Anesthetic considerations of CHILD syndrome

Dear Editor,

CHILD syndrome is a rare, X-linked dominant disorder seen almost exclusively in females.[1] Acronym of the CHILD syndrome consists of congenital hemidysplasia, ichthyosiform erythroderma, and limb defects. Systemic involvement may occasionally occur, typically on the same side of the body where skin and limb abnormalities are present. There is a lack of literature regarding the anesthetic management of patients with this condition. Written informed consent was obtained from the child's guardians to publish this report.

A 13-year-old female child (18 kg, 117 cm) presented for syndactyly release and split-thickness skin grafting of both the hands and donor graft site as the thigh. She had dry, scaly, erythematous patches predominantly on the left side of her body with a few patches present on the right side. The index patient had a few alopecic patches on the head with nail dystrophy of all limbs Figure 1a. No organ system involvement was apparent on history and physical examination. Airway examination revealed a high arched palate. Laboratory investigations were within normal limits. In view of the possible heart and other abdominal organ defects, we obtained an echocardiogram and abdominal ultrasound, which were normal. She was being treated with oral acitretin, a retinoid used for the management of skin lesions. Additionally, she

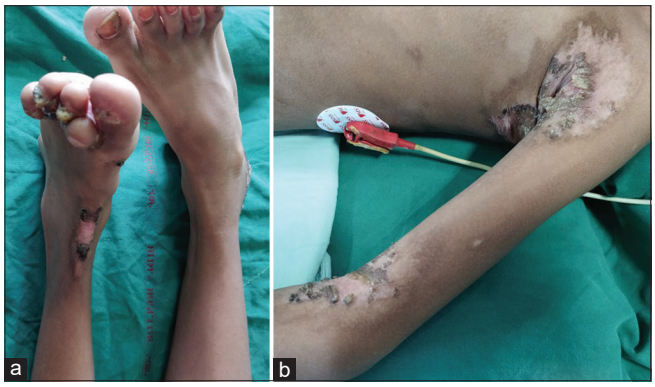

Figure 1: (a) Nail dystrophy of all limbs, (b) Widespread skin involvement of axilla

was on growth hormone therapy two years previously for 6 months for evaluation of short stature.

In the operating room, standard American Society of Anesthesiologists monitors were attached. Vital parameters were within the normal range (heart rate 80/min, non-invasive blood pressure 88/48 mm Hg, spO_2 100% on room air). Anesthesia was induced with fentanyl 35 μg, propofol 50 mg, and atracurium 10 mg, following which an AMBU Aura 40 (Ambu Inc. MD, USA) size 2 supraglottic device was inserted. A caudal block was administered with 0.2% ropivacaine 9 ml and 900 μg morphine. Anesthesia was maintained with isoflurane in O_2 and N_2O (50:50), using pressure control ventilation. Intraoperative glucose monitoring showed capillary glucose as 99 mg/dl. The rest of the postoperative course was uneventful.

CHILD syndrome occurs due to a faulty gene on the long arm of the X chromosome (Xq28), which normally encodes a protein (NSDHL) responsible for an essential step in cholesterol biosynthesis.[1-3] It is a very rare disorder with a prevalence of $<1/1,000,000$, with less than 60 cases reported to date; none reporting anesthetic concerns in these patients.[1,4]

There are various anesthetic concerns of this syndrome that need to be taken care of. Dystrophic nail changes may interfere with placement and accurate signal acquisition by the pulse oximetry probe. Hence, it is preferable to use Masimo SET® pulse oximetry. Widespread skin involvement can also make the performance of regional nerve blocks impossible. We were unable to give a brachial plexus block for this very reason [Figure 1b]. Systemic opioids and paracetamol were used to provide analgesia for the operated hand. The perianal region and natal cleft were similarly involved, but the sacral region was spared enabling us to use a caudal block for analgesia for the skin graft site. Intravenous catheter placement may also be difficult due to skin involvement. The erythematous patches also need adequate protection intraoperatively to prevent chafing. Pruritus after the use of epidural morphine was also a concern. However, weighing the benefits of a regional anesthetic technique versus opioid-induced pruritus, we decided to administer morphine in combination with a low concentration of local anesthetic. The combination of the two is associated with a low incidence of pruritus, and we administered ondansetron, which has demonstrated efficacy in preventing opioid-induced pruritus.[5]

Growth hormone therapy itself may lead to insulin resistance causing hyperglycemia and dyslipidemia with its associated problems, hence glucose monitoring was done.

