

Pregnancy outcome in Patient with Familial Partial Lipodystrophy & Insulin dependent Diabetes Mellitus

Background

Familial partial lipodystrophy is a rare genetic disorder characterised by selective, progressive loss of body fat from various areas of the body. Individuals have reduced subcutaneous fat in the arms and legs and head and trunk region. Excess subcutaneous fat accumulation in neck, face and intraabdominal region. The disproportional distribution of the fat usually result in insulin resistance and diabetes mellitus. Six different types have been identified The incidence is 1 in 10000000 in general population.

Case report

This is a case report of a 31 years old primigravida with history of Type 2 diabetes since 8 years and recently diagnosed with familial partial lipodystrophy since 2018 with marked insulin resistance and insulin dependent diabetes mellitus ,she had preconception counselling prior to embarking on her pregnancy. She was prescribed with high dose folic acid of 5mg preconceptionally. She required 870 units insulin on daily basis and her booking HBA1C level at gestation 7 weeks was 62mmol. Her booking BMI was 30.8 with normal blood pressure and no proteinuria. She had her recent retinal check which were normal. An early pregnancy scan at 8 weeks confirmed a viable intrauterine pregnancy. Otherwise, all her booking bloods were normal ,negative serology, rubella immune and rhesus positive.

All her antenatal visits were under the joint care of endocrinology and foetal maternal specialist. All her scans showed normal foetal growth with normal anatomy scan at 21 weeks. Over the course of her pregnancy, her insulin requirement increased exponentially which were adjusted in consultation with endocrinologist. She presented to the emergency room at gestation 29 + 5 weeks with symptoms of preeclampsia and proteinuria. She was reviewed and admitted for blood pressure monitoring and management as she was clinically stable with no clonus, normal reflexes. She was later on commenced on labetalol 200mg QDS to control her hypertension. During admission, she developed transient hypoglycaemic episodes and her insulin doses were withheld. Dexamethasone were administered in view of foetal lung maturity at 30 weeks gestation which stabilised her hypoglycaemic episodes . Her insulin doses were titrated according to her blood sugars . Over the next few days she developed significant proteinuria and worsening of preeclampsia in which magnesium sulphate was administered for maternal and foetal interest.

She underwent emergency caesarean section at gestation 32+6 weeks, delivering a baby girl weighing 2.8kg who was admitted to NICU for 4 weeks initially for hypoglycaemic episodes and later for blanket therapy for physiological jaundice. Mother and baby did very well postoperatively and subsequently discharged home from the hospital.

Discussion

Six different subtypes of familial partial dystrophy have been identified in which four forms are inherited autosomal dominant and one form is inherited as autosomal recessive. Marked insulin resistance is associated with this condition and pregnancy induced insulin resistance makes it challenging for good glycaemic control. Multidisciplinary team involvement including foetal maternal specialist, endocrinologist, diabetic clinical nurse specialist, dietician, and neonatologist will provide better maternal and foetal outcome. The preconception care is utmost important with education on risk associated with and the impact of pregnancy on this rare disorder

References

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