

Anaesthetic considerations in a child with Urbach Wiethe disease posted for removal of cheek swelling

Sir,

Urbach Wiethe disease or more commonly lipoid proteinosis (LP), is an autosomal recessive extremely rare disorder. So far, not more than 300 patients have been diagnosed with LP.^[1,2] The deposition of hyaline material in the mucosa of the arteriolar capillary wall and epithelial basement membrane causes thickening of the skin mucosa.^[3] This predisposes the patient to widespread scarring, even with minor trauma especially over the extensor surfaces of the extremities. Very little is known about the anaesthetic considerations in a patient with this disorder. Multisystem involvement and upper airway abnormalities in this syndrome mandates an adequate planning, preparation and a meticulous conduct to achieve favourable outcomes.

A 11-year-old male child weighing 22 kg presented for removal of an infected cystic swelling of the right cheek. On preanaesthesia examination, maculopapular rashes were observed on the exposed areas of the body [Figure 1]. He was a known case of lipoid proteinosis, diagnosed 10 years back and taking oral zinc sulphate and multivitamins. His mother was a known case of diabetes mellitus (DM) II, on oral hypoglycaemic medications. No history of consanguinity, seizures, visual disturbances or respiratory obstruction were reported. General Examination revealed profuse

hyperpigmented macules over the face and along the eyelid margins. Airway examination revealed woody and fissured tongue [Figure 2]. His voice was hoarse and weak. Routine haemogram, renal and liver functions were unremarkable. In operation theatre, the patient was premedicated with intravenous (IV) midazolam 1 mg and ondansetron 2 mg. Anaesthesia was induced with fentanyl 2 µg/kg and propofol 2 mg/kg. Muscle relaxation was achieved using suxamethonium 2 mg/kg. On direct laryngoscopy, vocal cords were normal but the glottic area was narrow. Hence trachea was intubated orally with 4.5 mm cuffed endotracheal tube (ETT). Anaesthesia was maintained with oxygen and sevoflurane to maintain a minimum alveolar concentration (MAC) value of nearly 1.8 to 2%. Inj atracurium 12 mg was given for neuromuscular blockade. Inj dexamethasone 4 mg was given to prevent airway edema post operatively. The course of surgery was uneventful. The patient was reversed with neostigmine 1.5 mg and glycopyrrolate 0.3 mg IV. The patient was extubated keeping in lateral position. Patient had an uneventful recovery and was then discharged on 3rd day.

Haider *et al.*^[4] reported a case of emergency caesarean section in which patient did not provide any history of her disease (LP) and later they faced difficulty during intubation. They had to use an ETT of size 5.5 while actually she was estimated for a size of ETT 7. As this was our concern too, we electively kept small sized ETTs as a part of anticipated difficult airway/narrow laryngeal opening. There is said to be an association with, and possible predisposition to, diabetes mellitus although there are no recent corroborative references.^[4] But here in our case, reference was made to the fact that the patient's mother was a known



Figure 1: Maculopapular rashes



Figure 2: Woody tongue

case of DM II. The reduced oropharyngeal secretions increases the problems of hoarseness and phonation which mitigates against the use of antisialagogue premedication. Hence glycopyrrolate was avoided in this case. To avoid any risk of bleeding because of the friable and inelastic tissue, laryngeal instrumentation and tracheal intubation should be as gentle as possible.^[5] These patients have reduced gag reflex hence care should be taken to prevent aspiration in the post-operative period.^[6] This mandates the need of lateral tilting at the time of extubation. Literature suggests that these patients have a propensity to develop seizures and therefore epileptogenic drugs are best avoided in these cases, so we induced this patient with propofol due to its anticonvulsant as well as antiemetic action and rapid, smooth onset compared to thiopentone.^[5,7] Although laudanose induced seizures are very uncommon in day care procedures, utmost care must be taken to avoid atracurium in these patients with a known history of epilepsy.

In conclusion, a thorough history for consanguinity and history of diabetes mellitus should be taken from parents. Also, acne-like facial lesions with hoarseness of voice and macroglossia should alert an anaesthesiologist. Difficult laryngoscopy because of macroglossia and difficult intubation due to upper airway narrowing should be anticipated. Anti-sialagogue premedication and epileptogenic drugs should be avoided.

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 parents have given consent for their child's images and other clinical information to be reported in the journal. The parents understand that his 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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