An IVF baby with neonatal lupus erythematosus

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Introduction

We report an infant with neonatal lupus erythematosus (NLE) born following in vitro fertilisation (IVF) to a mother who was unaware of an autoimmune illness in herself. NLE is rare in the general population but the literature shows at least seven other IVF babies with this condition [1, 2, 3]. Hormonal manipulation for ovulation induction causing a flare-up of lupus activity is a possible explanation.

Case report

A baby girl weighing 2.55 kg was born by caesarean section following an uncomplicated gestational period of 36 weeks and 5 days. The pregnancy had been assisted by ovulation induction and IVF. Apart from antenatally diagnosed acquired hypothyroidism for which the mother was on replacement therapy, she appeared healthy. Medical investigations performed before IVF did not include screening for SLE.

The baby was diagnosed to have congenital hypothyroidism because thyroid stimulating hormone (TSH) levels were 9.3 miu/ml and 9.4 miu/ml (normal reference < 8.8 miu/ml) at age 16 and 22 days. When treated with thyroxine 12.5 mg/day TSH fell to 4.34 miu/ml.

At the age of 4 weeks a reddish brown, slightly elevated, well defined pigmentation was noticed bilaterally in the preauricular regions. The rash progressed to involve the entire forehead and bridge of nose by 13 weeks. The cheeks, scalp, trunk and limbs were unaffected. Cardiovascular and other systems were clinically normal and she continued to thrive and develop normally. The mother complained of arthralgia and lassitude but had no abnormal physical signs.

When the baby was 13 weeks old serological investigations were performed on both baby and mother. The baby's results did not show either Ro-SSA antibody, or La-SBB antibody and the electrocardiogram, and hepatic and haematologic investigations were normal. The antinuclear antibody and Ro-SSA antibody were positive in the mother. Her serology was negative for La-SBB antibody and antidouble stranded DNA. Mother's ESR was 23 mm.

The rash (figure 1) began fading spontaneously from the age of 7 months and disappeared completely by 9 months.

Discussion

NLE is a rare autoimmune syndrome caused by passively transferred maternal auto-antibodies that react with skin and cardiac muscle. The characteristic rash resolves spontaneously during infancy but cardiac involvement causes lasting damage. Congenital heart block of varying severity caused by fibrosis of the bundle

Figure 1. Skin lesion in neonatal lupus erythematosus: brownish plaques on forehead at 13 weeks.

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of His, haemolytic anaemia, thrombocytopenia or hepatitis are noted in a minority of infants. The transient transplacental IgG antibodies are commonly anti Ro/SS-A, and sometimes anti La/SS-B.

NLE often brings to light latent or obvious autoimmune connective tissue disorder in the mother. Even if she is asymptomatic, the risk of developing systemic lupus erythematosus (SLE), subacute cutaneous lupus erythematosus or Sjogren’s disease in the future remains.

In our report two major criteria stipulated by the American College of Rheumatology i.e. the characteristic rash in the neonate and maternal antibodies to Ro-SSB were positive, confirming the diagnosis.

Literature retrieved from Pub Med database showed reports of 7 other cases of NLE following ovulation induction and IVF. Flare-up of lupus activity in women following ovulation induction is documented and rates are higher after gonadotrophins than after clomiphene therapy [3,4]. Awareness of this and its associated maternal and fetal complications will increase the safety of artificial reproductive technologies in women with SLE [5].

Introduction

Hyperhaemolysis syndrome (HS) is a rare condition where blood transfusions are followed by destruction of both donor and recipient red cells, in the absence of significant detectable red cell antibodies. This phenomenon is reported only in a few children and is extremely rare in thalassaemia syndromes [1, 2, 3, 4]. We report a case of HS in a child with low G6PD enzyme levels and Hb E beta thalassaemia, in whom intravenous immunoglobulin (IVIG), oral methylprednisolone and splenectomy failed but cyclophosphamide therapy resulted in a successful outcome.

Case report

A 5-year old boy born to non-consanguineous parents, from a remote village in the North Central Province of Sri Lanka was found to have severe anaemia and hepatosplenomegaly at 3 ½ years of age. Haemoglobin electrophoresis showed Hb F (11%) and Hb E in the child, Hb E in the mother and elevated Hb A (6.8%) in the father. He received four transfusions of ABO and Rh compatible non-washed red cells in the local hospital and post-transfusion Hb levels were maintained at 10-11 g/dl for ten months. Following the fourth transfusion he became jaundiced (serum bilirubin 94.5 μmol/l, indirect 75.6 μmol/l), passed dark urine (urobilinogen ++ haemosiderin +), and haemoglobin dropped to 3 g/dl. No trigger for acute haemolysis from G6PD deficiency was identifiable. He was admitted to Lady Ridgeway Children’s Hospital, Colombo where repeated blood transfusions with extended phenotypically matched leucocyte depleted red blood cells resulted in further haemolysis and reduction of haemoglobin to below pre-transfusion levels. Extensive

Hyperhaemolysis syndrome in haemoglobin E / beta thalassaemia responding to cyclophosphamide therapy

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