Journal of Pakistan Association of Dermatologists 2009; **19**: 109-111.

Case Report

Michelin tire syndrome

Farid-ur-Rehman, Gul Hussain*

Department of Dermatology, Foundation University Medical College, Rawalpindi
*Department of Pediatrics, Foundation University Medical College, Rawalpindi

Abstract We describe a 5-year-old girl who has symmetrical ringed skin folds associated with underlying smooth muscle hamartoma. Cutaneous findings in this child resemble those in other reported cases of Michelin tire syndrome. In addition to the skin changes she has multiple phenotypical anomalies. These include distinctive facial dysmorphia, and genital, ophthalmologic, skeletal and central nervous system anomalies.

Key words
Michelin tire syndrome

Introduction

Ross coined Michelin tire syndrome in 1969, when he reported a child with generalized skin folding who had a fanciful resemblance to the symbol of French tire manufacturer (**Figure 1**).[1]

This syndrome is also known as generalized folded skin. At birth, as the name implies, the skin is thrown into folds but it is not lax and feels rather thick. With age, the skin folds spontaneously improve[2] and in childhood these are mainly confined to the extremities. Hence, it is also termed as symmetrical ringed creases of the extremities. It is a rare syndrome and can be inherited in an autosomal dominant pattern,[3] as suggested by two reports in which several members of the same family were affected. The histopathological findings are variable[2] and include diffuse lipomatous hypertrophy, generalized smooth muscle hamartoma to unremarkable skin changes. Hypertrichosis has been described in cases associated with smooth muscle hamartoma.

Address for correspondence
Dr. Farid-ur-Rehman, Associate Professor
Dermatology Department,
Foundation University Medical College, Rawalpindi
E mail: dr_faridurrehman@yahoo.com

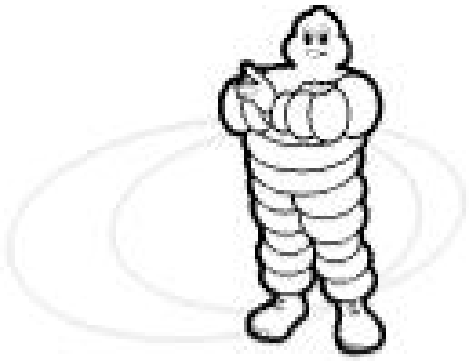

Figure 1 Symbol of French tyre manufacturer

Michelin tire syndrome may be an isolated abnormality or it may be associated with other phenotypic abnormalities.[2]

Case report

A five-year-old girl was referred from the Pediatrics Department to the Dermatology Department, Foundation University Medical College for peculiar skin changes.

She was born to consanguineous parents after a full term gestation through normal delivery. At birth, she had increased skin folds all over he body including the trunk and all four limbs, excessive hair growth and a dark complexion. There was no family history of similar disease. With advancing age, her parents observed

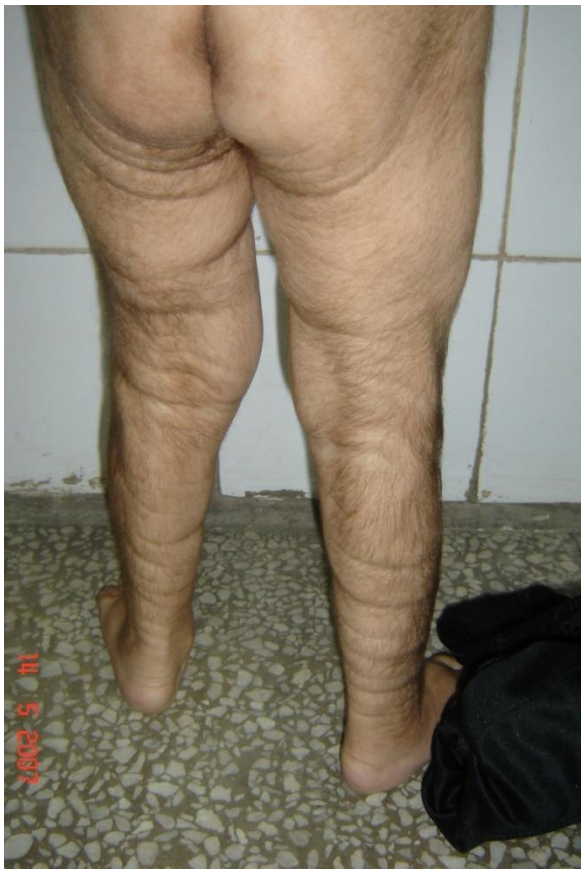

Figure 2 Lower limbs, showing ringed creases, hyertrichosis and hyperpigmentation.

