Anaesthetic management of a child with Farber's lipogranulomatosis posted for exploratory laparotomy

Sir,

Farber's lipogranulomatosis is a rare (<1:1,000,000) genetic metabolic disorder with autosomal recessive inheritance from mutation of N-acylsphingosine amidohydrolase 1 (*ASAH1*) gene in the lysosomes, resulting in deposition of ceramide in various tissues of the body.[1] This multisystem disease involves the neurons, myocardium, liver, spleen, lymph nodes, alveoli, pleural lining of the lungs, and the synovial lining of body joints.[2] Therefore, it is important to carefully examine and assess the functioning of all the involved organ systems before administering anaesthesia and be prepared for the anticipated complications.

An 8–year-old girl with a known case of Farber's lipogranulomatosis on regular medical treatment presented with abdominal pain since 2 days. She was diagnosed with acute intestinal obstruction requiring emergency exploratory laparotomy. The patient was diagnosed with Farber's lipogranulomatosis at 18 months of age, on oral steroid treatment and became progressively confined to bed over the last 3–4 years. The child was malnourished with psychomotor retardation and fixed flexion deformity of various body joints of the body [Figures 1 and 2]. There were multiple subcutaneous nodules present all over her body. On airway examination mouth opening was 2 cm with large tongue with only part of soft palate and hard palate visible. The upper lip bite test was grade 1 with no flexion or extension movement at the neck. On spine examination, lumbar scoliosis was noted. Her preoperative vitals were as follows: temperature - 101°C, pulse rate - 130/min, blood pressure - 100/60 mmHg, and SpO_2- 90% on room air with respiratory rate - 34/min. On chest auscultation the air entry was significantly decreased in the bases. Blood investigations revealed haemoglobin - 8.2 g%, total leucocyte count - 3,200/cm^3, platelet count - 1.6 lakhs/cm^3. Arterial blood gas (ABG) analysis reported pH - 7.52, pO_2 -59 mmHg, pCO_2-24 mmHg, HCO_3-17 mmol/L. The liver enzymes were mildly raised with hypoproteinaemia (serum albumin - 2.2 g/dL). Electrocardiogram showed sinus tachycardia and on chest x-ray fibrotic changes with calcification in bilateral lung fields were noted. Patient was taken up for surgery under American Society of Anesthesiologist's physical status III/E and the parents were prognosticated about the possibility of the need for postoperative mechanical ventilation. The child was wheeled inside the operation theatre and electrocardiography, pulse oximetry and non-invasive blood pressure were applied. Paediatric difficult airway cart was kept standby. From 22 G intravenous cannula *in situ* ringer lactate infusion was started. Nasogastric tube *(in situ)* was aspirated. The child was premedicated with Inj. Fentanyl 2 mcg/kg intravenously (IV). Preoxygenation was done with 100% O_2 for 3 min. Modified rapid sequence induction was done with Inj. propofol 1 mg/kg and Inj. succinylcholine 2 mg/kg intravenously. The child was intubated using McGrath videolaryngoscope with cuffed endotracheal tube (ETT) of 5 mm ID. Thereafter, the trachea was inspected using flexible bronchoscope for any evidence of trauma or lipogranulomas and few lipogranulomas were found embedded in the tracheal wall with no sign of injury to the tracheal wall. The ETT cuff pressure was maintained around 20 cm H_2O. Inj. dexamethasone 0.1 mg/kg and Inj. paracetamol 15 mg/kg was administered intravenously. Adequate padding of pressure points was ascertained and the

