CASE REPORT

Blueberry muffin rash at birth due to congenital rubella syndrome

Guruprasada Shetty, Rashmi Kalyanshetti, Habeeb Ullah Khan, Pavan Hegde
Department of Pediatrics, Father Muller Medical College, Mangalore, Karnataka, India

ABSTRACT

We report a term, small for gestational age, female neonate presenting at birth with generalized reddish blue papulonodular lesions, thrombocytopenia, hepatosplenomegaly, cataract, sensorineural hearing loss, patent ductus arteriosus and subependymal cyst. A diagnosis of blueberry muffin rash in a child with congenital rubella syndrome was made based on the clinical and laboratory findings. The rash disappeared by 2 weeks of life.

Key words: Blueberry muffin rash, cataract, congenital rubella syndrome

INTRODUCTION

The term blueberry muffin baby was initially coined by pediatricians to describe cutaneous manifestations observed in newborns infected with rubella during the American epidemic of the 1960s.[1,2] It is characterized by widespread maculopapular lesions of reddish-blue or magenta color caused by persistent dermal erythropoiesis. In congenital rubella, the typical lesion can present at birth, during the first 48 h or, rarely some months later.[2]

The differential diagnosis of blueberry muffin baby includes conditions associated with dermal extra medullary hematopoiesis, infiltrative neoplastic lesions of the skin and cutaneous vascular anomalies. In most cases, the lesions evolve into tan macules and fade completely within a few weeks. When lesions do not fade or when they progress and enlarge, a neoplastic disorder should be suspected.[1-3]

We report a case of congenital rubella infection in a term, small-for-gestational age (SGA) female neonate presenting with blueberry muffin rash.

Access this article online	
Quick Response Code	Website: www.ijpd.in
	DOI: 10.4103/2319-7250.122167

CASE REPORT

A term (37 week), SGA, female neonate was born to a primi mother by normal vaginal delivery. Antenatal period was uneventful except for fever with rash in the first trimester. The mother received basic antenatal care. Ultrasonography performed at 30 weeks showed intrauterine growth retardation (IUGR). Her toxoplasmosis, others, rubella, cytomegalovirus, herpes (TORCH) profile was not done. The baby cried immediately after birth and had multiple reddish blue firm nodular non blanching lesions, varying from 2 to 5 mm all over the body including face and trunk [Figures 1 and 2]. On examination, baby had normal vital signs. Anthropometry showed a Ponderal index of 2.2 favoring symmetrical IUGR. Systemic examination revealed hepatosplenomegaly and systolic murmur in the pulmonary area. Ophthalmological assessment showed bilateral cataract with microophthalmia. Hearing assessment revealed sensorineural hearing loss in both ears. Two dimensional echocardiography with color Doppler examination showed 6 mm size patent ductus arteriosus. Chest X-ray was normal and cranial ultrasound detected subependymal cyst. Hematological investigations showed hemoglobin of 16 g/dl, total leukocyte count of 13300/mm^3 with a differential count of neutrophil (55%), lymphocyte (41%), monocyte (4%) and absolute platelet count of 25,000/mm^3. Blood biochemistry

ADDRESS FOR CORRESPONDENCE
Dr. Guruprasada Shetty,
Department of Pediatrics, Father Muller Medical College,
Mangalore - 575 002, Karnataka, India.
E-mail: gpshettypg33@gmail.com

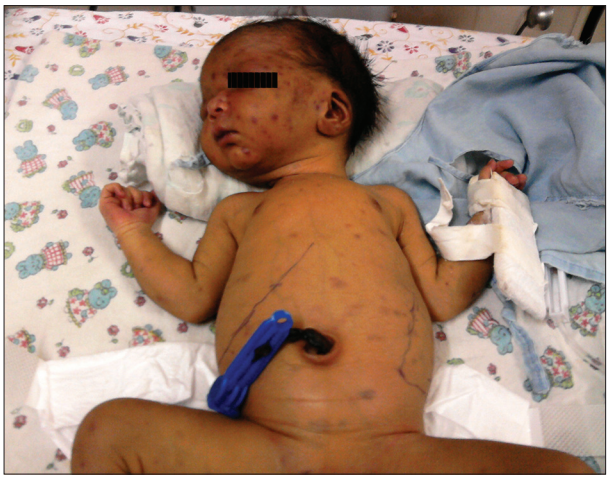

Figure 1: Baby with congenital rubella infection showing blueberry muffin rash. Line markings on abdomen shows hepatosplenomegaly

showed conjugated hyperbilirubinemia (total bilirubin-11.8 mg/dl and direct bilirubin 7.4 mg/dl). Serum rubella specific immunoglobulin (Ig) M was strongly positive in the baby. Mother's blood was positive for both IgG and IgM rubella specific antigens. Screening for other congenital infections was negative.

A diagnosis of congenital rubella infection with blueberry muffin rash was made. The baby received supportive management as per our unit protocol. The rashes began to fade at the end of the 1st week.

