Blueberry muffin syndrome owing to congenital rubella: case report

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Abstract A case of congenital rubella is reported in a 22-day-old boy presenting with a ‘blueberry muffin’ rash. Late-onset blueberry muffin syndrome following congenital rubella is very rare.

Introduction

Blueberry muffin syndrome is a cutaneous manifestation characterised by widespread maculopapular lesions of a reddish-blue or magenta colour caused by persistent dermal erythropoiesis. In congenital rubella, the typical lesion can present at birth, during the 1st 48 hours or, rarely, some months later. Blueberry muffin lesions have been recorded in patients with other congenital viral infections, and in a variety of congenital haematological disorders.1

Congenital rubella affects virtually all organ systems.2 The most common manifestation is intra-uterine growth retardation. Associated features include cataract (which is frequently associated with microphthalmia), myocarditis and structural cardiac defects, hearing loss, meningo-encephalitis and, rarely, blueberry muffin skin lesions. Persistent infection leads to pneumonia, hepatitis, thrombocytopenia and anaemia.

Case Report

The infant was first admitted to another hospital at the age of 3 days with vomiting, low birthweight and convulsions owing to hypoglycaemia. Blood chemistry showed hypoglycaemia and slightly elevated transaminases and there was occult blood in the faeces. Echocardiography showed peripheral pulmonary artery stenosis. He was discharged on the 6th of life with a diagnosis of transient hypoglycaemia because of low birthweight and peripheral pulmonary artery stenosis. The infant was referred to Gevher Nesibe Hospital at 22 days of age with skin lesions (Figs 1 and 2). His mother reported an undefined low-grade fever and generalised maculopapular rash at 8 weeks gestation. Physical examination showed multiple dark-bluish skin nodules with purple maculopapular lesions covering the face, limbs and all parts of the trunk, and there was hepatosplenomegaly. A diagnosis of blueberry muffin syndrome was made.

The results of investigations were as follows. Haemoglobin was 17.2 g/dl, white cell count $22.3 \times 10^9$/L, platelets $22 \times 10^9$/L, total serum bilirubin 356 mmol/L (conjugated 133 mmol/L, unconjugated 223 mmol/L),
serum aspartate aminotransferase 599 IU/ml and alanine aminotransferase 295 IU/ml. Rubella serology supported congenital rubella: IgM 16.5 IU/ml and IgG 236 IU/ml. The following congenital infections were excluded on serology: syphilis, cytomegalovirus, toxoplasmosis, herpes simplex, parvovirus, coxsackie B2 and hepatitis B and C. Abdominal ultrasound demonstrated hepatosplenomegaly with an unhomogeneous echotstructure. Ophthalmology examination was normal. A bone marrow examination to exclude malignancy was normal. The parents refused a skin biopsy on their infant.

The bilirubin and transaminase levels increased progressively up to 37 days of life. On the 47th day of admission, the laboratory findings began to improve but, unfortunately, the infant died of nosocomial infection on the 57th day of life.

Discussion

Blueberry muffin rash is symptomatic of an infection or disease and the prognosis depends on the cause. Blueberry muffin lesions are found in congenital rubella, cytomegalovirus, toxoplasmosis, herpes simplex, coxsackie B2 and parvovirus and also in a variety of congenital haematological and malignant disorders. Lesions resembling the blueberry muffin rash but with different histology also occur in neonatal neuroblastoma (skin metastases of neuroblastoma), congenital monoblastic leukaemia (leukaemia cutis) and congenital alveolar rhabdomyosarcoma (skin metastases of rhabdomyosarcoma). Cutaneous erythropoiesis is considered to be owing to persistence or reactivation of fetal dermal erythropoiesis secondary to prolonged, severe intrauterine anaemia. In this case, the positive rubella serology pointed to the diagnosis.

In congenital rubella, the blueberry muffin rash is observed at birth or, rarely, some months later. It is associated with vomiting, diarrhoea, poor weight gain, deafness, interstitial pneumonia and hepatitis. Histology usually shows dermal erythropoiesis. In most cases, the illness regresses without complications within 4–8 weeks.

References


