

## Midface toddler excoriation syndrome



**Figure 1:** Lichenified plaque on the bridge of the nose along with excoriation marks and atrophic scarring of the midface

A 7-year-old boy born of non-consanguineous parentage, presented with itchy lesions on his face, manifesting since the age of 11 months. His parents reported repeated scratching of the nose and the surrounding areas. Besides, they noticed sleep disturbances as well. The developmental milestones were not delayed. Similar lesions were not found elsewhere in the body. There was no loss of sensations or any neurological deficit; no history of seizures, or photosensitivity. Family history was non-contributory. Cutaneous examination showed bilaterally symmetrical lichenified lesions over the bridge of the nose and adjacent areas, along with multiple excoriations on the central portion of the mid-face [Figure 1]. The child was found to be constantly scratching and pulling the nose, even during clinical examination. Routine laboratory investigations including complete hemogram, uric acid, renal profile (urea and creatinine) and liver enzymes were within normal limits. Based on the

**How to cite this article:** Kumar P, Das A. Midface toddler excoriation syndrome. *Indian J Dermatol Venereol Leprol* 2021;87:528-9.

Received: July, 2019 Accepted: October, 2019 Published: June, 2021

**DOI:** 10.4103/ijdv.IJDVL\_323\_19 **PMID:** 31975696

This is an open-access article distributed under the terms of the Creative Commons Attribution-Non Commercial-Share Alike 4.0 License, which allows others to remix, tweak, and build upon the work non-commercially, as long as the author is credited and the new creations are licensed under the identical terms.

history, examination findings, and investigations, a diagnosis of midface toddler excoriation syndrome (MiTES) was made. Biopsy for histopathology and genetic testing could not be performed. The child was prescribed emollients and topical steroid–antibiotic combination, and moderate improvement in the clinical appearance was noted.

Dermatitis artefacta and neurotic excoriations were considered as clinical differentials. However, onset since infancy, constant scratching of the central face, and characteristic localization of lesions over mid-face favoured the clinical diagnosis of MiTES. In addition, psychological evaluation of the child and parents was non-contributory. Another important differential is Lesch–Nyhan syndrome, which is characterized by overproduction of uric acid, dystonia, mental retardation, aggressive and impulsive behaviours, and self-mutilating activities.

MiTES is a recently described entity linked to *PRDM12* gene mutation and is clinically characterized by infantile onset excoriations and sequelae of long-term scratching, present over mid-face, with a predilection for the nasal bridge. Bi-allelic mutations in *PRDM12* gene cause hereditary sensory and autonomic neuropathy type VIII (HSAN 8), a congenital pain insensitivity disorder, manifesting as ulceration of the digits, lips, tongue, as well as facial scratching. Since, MiTES is a genetically heterogeneous entity, genes other than *PRDM12* can also be involved. MiTES resembles HSAN 8 clinically and etiologically but lacks ulceration of the acral parts.

#### **Declaration of patient consent**

The authors certify that they have obtained all appropriate patient consent forms. In the form, the legal guardian has given his consent for images and other clinical information to be reported in the journal. The guardian understands that names and initials will not be published and due efforts will be made to conceal identity, but anonymity cannot be guaranteed.

#### **Financial support and sponsorship**

Nil.

#### **Conflicts of interest**

There are no conflicts of interest.

*Piyush Kumar, Anupam Das<sup>1</sup>*

Department of Dermatology, Katihar Medical College and Hospital, Katihar, Bihar, <sup>1</sup>Department of Dermatology, KPC Medical College and Hospital, Kolkata, West Bengal, India

#### **Corresponding author:**

Dr. Anupam Das,  
Building “Prerana” 19 Phoolbagan, Kolkata - 700 086, West Bengal, India.  
anupamdasdr@gmail.com