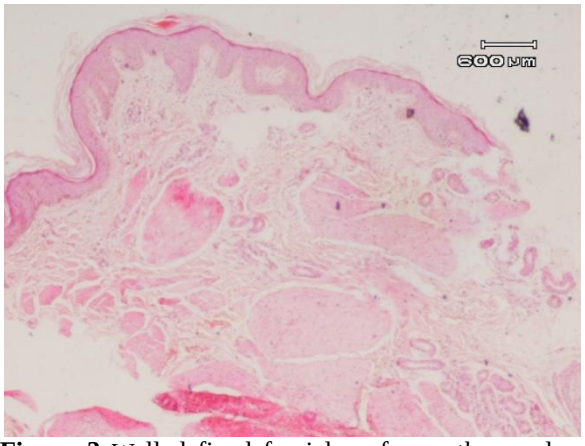

Figure 3 Well defined fascicles of smooth muscles arranged haphazardly in the reticular dermis.

marked reduction in the number and depth of skin folds along with the number of hairs and hyperpigmentation. At the age of two years she developed generalized tonic clonic fits.

At the time of examination, there were multiple, symmetrical skin folds on all four limbs. These skin folds were more numerous and deep at the distal ends (**Figure 2**). On palpation, the skin on the extremities was thick and was neither lax nor hyperelastic. There was bilateral symmetrical hypertrichosis and hyperpigmentation at the involved areas.

The remainder of the physical examination revealed a dysmorphic girl who was 90 cm tall (less than 3rd percentile), weighed 11 Kg (50% percentile of National Center for Health Statistics [NCHS]). The child had thick eyebrows with thick everted lips. The nasal bridge was wide. The oral cavity and teeth were unremarkable. The ears were low set. The neck was shorter and wider than normal with an excessive number of skin folds.

Gynecological examination revealed thin hair growth on mons, labia majora was flat without fat and labia minora were absent. There was clitoromegaly. Eye examination revealed disc pallor and multiple flecks on the retina. Skeletal abnormalities included valgus deformities of both knees and ankles. There was overlapping of toes. Joints were normal. The child had a wide based gait. Left ovary was absent, the only abnormality in abdominopelvic USG. Radiological examination of the bones revealed bone age of three years. Metabolic causes of seizures were ruled out, EEG was unremarkable and CT scan of brain showed an arachnoid cyst. Congenital adrenal hyperplasia was excluded, as the hormonal profile; including serum testosterone, 17-hydroxyprogesterone and ACTH were within normal limits.

Histopathologic examination

The histopathological examination **(Figure 3)** revealed numerous well-defined fascicles of smooth muscles arranged haphazardly in the reticular dermis. The epidermis and subcutaneous tissue was unremarkable.

Discussion

Based on skin folds, hypertrichosis, dysmorphic facial features, tonic clonic fits, abnormal genitalia including clitoromegaly and retarded bone age, Michelin tire syndrome and leprerchaunism were included in the differential diagnosis. Glucose tolerance test and plasma insulin levels fasting and post-parandial were unremarkable and the histopathological findings of smooth muscle hamartoma instead of few and fragmented elastic and collagen fibers as seen in Leprerchaunism,[4] confirmed the diagnosis of Michelin tire syndrome.

The characteristic histopathological findings in Michelin tire syndrome of well-demarcated fascicles of smooth muscles oriented haphazardly in the reticular dermis with or without connections to the hair follicles are similar to localized smooth muscle hamartomas.[5] But some of the latter show epidermal changes including, acanthosis, papillomatosis and keratinocyte hyperpigmentation.[3] Clinically, localized smooth muscle hamartomas present as patch, plaque or papules. Becker's naevi histopathologically, also show smooth muscle hyperplasia but have associated epidermal acanthosis, papillomatosis and hyperpigmentation with distinctive finding of increased number of melanocytes.[3]

Our patient shares some of the other previously reported phenotypic anomalies[3] including low set ears, overlapping of toes,[6] valgus deformities of knees and ankles,[6] seizures and bone growth retardation. Features that differentiate our patient from all other patients described earlier include absence of labia majora and minora, clitoromegaly, multiple flecks on the retina and disc pallor and arachnoid cyst on CT scan of brain.

References

1. Ross CM. Generalized folded skin with an underlying lipomatous nevus. *Arch Dermatol* 1969; **100**: 320-3.
2. Atherton DJ, Moss C. Naevi and other developmental defects. In: Burns T, Breathnach S, Cox N, Griffiths C, eds. *Rook's Textbook of Dermatology, 7th edition*. London: Blackwell Science; 2004: 15.1-15.114.
3. Schnur RE, Herzberg AJ, Spinner N *et al*. Variability in Michelin Tire syndrome. *J Am Acad Dermatol* 1993; **28**: 364-70.
4. Burrows NP, Lovell CR. Disorders of connective tissue. In: Burns T, Breathnach S, Cox N, Griffiths C eds. *Rook's Textbook of Dermatology, 7th edn*. London: Blackwell Science; 2004:46.1-46.71.
5. Albrecht S. Neoplasia and hyperplasia of neural and muscular origin. In: Freedberg IM, Arthur ZE, Wolff K *et al*., eds. *Fitzpatrick's Dermatology in General Medicine, 6th edn*. New York: McGraw Hill; 2003. p. 1033.
6. Mallory SB, Leal-Khouri F, eds. *An Illustrated Dictionary of Dermatological Syndromes.* New York: The Parthenon Publishing Group; 1994.