Limb defects in these patients may range from shortened metacarpals to the absence of the entire limb. In severe cases, the absence of ribs and vertebrae may lead to scoliosis. Cardiac involvement is common in left-sided cases, which in most instances leads to prenatal death.[4] Congenital hemidysplasia with ichthyosiform erythroderma and limb defects syndrome, also known as CHILD syndrome, is a rare condition that affects different parts of the body. It has been described as an X-linked, dominant condition with a male-lethal trait with most surviving patients being females. It is related to mutations in the NSDHL gene. This condition should be suspected at birth if a child presents with a unilateral epidermal nevus. Other commonly seen features include unilateral limb and skin, unilateral ichthyosiform erythroderma, inflammatory variable epidermal nevus, and congenital heart disease.[1,4] Lung hypoplasia, renal agenesis, unilateral central nervous system (CNS) defects, lissencephaly, cerebellar malformations, hypoplasia of the thyroid, and adrenal glands are also seen necessitating a thorough physical examination and focused investigations.

Hence, skin and nail involvement results in difficult intravenous access, peripheral nerve blocks, and pulse oximetry probe positioning challenging. The highly arched palate may intensify difficulty in mask ventilation and laryngoscopy. Possibility of systemic involvement necessitates intense vigilance and multidisciplinary involvement early for successful anesthetic management of these cases.

Declaration of patient consent

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

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

Ameya Pappu, Manpreet Kaur, Joel J. Gnanadhas, Rakesh Dawar[1], Ajay Singh

Departments of Anaesthesiology, Pain Medicine and Critical Care and [1]Plastic, Burn and Reconstructive Surgery, All India Institute of Medical Sciences, New Delhi, India

Address for correspondence: Dr. Manpreet Kaur, Department of Anaesthesiology, Pain Medicine and Critical Care, Burn and Reconstructive Surgery, All India Institute of Medical Sciences, New Delhi - 110 029, India.
E-mail: manpreetkaurrajpal@yahoo.com

References

1. Ramphul K, Kota V, Mejias SG. Child Syndrome. In: StatPearls [Internet]. Treasure Island (FL): StatPearls Publishing; 2020. Available from: http://www.ncbi.nlm.nih.gov/books/NBK507813/.[Last cited on 2020 Feb 25].
2. Bergqvist C, Abdallah B, Hasbani D-J, Abbas O, Kibbi AG, Hamie L, *et al*. CHILD syndrome: A modified pathogenesis-targeted therapeutic approach. Am J Med Genet A 2018;176:733-8.
3. Mi XB, Luo MX, Guo LL, Zhang TD, Qiu XW. Child syndrome: Case report of a Chinese patient and literature review of the NAD[P] H steroid dehydrogenase-like protein gene mutation. [Internet]. Available from: https://www.ncbi.nlm.nih.gov/pubmed/26459993. [Last cited on 2020 Feb 25].
4. Ichthyosis, CHILD Syndrome - NORD (National Organization for Rare Disorders) [Internet]. Available from: https://rarediseases.org/rare-diseases/ichthyosis-child-syndrome/. [Last cited on 2020 Feb 25].
5. Kumar K, Singh SI. Neuraxial opioid-induced pruritus: An update. J Anaesthesiol Clin Pharmacol 2013;29:303-7.

Access this article online

Quick Response Code:

Website: www.joacp.org

DOI: 10.4103/joacp.JOACP_425_19

How to cite this article: Pappu A, Kaur M, Gnanadhas JJ, Dawar R, Singh A. Anesthetic considerations of CHILD syndrome. J Anaesthesiol Clin Pharmacol 2021;37:130-2.

Submitted: 19-Dec-2019 **Revised:** 28-Jan-2020
Accepted: 12-Mar-2020 **Published:** 10-Apr-2021