

## **A rare case of Michelin tire baby syndrome with achondroplasia in a newborn baby**

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### **Abstract**

**Michelin Tire Baby Syndrome (MTBS) also known as Kunze-Riehm syndrome is a rare genetic condition with a rare disease of unclear etiology and pathogenesis with generalized folding of excess skin manifesting clinically as symmetric ring-like excessive skin folds mostly affecting the extremities. The familial cases have been reported to have an AD pattern of inheritance. It may or may not be associated with chromosomal abnormalities. A rare association of Michelin tire baby syndrome with Achondroplasia was Published in 01-Apr-2020 - India. The following report is the first case of a rare association of Michelin tire baby syndrome with Achondroplasia in Yemen.**

**Keywords:** Kunze-Riehm syndrome, Michelin tire baby syndrome, Achondroplasia.

*Accepted on 8<sup>th</sup> August, 2020*

### **Introduction**

A Michelin tire baby syndrome (also known as "Folded skin with scarring", It was originally described by Ross in 1969, it is characterized by multiple, symmetric, circular skin creases, or bands, on the forearms, lower legs, and often the neck that are present at birth. The creases disappear later in life. They are reminiscent of those of Bibendum, the mascot of the tire manufacturer, Michelin, hence the name of the syndrome. Associated abnormalities vary and may include facial dysmorphism, upslanting palpebral fissures, hypertelorism, cleft palate, genital anomalies, mild developmental delay, ureterocele, smooth muscle hamartoma, nevus lipomatosis, Laron syndrome (dwarfism with high growth hormone and low somatomedin activity), and other defects [1-5]. The familial cases have been reported to have an AD pattern of inheritance. It may or may not be associated with chromosomal abnormalities. Although the deletion of the short arm of chromosome 7 and paracentric inversion of the long arm of chromosome 11 have been implicated as the cytogenetic abnormalities in a few cases 5, however, some cases have not been reported to be associated with any chromosomal abnormalities. Achondroplasia is a condition inherited as an autosomal dominant disorder that results in disordered growth. The pathogenesis is unknown. A rare association of Michelin tire baby syndrome with Achondroplasia was Published in 01-Apr-2020 - India.

