
Anaesthetic management of a rare case of Langerhans cell histiocytosis

Sir,

Langerhans cell histiocytosis (LCH) or histiocytosis X is an uncommon haematological disorder affecting infants and young children. This uncontrolled proliferation of white blood cells has gone by several names, including Hand-Schüller-Christian disease, Abt-Letterer-Siwe disease, Hashimoto-Pritzker disease and Histiocytosis X.^[1] Patients with LCH present challenges to the anaesthesiologist as a consequence of potential hypothalamic pituitary axis (HPA), renal,

hepatic, lung and bone marrow involvement along with cutaneous manifestations inside oral cavity and overall body.^[2] A case of LCH with severe cutaneous manifestations in a patient as young as 10 months of age has not been reported.

A 10-month-old male baby weighing 6.5 kg with normal developmental milestones and a known case of LCH since birth was posted for Hickman catheter insertion under general anaesthesia. Multiple crusted papules with scaling of papular xanthomas were present on the body and notably over the cheek and perioral area [Figure 1]. Airway examination revealed ankyloglossia. However, there was no lung involvement as per available investigations. The results of all laboratory investigations including renal function test, liver function test and chest X-ray (PA view)



Figure 1: Multiple crusted papules along with scaling of papular xanthomas present on the body

were within normal limits for his age. The infant with parental cooperation was transferred undisturbed and gently to operation table and inhalational induction was carried out with sevoflurane 6%–8% through facemask prelubricated with liquid paraffin. Care was taken to prevent injury to skin lesions with moistened gauzes swab. In view of extensive skin lesions, intravenous access though difficult was secured in the left hand. Pulse oximetry was applied on and off over different fingers, and blood pressure cuff was applied over the left thigh which was with minimal lesion to prevent injury from friction. The adhesive portion of electrocardiogram electrode was removed, leaving only the electroconductive gel portion which was affixed over normal areas of skin. Minimal pressure was applied during head tilt and chin lift for manual ventilation. Atracurium 3.0 mg was given intravenously to enable tracheal intubation with a size 3 mm uncuffed tube using the Macintosh size 1 blade laryngoscope, which was lubricated with water-based jelly to prevent trauma. Anaesthesia was maintained with sevoflurane, oxygen and nitrous oxide. Intraoperative course was smooth and the patient was haemodynamically stable. The extubation of trachea was uneventful. Eighty millilitres of crystalloid was infused over a period of 40 min. Postoperative analgesia was provided with intravenous paracetamol. Postoperative period was uneventful. The patient was discharged from the hospital on the next day.

Patients with LCH might have osteolytic lesions that can predispose to pathological fractures. A gentle transfer to operating room can avoid tantrums and prevent injuries.^[2] Literature suggests that laryngoscopy might lead to intraoral bleeding.^[3] In this case, we performed gentle laryngoscopy with well-lubricated blades. The

skin lesions in patients with LCH can vary from scaly erythematous lesions to red papules with extensive eruptions on the scalp in 80% of children. Changing the site of pulse oximeter can avoid pressure erosion over fingers. Noninvasive pressure cuff should be applied over the lesion-free area or over the cotton pad to avoid skin pressure injury. Haemoglobin should be optimised as bone marrow infiltration causes anaemia. Hepatomegaly is present in one-fourth of cases which can affect drug metabolism. Blood sugar level and urine output have to be closely monitored as HPA involvement leads to diabetes insipidus. Chest X-ray in the preoperative period can reveal possible lung nodules.

Declaration of patient consent

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

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Conflicts of interest

There are no conflicts of interest.

**Shalendra Singh, Shreyas Kate, Rashmi Datta¹,
Virender Suhag²**

Department of Anaesthesiology and Critical Care, Armed Forces Medical College, ²Department of Radiation Oncology, Command Hospital (Southern Command), Pune, Maharashtra, ³Department of Anaesthesiology and Critical Care, Command Hospital (Western Command), Chandigarh, India

Address for correspondence:

Dr. Shalendra Singh,
Department of Anaesthesiologist and Critical Care, Armed Forces Medical College, Pune - 411 040, Maharashtra, India.
E-mail: drsinghafmc@gmail.com

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