mean while the baby had succumbed.

Discussion

CAVB has an incidence of 1 in 22,000 live births[5]. In a series of patients with complex CAVB with structural heart defects, only 14% survived the neonatal period, compared with 85% survival in patients with isolated CAVB.[6]

This emphasizes that when CAVB is diagnosed, it is essential to categorize into isolated CAVB or complex CAVB with structural heart defects. It is evident that prognosis is better for the asymptomatic isolated CAVB patients. Hence, the association of CAVB with structural heart defects makes the diagnosis of neonatal lupus unlikely.

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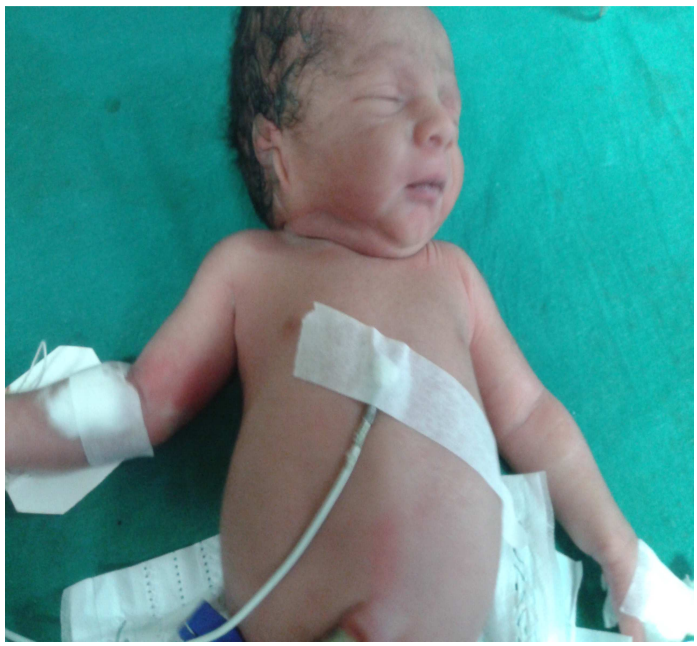

Figure 1.a. Neonate with non-immune fetal hydrops

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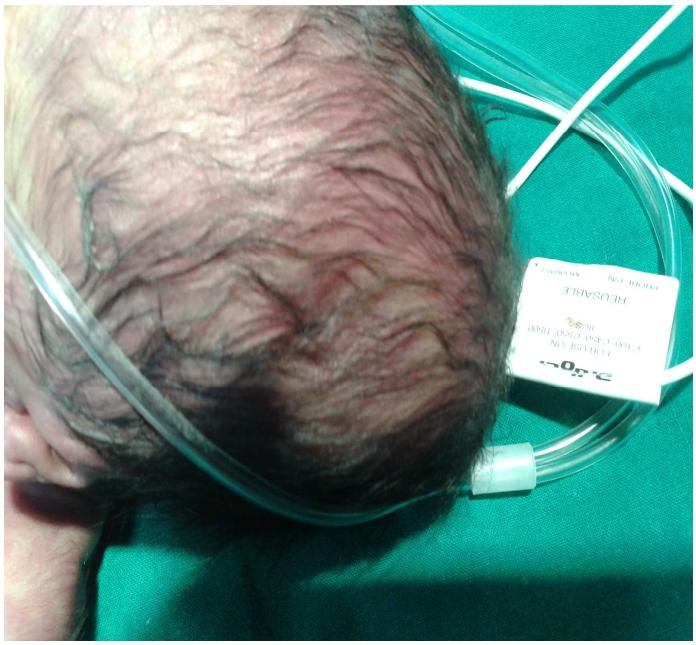

