Rings in the Neonate

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Abstract
Neonatal lupus erythematosus (NLE) is an uncommon disease of the neonate. It is believed to be caused by the transplacental passage of maternal autoantibodies to the ribonucleoproteins (Ro/SSA, La/SSB or rarely U1RNP) as these are almost invariably present in NLE sera.[1] The most common clinical manifestations include cutaneous lupus lesions and congenital complete heart block. Hepatobiliary and haematologic abnormalities are reported less frequently.[2,3] We describe a patient with cutaneous NLE to illustrate and raise awareness of the characteristic annular eruption of this condition. We also emphasize the need for thorough investigation for concomitant organ involvement and for maternal education regarding risk in future pregnancies.

Case Report
A 4-week-old female infant was referred with erythematous lesions on the neck, trunk and scalp. The lesions were first noted on the neck when the neonate was 2 weeks old. They spread to the abdomen and scalp despite antibiotic treatment for suspected cutaneous infection. The infant was full-term and delivered by lower segment caesarean section for breech position, with a birth weight of 2.575kg. Her mother, a 23-year-old "primagravida", had no known history of autoimmune disease. Clinical examination revealed erythematous plaques on the right anterior chest, upper abdomen, neck and scalp, which were annular with overlying scale. (Figures 1 and 2) The characteristic "owl eye" or "raccoon eye" appearance of NLE was also noted. Maternal skin examination was normal.

Figure 1: Erythematous annular plaques on the chest and upper abdominal area

Laboratory investigations revealed antinuclear antibody (ANA) positivity and antibodies to the ribonucleoproteins Ro/SSA and La/SSB in the infant and mother. Full blood count and liver function tests from the infant were within the normal range. Cardiac examination was normal and electrocardiogram showed normal sinus rhythm, rate 142 beats per minute. The infant was diagnosed with cutaneous NLE in view of the cutaneous findings and positive serology. Management involved strict photoprotective measures and hydrocortisone 1% ointment topically to affected areas. All the lesions resolved within 6 months with no residual scarring.

Figure 2: Annular lesions on the scalp and forehead

Discussion
NLE is a rare autoimmune disorder, which was first described by McCuiston and Schoch in 1954.[3] The incidence of NLE is estimated to be approximately 1 per 12,500-20,000 live births.[4,5] Infants present with cutaneous lesions in approximately 50% of cases and congenital complete heart block in 50% of cases. Both these manifestations are present concomitantly in 10% of presentations.[5] Other less common presentations include haematological and hepatic abnormalities.[2-5] In our patient cutaneous lesions were first noted at 2 weeks of age. Lesions can be present at birth, but are typically seen days to weeks after birth. They are frequently located on the face and scalp, however lesions can also be found on covered areas such as the trunk and nappy area.[2] The characteristic lesion is an annular macule or plaque sometimes with fine scale or crusting.[6] The diagnosis in our patient was based on characteristic cutaneous findings and positive serology in the infant and mother. Histological findings in NLE are similar to those seen in subacute cutaneous lupus erythematous.[6] In our case all lesions resolved within a 6 month period with no residual scarring. Persistent telangiectasia, dyspigmentation, atrophy, scarring and in rare cases a cutis marmorata telangiectatica congenita (CMTC) like eruption have been reported post cutaneous NLE.[6]

It is recognised that virtually all women who have a child with NLE are anti-Ro positive. Approximately 2% of individuals in the general population are known to be positive for this antibody on serological testing. However despite this, NLE is a rare disorder. It is estimated that the risk for an unselected anti-Ro positive woman to have a baby with NLE may be 2%.[2] Currently the greatest predictor of risk in future pregnancies is having previously had a child with NLE. The estimated risk reported is wide but is thought to be approximately 25%-33%.[2,7] The presenting feature in subsequent pregnancies cannot be predicted as there have been reports of heart block occurring in subsequent pregnancies when the first presentation had been cutaneous features only.[2] Fetal echocardiograms are recommended weekly for at risk pregnancies between 16 and 24 weeks, and biweekly between 26 and 32 weeks gestation.[8]

It is common for women who have a child with NLE to be asymptomatic at the time of birth. However it is recognised that many women go on to develop symptoms of autoimmune disease and should be followed.[8] There have also been reports of development of collagen vascular disease later in life in the affected infant which would suggest that these children are at a greater risk for the development of autoimmune disorders.[9] We report this case to raise the awareness of this rare condition. We emphasize the need for thorough investigation of concomitant organ involvement when a neonate presents with cutaneous NLE. We also stress the need for maternal education as to the risk in future pregnancies and the risk of development of autoimmune disease.

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Comments: