Circumferential skin folds in a child: A case of Michelin tire baby syndrome

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ABSTRACT

A six-month-old girl who presented with dermatitis was found to have multiple, symmetric, deep, gyrate skin folds involving her trunk and similar circumferential lesions on her extremities since birth. She had a characteristic round face with hypertelorism, depressed nasal bridge, thin, down-turned vermillion border of upper lip and short neck. Skin biopsy demonstrated increased smooth muscle fibers in the deeper dermis. A diagnosis of Michelin tire baby syndrome was made. Clinical features, histopathology, differential diagnosis and prognosis of this rare disorder have been discussed.

Key Words: Circumferential skin folds, Congenital diffuse lipomatosis, Michelin tire baby

INTRODUCTION

Circumferential skin fold is a rare finding at birth. One or few constrictions involving the limbs may be 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 Michelin tire baby syndrome (MTBS). It is a rare genodermatosis (MIM % 156610), approximately 20 cases of which have been reported to date.[1] The detailed description of this condition was given by Ross in 1969.[2] However, Wiedemann has described this as 'a congenital anomaly existing from the beginning of the mankind' as this feature was depicted in the son of Eve in a bronze door inscription at the cathedral of Hildesheim in northwestern Germany.[3]

CASE REPORT

A six-month-old female child with a clinical suspicion of Cushing's syndrome was referred from the Department of pediatrics for the management of itchy skin lesions. The child had an intensely pruritic, oozy dermatitis involving the face and extensor extremities. In addition, there were multiple, symmetric, deep, gyrate skin folds involving her trunk and similar circumferential lesions on her extremities [Figures 1 and 2]. She had a characteristic round face with hypertelorism, depressed nasal bridge, thin, down-turned vermillion border of upper lip and short neck [Figure 3]. The folds were present since birth and were asymptomatic without causing any physical discomfort to the child. The parents did not have any complaint regarding these as it was perceived by them as a sign of good health.

She did not have any major illness in the past necessitating prolonged treatment. Though there was a resemblance to Cushing's syndrome, detailed examination did not reveal any features suggestive of this condition. Her body weight was 8.5 kg. Other anthropometric measurements were within normal limits and developmental milestones were normal. Several of her family members had history of atopy but none had similar skin folds.

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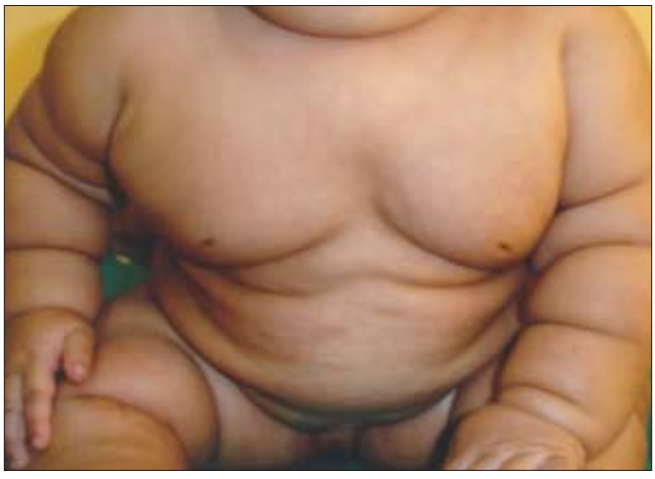