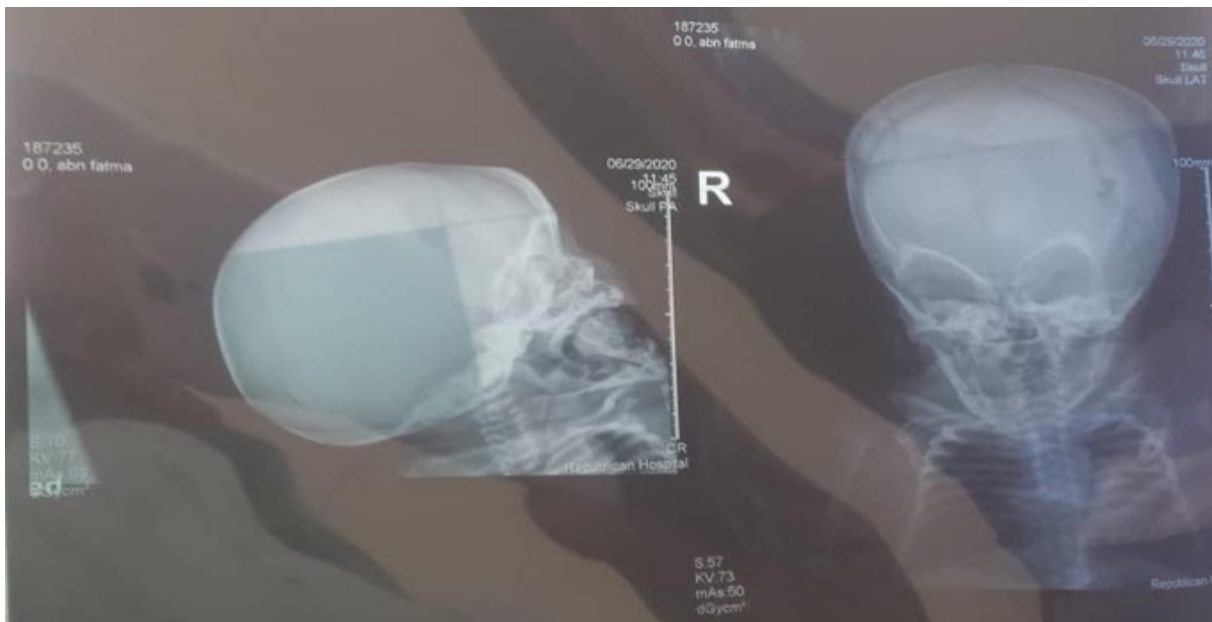
### **Case Presentation**

A Yemeni seven days old female baby, born of a consanguineous marriage, was presented to Algomhori

teaching hospital nursery complaint of the difficulty of breathing with peripheral cyanosis. The mother was primigravida with no medical complaint during pregnancy and has no regular antenatal care. Her last U/S report during antenatal period show 41weeks and 3 days fetus with polyhydramnios. The patient was born by C/S delivery. The one minute APGAR score was 4/10 and 7/10 at five minutes. Her birth weight 3.2 Kg (50th percentile). The length was 43 cm (below 5th percentile). The head circumference was 37 cm (95th percentile), the upper to lower segment ratio was 2:1. She had no family history of the same condition or any chronic illness. The physical examination revealed multiple, systemic excessive skin folds of her upper and lower extremities and bilateral epicanthic folds. Disproportionally large head to the body size difference, with prominent forehead (frontal bossing). Midfacial hypoplasia, with a flattened nasal bridge with anteverted nares. Low set ears, short neck, and narrow chest. Short arms and legs especially the upper arm and thigh. The proximal limbs are short (called rhizomelic shortening). She has short fingers and toes with a trident broad hand. The cardiovascular system revealed PDA, ASD, and pulmonary hypertension. The musculoskeletal system examination revealed mild generalized hypotonia. The rest of the systemic examination was unremarkable. Ultrasound of the abdomen and pelvis were normal as well. Skull X-ray show enlarged skull vault with a small skull base. The parents did not do a skin biopsy and genetic testing for Michelin tire and Achondroplasia due to financial limitations (Figures 1 and 2).



**Figure 1.** Clinical manifestation of Michelin tire baby syndrome with Achondroplasia.



**Figure 2.** Skull X ray, AP and lateral view.

## Discussion

Michelin tire baby syndrome (MTBS) was described in 1969 and so named because of the physical resemblance to the “Michelin Man” logo of the French tire manufacturer [6]. There are approximately around 30 cases reported in the literature. According to our literature review: the first case to be associated with achondroplasia was Published in 01-Apr-2020 - India by Sayan Banerjee. This case according to my knowledge is the second case of this association and is the first reported case of Michelin tire baby syndrome with Achondroplasia in Yemen. The diagnosis of MTBS is made clinically by the appearance of the characteristic skin creases. Various accompanying craniofacial abnormalities including microcephaly, cleft lip, and palate, thickened epiglottis, deformed ear, and nose, up- slanting palpebral fissures, have

been described [7]. These associations were different in each case. In our case, the baby has the characteristic skin folds of MTBS and bilateral epicanthic folds. Large head with frontal bossing, midfacial hypoplasia, with a flattened nasal bridge with anteverted nares, low set ears, short neck, and narrow chest. Short arms and legs especially the upper arms and thigh. The proximal limbs are short (called rhizomelic shortening). She has short fingers and toes with a trident broad hand. No hypertrichosis or other cutaneous lesions were found. The various anomalies that are known to be associated with MTB include, congenital heart disease, left-sided hemihypertrophy, hemiplegia, inguinal and umbilical hernia, hypoplastic scrotum, joint hypermobility, and stellate scarring. Epilepsy, developmental delay, growth retardation, and psychomotor retardation have also been reported [8]. In the literature search,

