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Case Report

A rare case of Kunze-Riehm syndrome in a neonate

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ABSTRACT

The Kunze-Riehm syndrome also called as Michelin tire baby syndrome (MTBS), is a rare genodermatosis, characterized by multiple symmetric circumferential folding of excess skin with the various phenotypic abnormality. The diagnosis is made on the basis of the characteristic clinical features in the literature there are approximately 31 cases reported and to the best of our knowledge, this would be the sixth case published from India. Herein authors report a rare case of Kunze- Riehm syndrome in a neonate.

Keywords: Circumferential, Clinical diagnosis, Genodermatosis, Kunze-Riehm syndrome, Michelin tire baby syndrome, Phenotype

INTRODUCTION

Kunze-Riehm syndrome is characterized by generalized folding of excess skin although it may be associated with additional phenotypic abnormalities. The term Michelin tire baby (MTB) derives from the cartoon mascot of the Michelin tire company and refers to the presence of multiple symmetric circumferential rings of folded skin described by Ross in 1969.[1]

CASE REPORT

A 39-week term male child was delivered vaginally with a birth weight of 2.9kg. He was born to a consanguineous union after the infertility treatment. The antenatal scans in the third trimester showed oligohydramnios (AFI=4), with fetal MRI showing corpus callosum agenesis with bilateral severe lateral ventriculomegaly. He cried immediately after birth. Anthropometry, revealed, weight of 2.9kg, length=50cm ,HC=33cm. He was born with short neck with circumferential skin folds in the neck, symmetrically in all the limbs (Figure 1), abnormal genitalia- anteriorly placed scrotum with unilateral undescended testis and penile shaft in the posterior part with hypospadias (Figure 2).

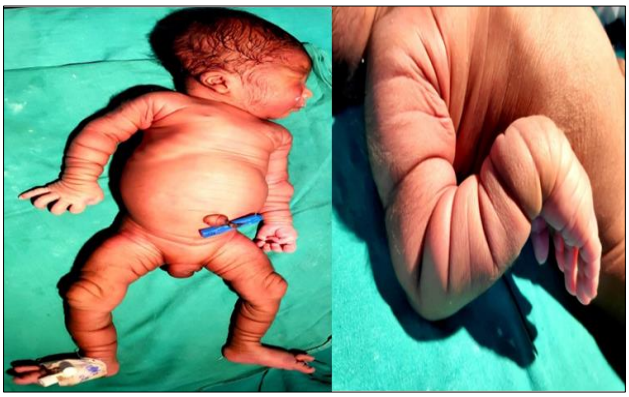

Figure 1: Symmetrical circumferential skin folds in the neck and body.

He was dysmorphic with brachycephaly, flat face, retrognathia, low set ears, thick helix, hypertelorism, periorbital fullness, blepharophimosis with Short

palpebral fissures, upslanting palpebral fissure, broad and depressed nasal bridge, microstomia, posterior cleft palate (Figure 3).

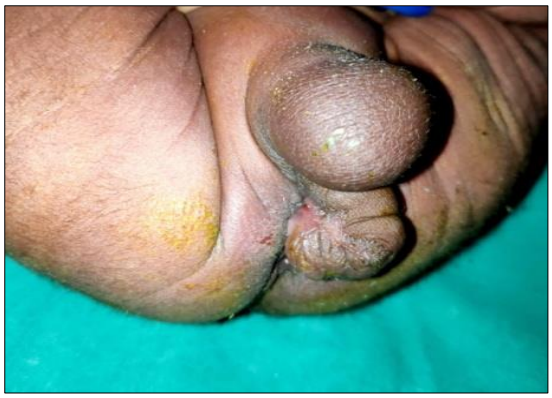

Figure 2: Abnormal genitalia with hypospadiasis.

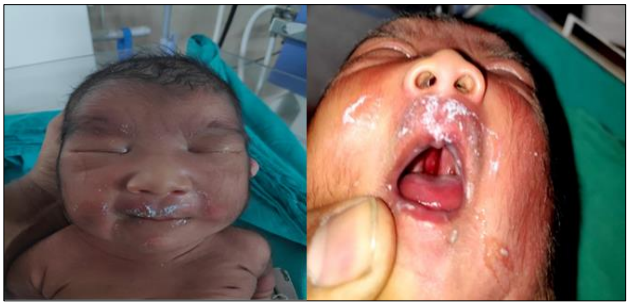

Figure 3: Facial features and posterior cleft palate.

DISCUSSION

The syndrome may be associated with various phenotypic abnormalities. The skin is thrown into folds and is mainly confined to the extremities. The most common site of involvement is extremities and it can also involve trunk, palms, and soles. The pathogenesis is yet unclear. The condition may be familiar. The autosomal dominant mode of inheritance has been reported in familial cases.Paracentric inversions of the long arm of chromosome 11and deletion of the short arm of chromosome 7 has been reported in association with this syndrome.[2,3]

The associated congenital anomalies are craniofacial anomalies, cleft palate, hypoplastic scrotum and hernias (inguinal and umbilical), left-sided hemihypertrophy, hemiplegia and microcephaly, psychomotor retardation, epilepsy, joint hypermobility, stellate scarring, developmental delay, and smooth muscle hamartoma which may be diffuse.

Diagnosis is made on clinical signs of this syndrome is mainly clinical, and the exact pathogenesis is unknown.[4] The nevus lipomatous and smooth muscle hamartoma are the main histological findings seen in this condition.[5]

This condition may be associated with other disorders such as Beare–Stevenson cutis gyrate syndrome associated with dermatomegaly localized to scalp, forehead, face, and neck, hearing impairment, undescended testis, circumferential skin creases, and mental handicap syndrome, and multiple congenital anomalies/mental retardation syndrome. The prognosis is dependent upon the associated clinical condition. The skin folds gradually disappear without any intervention.[6]

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