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SEVER NEONATAL THROMBOCYTOPENIA IN CASE WITH PATAU SYNDROME (TRISOMY 13)

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ABSTRACT

Newborns with Patau syndrome are prone to autoimmune thrombocytopenia more than normal population. There isn't Statistics regarding the prevalence of autoimmune thrombocytopenia in this syndrome. We present a newborn with Patau syndrome involved in sever autoimmune thrombocytopenia which is confirmed during hospitalization due to respiratory distress and growth restriction. The patient didn't has any intracranial hemorrhage. We discuss clinical diagnostic criteria in our presentation.

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Introduction

Patau syndrome is a rare congenital abnormality in newborns. The prevalence of it is about 1 case in 10000 alive births. Patau syndrome caused in add a copy of chromosome 13. Advanced maternal age often is associated with this abnormality. The additional copy in neonates induces mental and physical abnormalities and cardiac defects. The Syndrome has higher incidence in females Vs males and involves all races. Patau syndrome as well as Down syndrome has a positive correlation with mother's age (1).

Clinical Menifestations

Patau syndrome is including of the spectrum of different anomalies and multi-organ involvement like as microcephaly, deformed and lower than normal line ears, Cleft lip and

Palate, memingomielocele, microphetalemia, anophetalemia, hypotelorism as well as polidactilia, clitodactilia, heel anomaly and lack of rib. The other anomalies are genital anomalies like as criptorcydism and ambiogony. Almost the patients have cardiac abnormalities like as dextrocardia, ASD, VSD. Some patients have digestive and renal anomaly (2, 3).

Diagnosis

The syndrome is diagnosed by Prenatal sonography , Amniocyntesis and Chorionic villus biopsy. Determine of chromosome pattern by karyotype is Gold standard test for approving the diagnosis of Patau syndrome in newborn which is confirmed by three chromosome 13 (4, 5).

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Prognosis

In alive newbon with patau syndrome will de died nearly 45% in first month, 70% in first six months and finally the end of first year of birth (6).

Case presentation

We present the female newborn case with 37 weeks weighing 2100 gr and Head circumference 30 cm by normal vaginal delivery (NVD) from 34 old year mother an outcome of non-consanguineous marriage. Apgar in 1th and 5^{th} minute was ordinary 6/10 and 7/10. All Prenatal sonography during pregnancy were remarkable note. Atrial Blood Gas (ABG) of umbilical cord first hour ABG were in normal range. The newborn hospitalized due to Low Birth Weight, Symmetric IUGI, Respiratory distress and multiple anomalies. We used antibiotic Ampicillin and amikacin for the Patient. We performed laboratory examination such as CBC diff, BS, Sepsis screening and CXR. The Lab Exam are

HB= 19 PLT= $22*10^6$ $26*10^6$
Na=142 K=3.7
WBC=6700
poly45% Lymph50%
BS=61 Ca=9.1
Urea=14 Cr=0.8

Peripheral blood sample of her mother was examined for CBC diff and Platelet which was in normal range.

Clinically, we found microcephaly, hypotelorism, symmetric IUGI, VSD, ASD, mild PH, RVE and RAE in this patient. As well as we found Trisomy 13 in Karyotype evaluation and Patau syndrome was confirmed. In auditory screening, there was sensory and neural hearing loss.

Seven days after hospitalization, the patient was without respiratory distress and supplementary Oxygen discontinued. Meanwhile, we started single dose IVIG as 15mg/kg due to autoimmune thrombocytopenia suspected, then after some days we detected increasing of platelet step by step as it increased to 100000 after ten days.

The patient was transferred to the Neonatal ward and started feeding with her mother's milk with bottle milk for Cleft palate. Finally, the neonate started breast feeding as full amount and was discharged after three weeks by personal satisfaction not by physician order.

Discussion

Previous studies reported that the autoimmune thrombocytopenia in congenital syndrome like as Patau syndrome (Trisomy 13) (7). With regard to prevalent thrombocytopenia (platelet < $150000 * 10^9$) specially in NICU patients is about 35 – 45% and 20% NICU patients have thrombocytopenia lower than $50000 *10^9$, if its confirmed by platelet count, we must consider important causes of thrombocytopenia such as immune, infection and genetic abnormality.

Based on this evidence, we recommend to consider sever thrombocytopenia in chromosomal abnormality like as Patau syndrome and Start the therapeutic plan as well as continues and regular follow up for neonates who survive.

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