

The Third Case of the Extended Michelin Tire Baby Syndrome Associated With Undescended Testis, Mental Retardation and Hearing Impairment

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1. Abstract

Michelin tire baby syndrome is a very rare heterogeneous disorder, characterized by congenital generalized circumferential folding of excess skin occurring as an isolated abnormality or in association with various phenotypic abnormalities. About 30 cases of Michelin tire baby syndrome have been reported in the literature, including very few cases with the extended form of the syndrome which is associated with multiple congenital abnormalities and mental retardation [6] reported the second case of another type of the extended Michelin tire baby syndrome associated with significant non-cutaneous abnormalities and mental retardation associated with hearing impairment, and undescended testis. The aim of this paper is to report the third case of the extended Michelin tire baby syndrome associated with undescended testis, mental retardation associated and hearing impairment, which occurred in an Iraqi child with the novel occurrence of excessive gyration of brain tissues seen on ultrasonography.

2. Keywords: Michelin tire baby syndrome; Undescended testes

3. Introduction

Michelin tire baby syndrome is a very rare heterogeneous disorder, characterized by congenital generalized circumferential folding of excess skin occurring as an isolated abnormality or in association with various phenotypic abnormalities [1,3]. The skin abnormality was also called congenital diffuse lipomatosis as it was attributed to a diffuse lipomatous nevus in the deeper dermis [1,2].

Diagnosis of the condition is primarily clinical, and the skin folds may gradually diminish and disappear with age without any intervention. The skin abnormality of multiple folding of skin is symmetric and like circular skin creases, or bands occurring on the forearms, lower legs, and often the neck [1,2].

The condition was first described by Cyril M. Ross in 1969, and was named after the mascot "Michelin Man" (Figure-1) of Michelin, the French tire manufacturer [1]. About 30 cases of Michelin tire baby syndrome have been reported in the literature, including very few cases with the extended form of the syndrome which is associated with multiple congenital abnormalities and mental retardation [1-7].

The syndrome was reported from many countries including Italy (Patrizi and colleagues, 1989), United States (Bass and colleagues, 1993; Samarin and colleagues, 2010), Canada (Elliott and colleagues, 1996; Leonard (2002), Japan (Sato et al, 1997; Kondoh et al, 2004; Nomura and colleagues, 2010; Kato and colleagues, 2015), India (Sardana et al, 2003; Palit and Inamdar, 2007; Metta and colleagues (2012), Tunisia (Kharfi et al, 2005; Tinsa et al (2009), Turkey (Ulucan et al, 2013), Iran (Haghshenas and colleagues, 2014), and Pakistan (Uzair while and colleagues, 2015). However, it has not been reported in Iraq [1].

The aim of this paper is to report the third case of the extended Michelin tire baby syndrome associated with significant non-cutaneous abnormalities undescended testis, mental retardation and hearing impairment, which occurred in an Iraqi child with the novel occurrence of excessive gyration of brain tissues seen on ultrasonography.

4. Case Report

An eighteen-month old boy, born from related parents (first degree parental consanguinity) was seen because of poor hearing and markedly delayed development with psychomotor

retardation. The boy was born at term after an uncomplicated pregnancy. His birth weight was not recorded, but most probably was above 2 Kilograms. At birth, there were multiple skin folds on the upper, lower, neck and genitalia.

He has not shown social smile.

The boy was not sitting or crawling.

He was not responding to name nor looking at faces.

He had poor feeding and growth failure and his weight at the age of eighteen months was 5 kilograms.

He had characteristic craniofacial dysmorphic features including:

1. Narrow palpebral fissures (Figure-2A).
2. Low set ears with over folded thick helices.
3. Upslanting palpebral fissures (Figure-2A).
4. Protuding tongue (Figure-2A).

The congenital multiple skin folds of Michelin tire baby syndrome on the upper limbs (Figure-2B), lower limbs (Figure-2C), neck, and genitalia were still present (Figure-2C). The boy also had undescended testes and abdominal ultrasound showed that both testes were normal in texture and vascularity, but they were ectopic and located in the lower abdomen. The size of the right testis was (1 X 7.5) centimeters, while the size of the left testis was (1 X 7) centimeters. Brain ultrasound showed excessive gyration of brain tissues. Chromosomal analysis showed a normal male karyotype with 46, XY.

Table-1 summarized the findings in the third case of the extended Michelin tire syndrome associated with significant non-cutaneous abnormalities and mental retardation associated with hearing impairment, undescended testis

Table 1: The findings in the third case of the extended Michelin tire syndrome associated with significant non-cutaneous abnormalities and mental retardation associated with hearing impairment, undescended testis.

Sporadic
First degree parental consanguinity
Congenital multiple skin folds on the upper, lower, neck and genitalia
Poor feeding
Growth retardation
Hearing impairment
Psychomotor retardation
Dysmorphic facial features
Narrow palpebral fissures
Low set ears with overfolded thick helices
Upslanting palpebral fissures
Protruding tongue
Undescended testes
Excessive gyration of brain tissue



Figure 1: A sketch the mascot “Michelin Man” of Michelin ,the French tire manufacturer.



Figure 2: The third case of the extended Michelin tire baby syndrome associated with significant non-cutaneous abnormalities undescended testis, mental retardation and hearing impairment, which occurred in an Iraqi child.

5. Discussion

The extended Michelin tire baby syndrome associated with significant non-cutaneous abnormalities and mental retardation is extremely rare and was reported by very few authors including [3-6], The Elliott type of the extended Michelin tire baby syndrome associated with significant non-cutaneous abnormalities and mental retardation is associated with aural and ocular abnormalities including microphthalmia. [5] From Canada reported the second patient with this type, while [7] most probably reported third case.

[6] Reported the second case of another type of the extended Michelin tire baby syndrome associated with significant non-cutaneous abnormalities and mental retardation associated with hearing impairment, undescended testis [6], and in this paper the third case of this is described.

Reference

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