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Blue berry muffin syndrome: a case report

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Abstract: Blueberry muffin rash is a characteristic multiple bluish skin nodules associated with perinatal infection, severe anaemia or neoplastic diseases. We report a term, IUGR, female neonate presenting at birth with reddish blue papulonodular lesions, hepato splenomegaly, cataract, sensorineural hearing loss and congenital heart disease. Investigation revealed, it was a case of congenital rubella syndrome. The skin lesion disappeared spontaneously within five weeks.
Keywords: Blueberry muffin rash, systemic features, congenital rubella syndrome

INTRODUCTION

Blueberry muffin syndrome is the descriptive term used when an infant is born with multiple blue/purple marks or nodules in the skin. The lesion are often generalized, but occur more often on the trunk, head, and neck [1]. These are due to the presence of clusters of blood-producing cells in the skin (extramedullary erythropoiesis), or bleeding into the skin (purpura) or spreading cancer (metastases).The condition was originally considered characteristic of rubella, but is now considered to be potentially associated with many other conditions. In most cases, the lesions evolve into tan macules and fade completely within a few weeks [2]. We report a case of congenital rubella infection in a female neonate presenting with blueberry muffin rash.

CASE REPORT

A term, IUGR, female baby was born to a primi mother by vaginal delivery. Mother was a booked case. She received regular iron and folic acid tablet. Ultrasonography performed at 32weeks did not reveal any abnormality. Antenatally, mother gave history of fever with rash in the first trimester. After routine care it was noticed that, the baby had multiple violaceous maculopapular non blanching lesions, predominantly over the upper trunk and face (Fig A & B). On examination, baby had normal vital signs. Head circumference was 31 cm, chest circumference 28 cm. Liver was palpable 4 cm below right costal margin and spleen was palpable for 2 cm. A systolic murmur was heard in right parasternal area. Echocardiography diagnosed it as a case of small perimembranous VSD. Chest x-ray was normal. USG brain and abdomen did not reveal any abnormality except hepato-splenomegaly. There was bilateral cataract. BERA study revealed sensorineural hearing loss in both ears. Complete blood count showed haemoglobin- 15.4 g/dl, leucocyte count-8,200 but there was thrombocytopenia (platelet count- 50,000). LFT showed conjugated hyperbilirubinemia (bilirubin-8.4, conjugated-4.4) and rise in transaminases. Serology of the neonate showed IgM antibody to rubella virus. Mother was also positive for both IgG and IgM against rubella. VDRL was non reactive. A diagnosis of congenital rubella infection with blueberry muffin rash was made. The baby received supportive management. The rashes began to fade from early 2nd week.

DISCUSSION

The term blueberry muffin baby was initially coined to describe cutaneous manifestations in newborns infected with rubella during 1960s [1,3]. It is a rare neonatal dermatitis characterized by widespread non blanch able, maculopapular bluish colour lesions, due to persistent dermal erythropoiesis [3,4]. During fetal period, haematopoiesis occurs in a number of organs, including the dermis. This activity persists until 34 weeks of gestation. It has yet to be shown whether blueberry muffin lesions are due to persistence or recurrence of this fetal potential[5]. Conditions that cause extra medullary haematopoiesis include intrauterine TORCH group of infections, hematologic dyscrasias like congenital spherocytosis, Rh & ABO blood group incompatibility and anaemia caused by twin-to-twin transfusion. It may also occur in some neoplastic condition like neuroblastoma, congenital leukaemia and Langerhans cell histiocytosis [5, 6]. Initial Evaluation thus should begins with pregnancy history and prenatal laboratory studies. Thereafter complete blood count, LFT, maternal and fetal TORCH, bone marrow examination and skin biopsy may be needed for further delineation of the cause. In our case, clinical examination and the positive rubella serology

helped us to reach the diagnosis. In congenital rubella, rash is observed at birth or rarely some months later[3] In most cases, the rashes regresses without complications within 4-8 weeks. [7].

Fig-A & B: showing violaceous maculopapular non blanching lesions, over the upper trunk and face

CONCLUSION

Blueberry muffin rash is never idiopathic and it may occur because of various causes. Thus, proper examination and investigation is necessary for specific diagnosis and further treatment.

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