Figure 1.b. Picture showing diffuse scalp edema

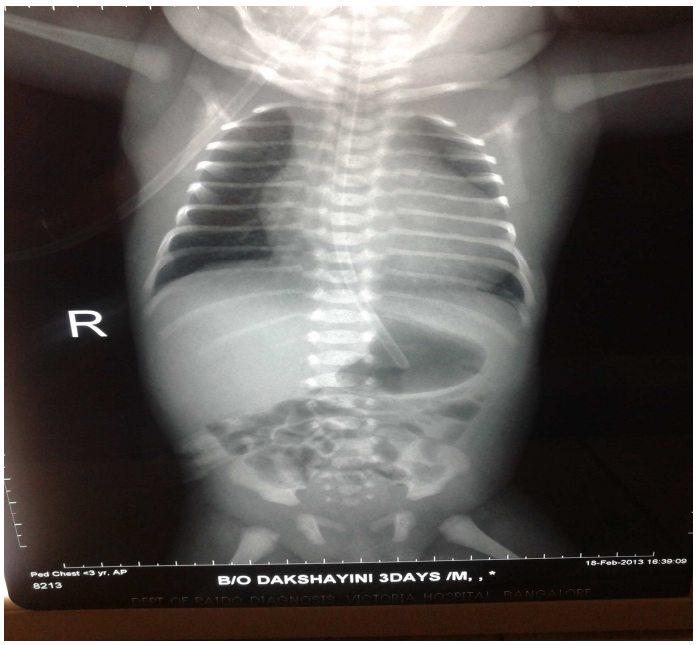

Figure 2. Chest roentenogram showing cardiomegaly

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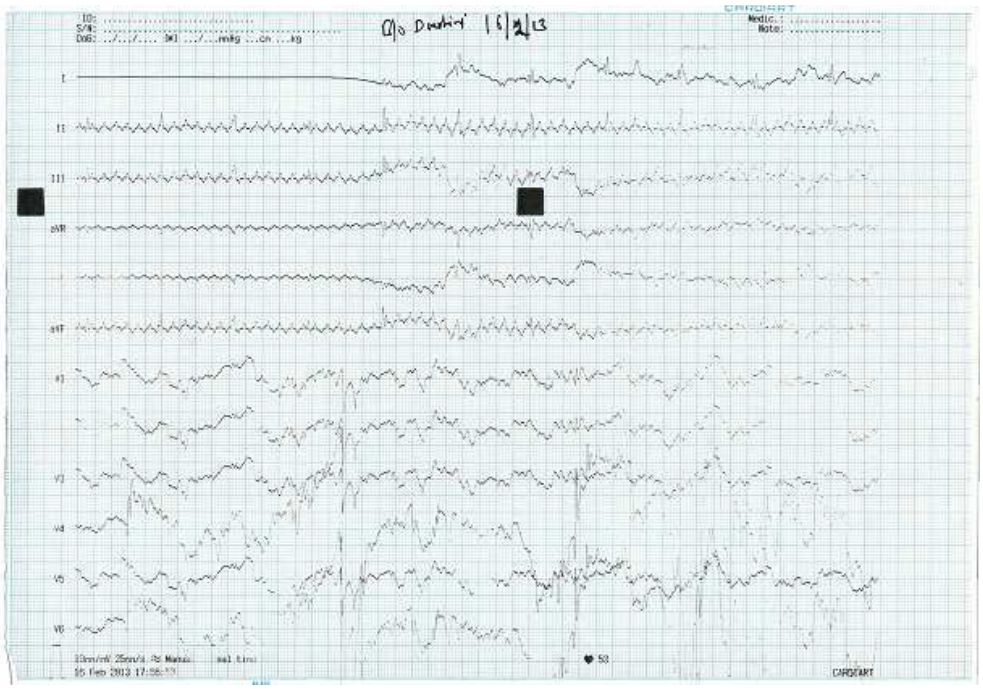

Figure 3. Echocardiogram showing complete atrio-ventricular block with atrial flutter

Abbreviations

CAVB - Congenital atrio-ventricular block
NLE - Neonatal lupus erythematosus
dsDNA - Double stranded DNA
ANA - Antinuclear antibodies
ECG - Electrocardiogram
ECHO - Echocardiogram