a case of MTBS was associated with panhypopituitarism. MTB has been reported in otherwise healthy individual's too [9]. In the case, the baby presented with congenital heart disease in the form of PDA, ASD, and pulmonary hypertension, with revealed mild generalized hypotonia. The patient did not have any organomegaly and the remaining systemic examination was unremarkable. Ultrasound of the abdomen and pelvis were normal as well. Skull X-ray show enlarged skull vault with a small skull base. The familial cases have been reported to have an AD pattern of inheritance. It may or may not be associated with chromosomal abnormalities. Although the deletion of the short arms of chromosome 7 and paracentric inversion of the long arm of chromosome 11 have been implicated as the cytogenetic abnormalities in a few cases [5], however, some cases have not been reported to be associated with any chromosomal abnormalities. The most common histological findings in the affected skin of MTB are smooth muscle hamartoma and nevus lipomatosis (with or without overlying hypertrichosis) [9]. In the present case, the diagnosis of Michelin tire baby syndrome with Achondroplasia was made by clinical presentation, because the parents did not give their consent for skin biopsy for histopathologic examination, and genetic testing for Michelin tire and achondroplasia could not be undertaken due to financial limitations. Sardana et al. in 2003 [10], have reported a case of spontaneously partially improving MTB syndrome in one year of follow-up. The baby of the present case was scheduled for frequent visits to monitor the progress of the skin folds and manage congenital heart defects and short stature.

## Conclusion

Michelin tire baby syndrome is one of the rarest cases worldwide in which its association with other syndromes and its manifestation and complication is still obscured. Those babies need close and long term follow-up and management by the multi-disciplinary team includes paediatricians, dermatologists as well as genetics specialists.

## Informed Patient Consent

The authors certify that he has obtained all appropriate parents' consent forms. In the form of; the parents have given their consent for their baby images and other clinical information to be reported in the journal. The parents understand that their baby names and initials will not be published.

## References

1. Bass HN, Caldwell S, Brooks BS. Michelin tire baby syndrome: Familial constriction bands during infancy and

early childhood in four generations. *Am J Med Genet* 1993; 45: 370-372.

2. Schnur RE, Herzberg AJ, Spinner N, et al. Variability in the Michelin tire syndrome. A child with multiple anomalies, smooth muscle hamartoma, and familial paracentric inversion of chromosome 7q. *J Am Acad Dermatol* 1993; 2: 364-370.
3. Schnur RE, Zackai EH. Circumferential ringed creases ("Michelin tire babies") with specific histologic findings and/or karyotype abnormalities: Clues to molecular pathogenesis?. *Am J Med Genet* 1997; 2: 221.
4. Pivnick EK, Wilroy RS, Martens PR, et al. Hypertrichosis, pigmentary retinopathy, and facial anomalies: A new syndrome. *Am J Med Genet* 1990; 4: 386-390.
5. Oku T, Iwasaki K, Fujita H. Folded skin with an underlying cutaneous smooth muscle hamartoma. *Br J Dermatol* 1993; 5: 606-608.
6. Vora RV, Pilani AP, Diwan NG, et al. Miochelin tire baby syndrome. *Indian J Paediatr Dermatol* 2016; 17: 226-228.
7. Ebbert JO, Limper AH. Respiratory syncytial virus pneumonitis in immunocompromised adults: Clinical features and outcome. *Respiration* 2005; 72: 263-269.
8. Hall CB, Powell KR, MacDonald NE, et al. Respiratory syncytial viral infection in children with compromised immune function. *N Engl J Med* 1986; 315: 77-81.
9. Haghshenas Z, Tajziehchi L, Ghavami F. Association between Michelin tire baby syndrome and congenital panhypopituitarism in an Iranian girl. *Arch Iran Med* 2014; 17: 585-586.
10. Sardana K, Mendiratta V, Kakar N, et al. Spontaneously improving Michelin tire baby syndrome. *Pediatr Dermatol* 2003; 20: 150-152.

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