PAEDIATRIC DERMATOLOGY

CONGENITAL SELF-HEALING RETICULOHISTIOCYTOSIS PRESENTING AS A BLUEBERRY MUFFIN RASH

W Hua [1] - L Li [1]

West China Hospital, Sichuan University, Division Of Dermatovenerology & Cosmetics Evaluation Center, Chengdu, China [1]

Background: Congenital self-healing reticulohistiocytosis (CSHRH) is a rare, cutaneous, self-limited form of Langerhans cell histiocytosis. There have been very few reports in Asian populations.

Observation: A baby girl born at term via spontaneous vaginal delivery to a healthy mother was noted to have a widely-disseminated blueberry muffin rash at birth. The pregnancy had been uncomplicated and the patient was otherwise healthy-She was active, afebrile, and feeding well. She had Apgar scores of 9 at 1 min and 10 at 5min. On examination she had numerous purple, red or dull red papules (1-6 mm diameter), affecting the face, scalp, trunk, back, diaper region, and four extremities. There was no involvement of the mucous membrane, palpable lymph node or hepatosplenomegaly. Routine blood cell counts and blood chemistry, urine and stool routines were within normal limits. Congenital infections including TORCH, TPPA, Tzanck smear, HIV antibody showed normal or negative findings. A skin biopsy was done on a papule 9 days after birth. The lesion showed a diffuse mononuclear cell infiltrate in the epidermis and dermis. Immunohistochemistry showed S100, CD1a, Langerin (CD207) and CD68/PGM-1 positivity, characteristic of Langerhans cell histiocytosis. BRAF mutation testing (for prognostic evaluation) was negative. Rapid spontaneous involution of the lesions was noted. By day 7, the blue-purple papules had flattened and faded. By day 19, about a quarter of the initial size of lesions remained, some lesions leaving postinflammatory hyperpigmentation or scars only, and 4 months later, the skin lesions had cleared. Based on the clinical course and typical histopathological findings, a diagnosis of CSHRH was made.

Key message: The “blueberry muffin baby” was usually caused by infectious diseases. We report on a rare case of CSHRH which presenting as a blueberry muffin rash.