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A Rare Case of Landau-Kleffner Syndrome in Adults: Anesthetic Challenges

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Sir,

A patient diagnosed with Landau-Kleffner syndrome was admitted to a tertiary care hospital for an emergency exploratory laparotomy. The patient had ingested a large piece of meat and landed in acute intestinal obstruction. This letter presents a rare syndromic case to discuss the anaesthetic management and challenges faced due to the difficult airway anatomy, a communication barrier and a long-standing seizure disorder. Landau-Kleffner syndrome is a rare neurological disorder of early childhood characterised by epileptiform aphasia, auditory agnosia and electroencephalographic abnormalities[1].

A 43-year-old male, a known case of Landau-Kleffner syndrome was admitted to the emergency department with the chief complaint of pain abdomen associated with abdominal distension and non-passage of stools for 4 days. He was diagnosed with acute intestinal obstruction on a CT scan and was posted for emergency exploratory laparotomy under general anaesthesia. The history revealed that the patient had accidentally swallowed a large piece of meat (Figure 1). He had a history of seizure disorder since childhood with the last episode of seizure eighteen months ago. He had been taking sodium valproate and lacosamide since childhood. Neurological consultation was taken and he was advised to continue with the same treatment.

On general physical examination, the patient was found to be obese with a Body Mass Index (BMI) of 31.5kg/m^2, awake, not oriented to time, place and

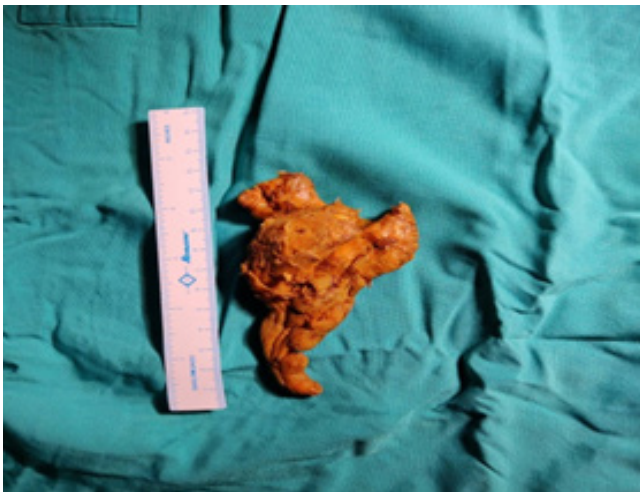

Figure 1. The large piece of meat removed during surgery.

Figure 2. Facial deformity in an adult patient with Landau-Kleffner syndrome.

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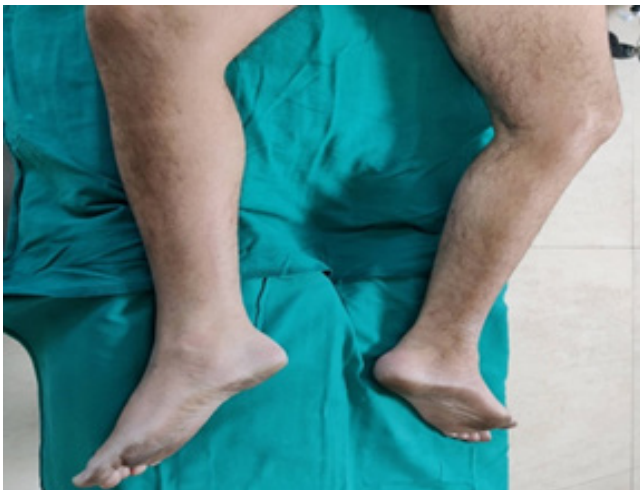

Figure 3. Muscle wasting seen in the bilateral lower limb.

person with very limited verbal comprehension. Facial deformities in the form of a broad nasal cavity, protruded mandible, short thyromental distance and a thick beard made the airway difficult. The airway examination could not be conducted properly as the patient was not following commands and was irritable (Figure 2). Muscle wasting was present in bilateral lower limbs (Figure 3).

Preoperative investigations included a normal haemogram, normal kidney function tests, liver function tests and 2D echocardiography. CT chest done outside suggested alveolar collapse and consolidation in bilateral lower lobes, ground glass appearance and haziness in both lungs along with mild right pleural effusion. It also reported an enlarged thymus. However, the actual Nucleic Acid Test (NAT) antigen test for COVID-19 was negative.