DISCUSSION

Blueberry Muffin Syndrome is a rare neonatal skin disorder characterized by widespread non blanchable, maculo papular lesions of reddish-blue or magenta color, due to persistent dermal erythropoiesis in patients with congenital viral infections.[2,4,5] Although the exact cause of prolonged dermal erythropoiesis is unknown, during normal embryologic development extramedullary hematopoiesis occurs in a number of organs, including the dermis; this activity persists until the 5th month of gestation. The presence of blueberry muffin lesions at birth represents postnatal expression of this normal fetal extramedullary hematopoiesis.[1,2]

Conditions that cause extramedullary hematopoiesis include intrauterine infections and hematologic dyscrasias. Among the TORCH group of infections, cytomegalovirus and rubella are the most common viral agents. Non infective causes of blue berry muffin rash are congenital spherocytosis, rhesus hemolytic disease, ABO blood group incompatibility and anemia caused by twin-to-twin transfusion. Lesions resembling the Blueberry Muffin Rash, but with different histology

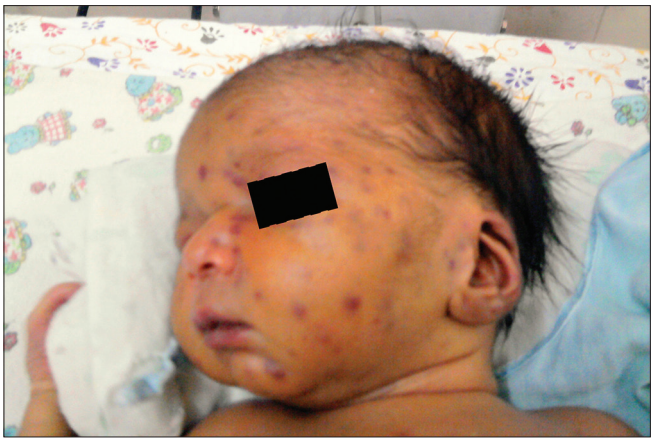

Figure 2: Blueberry muffin rash over face

also occur in neonatal neuroblastoma in congenital monoblastic leukemia and in the congenital alveolar rhabdomyosarcoma.[3,6] Evaluation of blueberry muffin baby begins with careful review of the pregnancy history and prenatal laboratory studies with a special focus on infectious serologies as well as blood type and antibody studies.[3,7,8] In our case, clinical examination and the positive rubella serology pointed to the diagnosis.

In congenital rubella, the blueberry muffin rash is observed at birth or rarely some months later. Histology usually shows dermal erythropoiesis. In most cases, the illness regresses without complications within 4-8 weeks.[5,6]

CONCLUSION

To conclude, though differentials for blueberry muffin baby are extensive, proper examination and investigation is helpful in arriving diagnosis.

REFERENCES

1. Mehta V, Balachandran C, Lonikar V. Blueberry muffin baby: A pictoral differential diagnosis. Dermatol Online J 2008;14:8.
2. Basu S, Gupta AK, Kumar A, Singh P, Kumar A. Congenital rubella syndrome presenting at birth with blueberry muffin rash and communicating hydrocephalus. J Pediatr Neurol 2009;7:423-6.
3. Brisman S, Gonzalez M, Morel KD. Blueberry muffin rash as the presenting sign of Aicardi-Goutières syndrome. Pediatr Dermatol 2009;26:432-5.
4. Hsiao YW, Tseng FW, Shih YL, Kuo TT, Jaing TH, Hui CY. Blueberry muffin baby with acute myeloid leukemia and spontaneous remission. Dermatologica Sin 2011;29:47-9.
5. Vozza A, Tolone C, Carrano EM, Di Girolamo F, Santinelli R, Ascierto PA, *et al*. Late onset Blueberry Muffin Syndrome following congenital rubella. J Eur Acad Dermatol Venereol 2003;17:204-5.
6. Koklu E, Kurtoglu S, Akcakus M, Koklu S, Gunes T, Kose M.

Blueberry muffin syndrome owing to congenital rubella: Case report. Ann Trop Paediatr 2006;26:149-51.

7. Gaffin JM, Gallagher PG. Picture of the month. Blueberry muffin baby (extramedullary hematopoiesis) due to congenital cytomegalovirus infection. Arch Pediatr Adolesc Med 2007;161:1102-3.
8. Piersigilli F, Danhaive O, Auriti C. Blueberry muffin baby due to alveolar rhabdomyosarcoma with cutaneous metastasis. Arch Dis Child Fetal Neonatal Ed 2010;95:F461.

How to cite this article: Shetty G, Kalyanshetti R, Khan HU, Hegde P. Blueberry muffin rash at birth due to congenital rubella syndrome. Indian J Paediatr Dermatol 2013;14:73-5.

Source of Support: Nil, **Conflict of Interest:** None declared