Case Report

Michelin Tire Baby Syndrome — A case report and literature review

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Abstract

Michelin tire syndrome is described in a two month old infant of Filipino-Saudi parents. The infant had generalized excessive folding of skin and facial dysmorphism. The skin biopsy showed excessive adipose tissue in reticular dermis, papillary dermis and around adnexa. Spontaneous partial improvement in skin folding was noted on follow up. To the best of our knowledge, this is the first case ever reported locally of Michelin tire.

Introduction

Michelin Tire Baby Syndrome (MTBS) (Synonym: Kunze-Riehm syndrome) is a rare genetic disorder, characterized by generalized folding of excess skin that may be isolated,[1] or may be associated with additional phenotypic abnormalities[2,3] and probably reflect multiple underlying

disorders. Circumferential skin fold is a rare finding at birth. One or few constrictions involving the limbs may be a part of amniotic band sequence. Multiple, symmetric, ring-like lesions involving the extremities and trunk are seen in a benign hamartomatous condition of the skin known as MTBS. It is a rare genodermatosis (Mendelian Inheritance in Man - ID no 156610 - skin creases, multiple benign ring-shaped, of limbs).

Since the first description by Ross in 1969,[1] only 20 patients have been reported in literature.

Case Report

A two months old baby was brought to our clinic for vaccination. This baby was the first child of Saudi-Filipino parents and was delivered at term after normal pregnancy with birth weight of 3.9Kg and length of 50cm. Clinical examination revealed multiple, asymmetric, deep, gyrate skin folds involving extremities and shoulders as shown in Figure 1 and 2. She had a characteristic round face with hypertelorism, depressed nasal bridge and a thin, down-turned vermillion border of upper lip. There was no hypertrichosis and no other cutaneous or extracutaneous abnormalities were noted. Anthropometric measurements were within normal limits. Systemic examination was unremarkable. None of her family members had history of similar skin folds. Baseline investigations did not reveal any abnormal findings and ultrasound of abdomen and pelvis were normal.

A diagnosis of Michelin Tire Baby Syndrome (MTBS) was made. A skin biopsy was done from a deep fold on the right arm. Histopathology Hematonylin and Eosin (H and E) showed normal epidermis and dermis of normal thickness but with excessive adipose tissue in the reticular dermis, papillary dermis and around adnexa. Special staining with mucin was negative and histological diagnosis was suggestive of lipomatous nevus. The child was regularly followed in the clinic and spontaneous partial improvement in skin folds was observed over shoulders, upper limbs and lower limbs. Systemic examination was normal and the developmental milestones were normal for age.

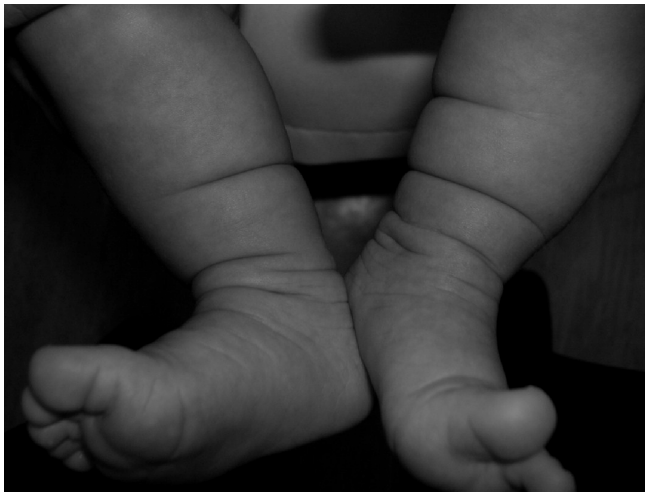

Figure-1: Showing circumferential skin folds over legs.