After taking written informed high-risk consent, the patient was shifted to the operating room and standard American Society of Anaesthesiologists (ASA) monitors were attached along with Bispectral (BIS) monitoring. RAMP position was made and the difficult airway cart were kept ready. Intravenous access was achieved with a 20G cannula. The patient was pre-oxygenated with 100% oxygen for 5 minutes and premedicated with injection fentanyl 100µg. A check video laryngoscopy with a sedation dose of injection propofol was done. As the medical team was able to appreciate the true cords, anaesthesia was induced with 2mg/kg iv propofol. After achieving complete neuromuscular blockade with an injection of atracurium 0.5mg/kg, the trachea was intubated with an 8mm endotracheal tube. Another 16-gauge intravenous cannula was also secured thereafter.

Anaesthesia was maintained according to the BIS value between 50 ± 5 with total intravenous anaesthesia with propofol infusion and oxygen/air mixture with a fresh gas flow of 1-2 L/min. Neuromuscular block was maintained with 0.1 mg/kg of atracurium to have zero twitch on the Train of Four (TOF) count monitoring, which was applied post-induction. Additional fentanyl boluses (0.5–0.75 µg/kg) were given to maintain analgesia. Continuous monitoring of Heart Rate (HR), Non-Invasive Blood Pressure (NIBP), SpO_2, $ETCO_2$, BIS and TOF was done. Propofol infusion was titrated to maintain a BIS value of 60–70 during the last 15 minutes of the surgery and an injection of ondansetron (0.1 mg/kg) was given. Anaesthesia was discontinued after the last stitch was placed. Neuromuscular blockade was reversed with neostigmine bromide (0.05 mg/kg) and glycopyrrolate (0.08 mg/kg) when a TOF ratio of 0.7 was reached. The trachea was extubated on return of spontaneous breathing with a minimum tidal volume of 6–8 ml/kg. The patient was shifted to the postoperative recovery room once awake and breathing spontaneously.

Landau-Kleffner syndrome also known as acquired epileptic aphasia, usually begins in childhood at the age of 2-8 years affecting mainly male children. The patient develops an inability to understand the spoken language along with affected speaking ability. Temper outbursts and aggressiveness due to the inability to communicate are commonly seen in older children[2]. Focal motor seizures are the most common type of seizure seen in 70% of affected patients who are usually treated with antiepileptic medications. Electroencephalogram (EEG) pattern during sleep shows frequent spikes in these patients.

Etiopathogenesis although unknown includes slow viral infections and demyelinating diseases of inflammatory origin. Also, autoimmunity has been postulated as one of the mechanisms of this disease pathology[3]. EEG abnormalities and language disturbances tend to persist in most patients[4]. Rehabilitation therapy in the form of speech and language therapy may be needed to improve the quality of life of these patients.

The patient in this case study, was an adult with impaired cognition who was unable to comprehend speech and language with the mental age of a two-year-old child. In literature review, only a single case report of general anaesthesia in a child with Landau Kleffner syndrome was found[5] but no data was found on anaesthetic concerns in the adult group of such patients.

Our main concerns in this patient were maintaining the dosing of antiepileptic drugs during the perioperative period and the interaction of these drugs with anaesthetic agents. Valproate is an inhibitor of hepatic microsomal enzyme systems and may reduce the clearance of many concurrently administered medications, hence careful drug dosing needs to be done in such patients. Many anaesthetic agents affect the seizure threshold. In patients with known seizure disorders such as this patient, medications which can potentiate myoclonus (etomidate, nitrous oxide) and provoke seizure-like activity (sevoflurane, enflurane) should be avoided[6]. Total intravenous anaesthesia with propofol was preferred over inhalational agents for the maintenance of anaesthesia because of its antiepileptic properties. All factors that can precipitate seizure activity such as hypoxia, and hypercarbia were carefully avoided. Maintaining a calm and quiet operating room environment to prevent apprehension in this patient was also important. Difficult airway cart and RAMP position were saviours in this case as the patient was a class I obese with an abnormal airway anatomy. Although awake fibreoptic intubation always remains a gold standard for difficult airway but video laryngoscopy has proven to be a beneficial alternative.

Limited literature on rare diseases makes the anesthesiologist more prone to unknown complications that can be encountered during the procedure which were fortunately not seen in this case. This report aims to create awareness about such uncommon diseases and their possible complications which is pivotal for successful anaesthetic management, especially during emergencies.

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