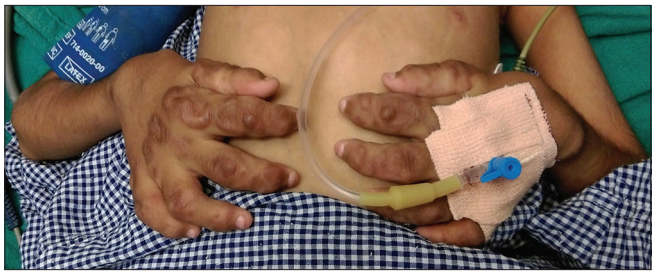

Figure 1: Multiple lipogranulomas with fixed flexion deformity in both hand

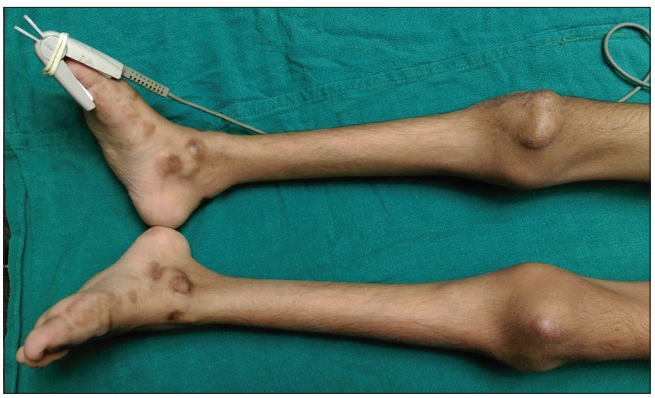

Figure 2: Joint swelling with fixed flexion deformity of lower limbs

body temperature was monitored. Adequate depth of anaesthesia was maintained with sevoflurane (1--2%) with 100% O_2 and muscle relaxation was achieved with injection atracurium under continuous neuromuscular blockade monitoring. Resection anastomosis of small intestine was performed lasting 2.5 h mounting to a blood loss of 200 ml which was replaced with packed red blood cells (PRBCs). Intraoperative ABG showed pH - 7.38, pO_2 - 259 mmHg, pCO_2 - 28 mmHg, HCO_3 - 19 mmol/L.

After the completion of surgery, bilateral transversus abdominis plane block and rectus sheath block were performed with Inj. bupivacaine 0.125% (30 ml). Subsequently, the inhalational anaesthetic was stopped; the neuromuscular blockade was assessed and reversed with Inj. neostigmine 50 mcg/kg and Inj. glycopyrolate 10 mcg/kg intravenously. The trachea was extubated after return of airway reflexes. Postoperatively the vitals were stable and the patient was pain free with no signs of respiratory distress.

The three characteristic signs of Farber disease include hoarseness of voice, small nodules under the skin (lipogranulomas), and painful swollen joints.[2] These patients are usually on long-term steroid therapy and intraoperative steroid supplementation should be provided as per the protocol to help tide over surgical stress and to aid in the medical management of airway edema following inadvertent trauma during airway manipulation.[3] One should also rule out the possible systemic side effects of long-term steroid therapy and to address them judiciously. These children usually have some degree of psychomotor retardation and require patient, unhurried approach while communicating with them.[2]

These patients present with anticipated difficult airway as the disease associated arthropathy may involve the cervical spine and the temporomandibular joint resulting in restricted mouth opening with limited range of movements at the neck. Also multiple growths (lipogranulomas) along the airway tract may present with upper airway obstruction, obscure the laryngeal view, bleed or may get dislodged during traumatic intubation and present as foreign body in the trachea which may worsen ventilation and can precipitate any catastrophic event during airway management.[4] Therefore, it is advised to avoid endotracheal intubation in these patients.[5] However, our patient was considered to have a full stomach; most likely with sepsis with the possibility of need for postoperative mechanical ventilation. We therefore planned to intubate the trachea under videolaryngoscope guidance to circumvent the anticipated difficulty during laryngoscopy because of limited mouth opening and neck movement. However, it is advised that an experienced anaesthesiologist who is routinely performing videolaryngoscope-guided endotracheal intubation in paediatric patients should perform it to avoid increase in time to intubation. Also a size smaller ETT helped to avoid any undue airway trauma. The continuous ETT cuff pressure monitoring helped in preventing the increased risk of airway morbidity following pressure induced injury to any lipogranuloma entrapped between the tracheal wall and the ETT cuff.

Hypoxaemia in ABG pointed toward the involvement of the respiratory system which mainly presents as restrictive lung disease.[6] The disease associated arthropathy involving the dorsal spine which may further restrict the chest wall movement during respiration. Also the abdominal compartmental syndrome due to acute intestinal obstruction would have pushed the diaphragm cephalic with resultant basal atelectasis of the lung and ventilation perfusion mismatch.

These patients may present with cardiac conduction defects and one must be vigilant and prepared for management of any cardiac arrhythmia.[4] It is safer to use shorter acting drugs which can safely be administered in presence of altered liver and kidney function.[5] Opioids should be used judiciously as there may be respiratory depression in the postoperative period. Central neuraxial blockade may present with technical difficulty due to associated joint arthropathy and the spread of drug may be unpredictable.[5] Adequate postoperative pain relief should be ensured to decreases the incidence of pulmonary complication. Also ensure adequate nutrition in the postoperative period for faster recovery.[6]

Successful anaesthetic management of these patients requires meticulous planning and multimodal approach outweighing the risks and benefits. There is no substitute for vigilant monitoring, however with more advanced devices, latest techniques of regional anaesthesia and use of newer shorter acting anaesthetic agents the morbidity can be reduced significantly.

Declaration of patient consent

The authors certify that they have obtained all appropriate consent forms from the parents of the patient. In the form, the parent/s has/have given consent for their child's images and other clinical

information to be reported in the journal. The parent/s understands that her name and initial will not be published and due efforts will be made to conceal her identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

Nitin Choudhary, Sonia Wadhawan, Rahil Singh, Poonam Bhadoria

Department of Anaesthesiology and Critical Care, Maulana Azad Medical College, New Delhi, India

Address for correspondence:
Dr. Nitin Choudhary,
Flat No.-F/2, Plot No.-853, Vaishali Sector-5, Ghaziabad - 201 010, Uttar Pradesh, India.
E-mail: drnitinchoudhary@yahoo.in

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