Figure 1: Deep ring-like folds over extremities

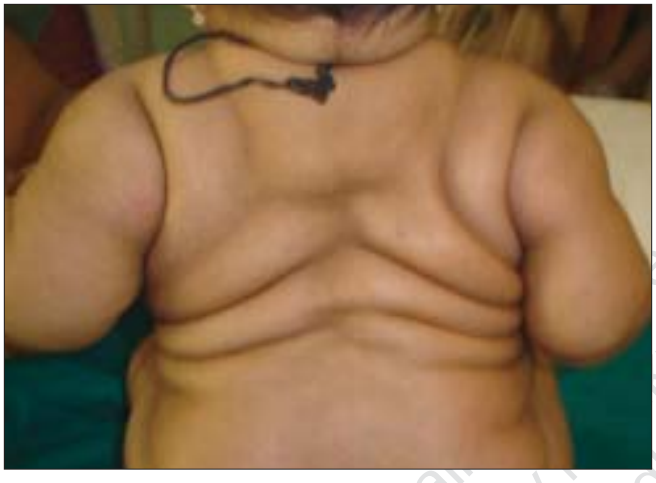

Figure 2: Symmetrical gyrate skin creases on back

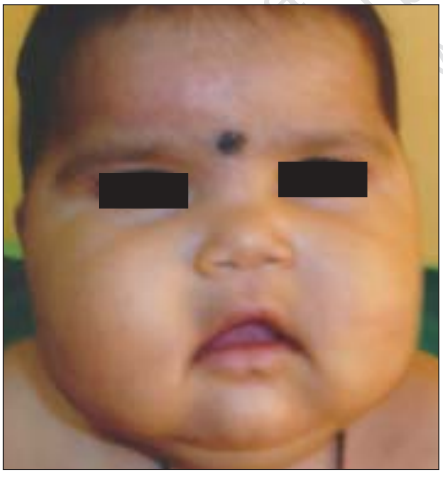

Figure 3: Typical facies with hypertelorism, depressed nasal bridge and down-turned upper lips

A diagnosis of MTBS was made. A skin biopsy was done from a deep fold on the right arm. Histopathology (H and E) showed normal epidermis and papillary dermis. There were scattered bundles of smooth muscle fiber in the reticular dermis. Masson trichrome stain revealed presence of increased smooth muscle fibers.

The child's dermatitis improved with a short course of systemic steroids, antihistamines and emollients. The cause and nature of the skin folds were discussed with the parents and they were counseled about the self-limiting course of this disorder.

DISCUSSION

Ross coined this descriptive name 'MTBS' because of the physical resemblance of these patients to the mascot of a French tire manufacturer.[2] It is a hamartomatous disorder involving either adipose tissue or smooth muscle in the deeper dermis of the skin.

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] The pathogenesis is yet unclear; chromosomal anomalies detected in this condition are deletion of the short arm of chromosome 11[5] and paracentric inversion of the long arm of chromosome 7.[6] Multiple, asymptomatic, circumferential folds, simulating skin bands are the most prominent feature of MTBS. These are usually present since birth. Extremities are the commonest sites of involvement, others being the trunk, palms and soles. Hypertrichosis of the involved areas may be observed.[7,8]

Multiple congenital anomalies have been described as associations. These include facial dysmorphism, as described in our patient, long and curled eyelashes, bushy eyebrows and bilateral epicanthic folds.[8] Low-set ears, cleft lip and palate, hypoplastic teeth and mandible may be present.[8] Other features include developmental delay, seizure disorders, pectus excavatum, congenital heart defect and undescended testis with abnormal histology.[3,8,9]

The cause of the dermatomegaly may be a diffuse lipomatous nevus involving the deeper dermis as reported in the first patient.[2] Hence the name 'congenital diffuse lipomatosis' has been attributed to this condition.[2,8] Underlying smooth muscle hamartoma may be present[8] as seen in the index case. Scarring, instead of increased adipocytes or muscle fibers has been reported.[10] Sato *et al.*[11] have reported fragmentation of elastic fibers in addition to increased smooth muscle tissue in their patient and suggested that abnormal elastic fiber formation may be a pathogenetic factor in this condition.

The affected children are otherwise normal. The skin folds gradually diminish and disappear without any intervention.[8,12] In familial cases, older family members may show few remnants of deep skin folds.

Congenital amniotic bands are usually solitary and present on limbs. These may be part of amniotic band sequence, present in association with multiple congenital anomalies. Beare-Stevenson cutis gyrata syndrome is associated with dermatomegaly which is localized to the scalp, forehead, face and neck.[13] Some authors believe that MTBS may not be a single disorder but a clinical finding associated with many other conditions.[13]

Circumferential skin creases have been included as a component of some recently described entities like HITCH syndrome (Hearing Impairment, undescended Testis, Circumferential skin creases and mental Handicap)[14] and multiple congenital anomalies and mental retardation (MCA/MR) syndrome.[15]

One patient of MTBS has been reported from India, who had mild affection and there was spontaneous involution of the skin creases during childhood.[12] Our patient has been kept under regular follow-up for the evaluation of course of the disease.

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