Sudden Stillbirth – A Rare Combination of Maternal Common Variable Immunodeficiency Disease, Placental Mass and Triploid Mosaicism.

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Introduction:-

Stillbirth has varied etiology ranging from unknown to either maternal, fetal and placental disorders or a combination of any. Large placental masses > 4cms are associated with obstetric and perinatal complications. Maternal immunodeficient conditions need efficient immunoglobin replacement to ensure adequate fetomaternal immunity in preventing infections. The combination of such multiple risk factors warrant vigilant monitoring to prevent adverse outcomes like stillbirth

Aim:-

Our aim is to attempt reconsideration of surveillance measures in presence of unusual risk factors.

Case:-

A retrospective case analysis of a 31-year-old obese multiparous lady with previous vaginal birth of twins and currently having long-term ongoing optimal immunoglobulin replacement therapy for her underlying common variable immune deficiency condition was done.

In the index pregnancy, her combined first-trimester aneuploidy screen and morphology scan were low risk for fetal abnormalities. She was diagnosed to have an abnormal placental cystic mass of 4-5 cms, likely chorangioma, at NT screening which progressively increased to 6 cms by 29 weeks with normal fetal biometry and no hyperdynamic circulatory disturbances or polyhydramnios as surveilled by the feto-maternal unit. At 33 weeks of gestation, she had sudden reduced fetal movements and was found to have fetal death inutero (FDIU) with no infective or obstetric symptoms leading to it. USS at 33 weeks showed 10 cms placental mass with FDIU. Her labour was induced and progressed to vaginal birth. She had intrapartum sepsis which could likely contribute to the fetal and maternal inflammatory response evidenced on autopsy.

Result:-

Fetal demise in an asymptomatic immunodeficient mother with optimal surveillance of placental mass.

Literature review:-

Placental Tumours:-

Placental tumours greater than 5 cm are associated with maternal and perinatal complications as the big tumors act like arteriovenous shunts leading to fetal congestive heart failure, hydrops, and even in utero death (1). Zanardini et al (2) case series reflected varied results of uneventful perinatal outcomes to IUGR in giant placental tumours.

Common variable immunodeficiency(CVID):-

CVID is the most prevalent symptomatic primary immunodeficiency characterized by impaired antibody responses, however, a case series by Kurevoca et al (3) suggests treatment with regular periodic adequate replacement therapy with intravenous immunoglobulins (IVIG) in pregnant women resulted in healthy newborns.

Old case report on Triploid fetus with Non-Trophoblastic Tumour in 1995(4) concluded that triploidy does not always imply the presence of a partial mole and the dictum, that pre-eclampsia, if it occurs under 20 weeks gestation, must be associated with a molar pregnancy, may not hold when placental aneuploidy is present. **There may be an association between choriohemangioma and polyploidy**.

Discussion & Conclusion:-

Prevention of stillbirth warrants vigilant fetomaternal surveillance based on the risk factors for it.
Available literature has revealed that placental masses are associated with fetal growth concerns or hyperdynamic complications.

Our case, with rare combination of large placental tumour and CVID, did not have any infections or fetal growth concerns or hyperdynamic complications like hydrops or polyhydramnios diagnosed antenatally. Inspite of optimal management and surveillance of risk factors in a multidisciplinary team approach, unfortunately, she had sudden FDIU.

Interestingly fetal autopsy revealed abnormal mosaic triploid cell line 69XXY of Umbilical cord along with hydropic placentomegaly having aneurysmal arterial dilatation on fetal surface. There was also evidence of maternal and fetal inflammatory response.

A similar old case reported in PUBMED in 1995 suggestive of a likely association between placental choriohemangioma with triploid fetus, but no CVID association. (4)

This case highlights that abnormal placental masses may have an association with abnormal fetal and/or placental karyotype. Large placental masses, especially more than 4cms in size, should have frequent fetal surveillance to detect acute hyperdynamic changes along with due consideration to non-invasive and invasive prenatal testing to rule out potential possibilities of fetal aneuploidy.

References:-

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