Anaesthetic management in midface toddler excoriation syndrome (MiTES)- A case report

Dear Editor,

Midface toddler excoriation syndrome (MiTES) is a novel disorder reported in toddlers, characterised by self-inflicted, habitual scratching over the glabellar area, causing severe scarring and disfigurement, associated with a positive regulatory domain-containing protein 12(*PRDM12*) gene mutation, which is involved in the development of neurons that aid in pain sensation and perception.[1]

A three-year-old male child weighing 11 kg visited the otorhinolaryngology clinic with complaints of persistent nasal discharge for three months and was diagnosed with a rhinolith in the left nostril. The child was posted electively for diagnostic nasal endoscopy and rhinolith removal. He was regularly followed up by the neurology team for seizure disorder, developmental delay, and intellectual disability. At two years of age, he was clinically diagnosed with MiTES [Figure 1] and was on treatment with oral risperidone 0.25mg and olanzapine 2.5mg once daily. On admission, samples for genetic testing were sent for confirmation. His cardiac and respiratory status was normal. After a pre-anaesthetic examination, informed consent was taken from the parents and the child was fasted as per standard guidelines. The intravenous (IV) line was secured on the day before surgery by using a patch with a eutectic mixture of local anaesthetics 45 min before the IV cannulation. Upon arrival, the child was pre-medicated with 0.05mg/kg of IV midazolam and shifted to the operation theatre. Anaesthesia was induced with IV ketamine 1mg/kg and propofol 2mg/kg, and the trachea was intubated with a 4.5 mm microcuffed endotracheal tube after adequate neuromuscular blockade with 0.5mg/kg of atracurium. Anaesthesia was maintained with isoflurane with a minimum alveolar concentration of 1.5 and intermittent IV atracurium 0.1mg/kg. The foreign body found in the nasal cavity was removed, and haemostasis was achieved. The child was haemodynamically stable throughout the procedure and extubated after administering IV neostigmine 0.05mg/kg and glycopyrrolate 0.005mg/kg at the end of the procedure for 60 min. A prophylactic dose of IV dexamethasone 0.1mg/kg was administered during induction, and IV ondansetron 0.1mg/kg was administered before extubation to block the histamine receptors. The post-operative period was uneventful.

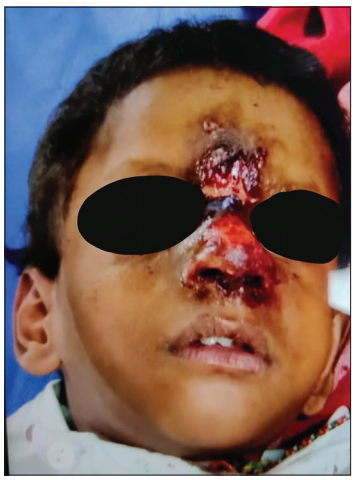

Figure 1: Image of the child taken at the time of diagnosis of MiTES, with severe mid-facial lesion caused due to excessive scratching of the face

MiTES is a newly described pain disorder with heterogenous genetic mechanisms, including an autosomal recessive type caused by biallelic PRDM12 polyalanine tract expansion and an autosomal dominant type due to heterozygous mutations in sodium voltage-gated channel alpha subunit 11. It resembles hereditary sensory and autonomic neuropathies (HSAN) type VIII, wherein biallelic mutation in the *PRDM12* gene is seen, with the afflicted individual often suffering from ulceration of the distal digits and facial scratching. Light touch, vibration, and proprioception sensations carried via the large sensory fibres remain unaffected. However, patients with HSAN type VIII can have reduced sweating and tearing with autonomic dysfunction.[2] Naturally occurring mutations of the *PRDM12* gene cause congenital insensitivity to pain and HSAN.[3]

The clinical profile of the child matched with the existing literature report.[1] However, seizures and developmental delays are not associated with MiTES syndrome. We focused on the safe use of anaesthetic drugs with minimal influence on pain and pruritus pathways and the possible dysregulated autonomic pathway. The use of opioids leads to pruritus, most noticeably in the areas of trigeminal nerve distribution, mainly mediated by subnucleus caudalis in the trigeminal ganglion. In addition, due to the loss of peripheral sensory nerve fibres, opioids are not required in cases of congenital insensitivity to pain.[4] MiTES is

a localised area of intractable itching associated with pain insensitivity in the same region. Thus, pain from other sources, such as tracheal intubation, positioning, and suctioning, needed to be managed without opioids, which could worsen the clinical situation. Autonomic dysfunction manifesting as impaired coordination in swallowing, gastroparesis, reduced lower oesophageal sphincter tone, bradycardia, postural hypotension, orthostatic hypotension, and hyperpyrexia are concerns that must be addressed.[5]

Declaration of patient consent

The authors certify that they have obtained all appropriate consent forms. In the form, the child's parents consented to images and other clinical information to be reported in the journal. The parents understand that the name and initials will not be published and due efforts will be made to conceal his identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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REFERENCES

1. Srinivas SM, Gowda VK, Owen CM, Moss C, Hiremagalore R. Mid-face toddler excoriation syndrome (MiTES): A new paediatric diagnosis. Clin Exp Dermatol 2017;42:68-71.
2. Elhennawy K, Reda S, Finke C, Graul Neumann L, Jost Brinkmann PG, Bartzela T. Oral manifestations, dental management, and a rare homozygous mutation of the PRDM12 gene in a boy with hereditary sensory and autonomic neuropathy type VIII: A case report and review of the literature. J Med Case Rep 2017;11:1-1.
3. Rienzo M, Di Zazzo E, Casamassimi A, Gazzerro P, Perini G, Bifulco M, *et al*. PRDM12 in health and diseases. Int J Mol Sci 2021;22:12030.
4. Qiu Y, Zhao L, Yao D, Jia Y. Anesthetic management of children with congenital insensitivity to pain with anhidrosis. Pediatr Investig 2019;4:296-8.
5. Dave N, Sonawane A, Chandolkar S. Hereditary sensory autonomic neuropathy and anaesthesia-A case report. Indian J Anaesth 2007;51:528-30.

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