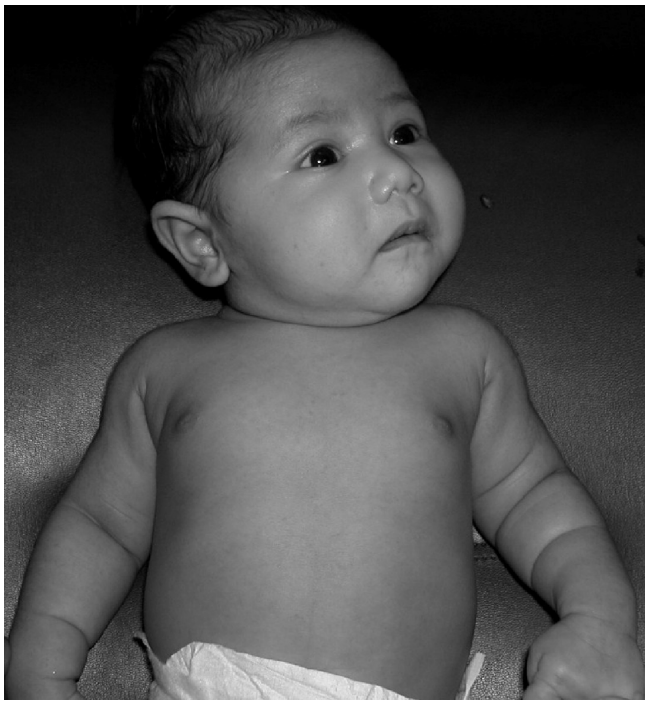

Figure-2: Shows skin folds over upper limbs.

Discussion

Michelin tire baby syndrome (MTBS) is characterized by ring-shaped skin creases involving primarily the limbs and neck and was first described by Ross,[1] who used this descriptive term MTBS because of physical resemblance of these patients to the mascot of a French Tire manufacturer. The pattern of skin creases tends to be symmetrical, temporary and does not adversely affect the patient's health.

The condition may be familial and there is evidence of autosomal dominant mode of inheritance affecting several members in successive generations of a family.[4] Kunze and Riehm[5] and Niikawa et al[6] reported the disorder in two families with this syndrome. In the first family the trait occurred in three generations.[6] Bass et al[4] described the disorder in seven individuals in four generations with three instances of male-to-male transmission. Spontaneous resolution took place in cases reported by Kunze and Riehm[5] and Bass et al.[4] The pathogenesis is yet unclear. Chromosomal anomalies detected in this condition are deletion of the short arm of chromosome 11[2] and paracentric inversion of the long arm of chromosome 7.[7]

In MTBS multiple, asymptomatic, circumferential folds simulating skin bands are usually present since birth, and the most common presentation is involvement of extremities, trunk, palms and soles. Other congenital anomalies have been described including craniofacial anomalies, cleft palate, hypoplastic scrotum and hernias (inguinal and umbilical).[8] Other abnormalities reported includes left sided hemihypertrophy,[1] hemiplegia and microcephaly,[2] stellate scarring,[3] developmental delay, and smooth muscle hamartoma which may be diffuse.[3,9] Psychomotor retardation, epilepsy and joint hypermobility have also been described.[2] Hypertrichosis has been observed in two patients.

The cause of the dermatomegaly may be a diffuse lipomatous nevus involving the deeper dermis as reported in the first patient.[1] Hence the name 'congenital diffuse lipomatosis' has been attributed to this condition.[1] Underlying smooth muscle hamartoma may be present. Scarring, instead of increased adipocytes or muscle fibers has been reported. In HITCH syndrome, degenerative changes in the deep dermis with normal distribution of elastic fibers were demonstrated.

In our case the patient presented with multiple skin creases, some facial dysmorphism with normal systemic examination and developmental milestones. Most of the investigations done were normal and histologic examination showed an underlying lipomatous naevus. There was no family history of similar condition and the patient showed improvement over a period of ten months. It may be concluded that MTBS is not only variable in clinical presentation but shows two distinct histologic abnormalities. These may be better studied by biochemical and molecular biologic analysis to delineate exact structural alterations in the mesenchymal tissue and by a karyotype analysis.

References

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