Airway management of a paediatric patient with temporomandibular joint ankylosis with extra hepatic portal vein obstruction, splenomegaly, hypersplenism, and obstructive sleep apnoea for shunt surgery: A unique challenge

Sir,

We encountered a unique challenge while managing a 9-year-old-male patient with features of growth retardation who was admitted in paediatric surgery for proximal lieno-renal shunt surgery. The patient had abdominal distension and thrombocytopenia due to splenomegaly. He also had temporomandibular joint (TMJ) ankylosis with severely restricted mouth opening. The child was unable to lie supine for an extended period with features suggestive of obstructive sleep apnoea (OSA). History of haematemesis and 2–3 episodes of melena were present in the last 6 months.

He was a known case of oesophageal varices with extra hepatic portal vein obstruction (EHPVO), portal hypertension and splenomegaly for the last 6–7 years. Corrective surgery for TMJ ankylosis release had been planned earlier but was abandoned because of thrombocytopenia. Examination showed severe restricted mouth opening with retrognathia. His pulse rate was 84/min; blood pressure 102/64 mmHg and all other systemic examination were within normal limits. Haemoglobin (Hb) was 8.5 gm% after 2 units of blood transfusion, platelet count was 72,000/cmm. Ultrasound of the abdomen showed a huge spleen of about 18 cm in size. Computed tomography face showed mandibular hypoplasia with bilateral TMJ ankylosis.

Fibreoptic intubation was planned, with readiness for tracheostomy. In the operation theater, monitors like ECG (Electrocardiogram), NIBP (Non Invasive Blood Pressure) and Pulse oximeter for SPO2 were attached. After checking for patency of nares, 2–3 drops of xylometazoline nasal drops were put in nares. A 20-gauge intravenous cannula was secured in the left upper limb before induction of anaesthesia.

Inhalational induction of anaesthesia with oxygen and sevoflurane was done while maintaining spontaneous ventilation. After attaining an adequate depth of anaesthesia, fibre optic-guided intubation was done and a 6.0 mm cuffed endotracheal was secured. Fentanyl 1 μg/kg and atracurium 0.5 mg/kg were administered. The total operative time was around 4 h, and blood loss was around 300 ml. Intraoperatively, the patient was transfused 1 unit of packed red blood cells. The patient was shifted to Intensive Care Unit and was ventilated overnight. The post-operative investigations were normal. The trachea was extubated following morning and then shifted to the ward. Further post-operative course was normal. He was discharged on 8th post-operative day.

Multiple etiologies of TMJ ankylosis have been postulated with trauma and infection as the leading causes.[1] Other causes include congenital, forceps delivery, rheumatoid arthritis, ankylosing spondylitis, infectious diseases such as measles or fibrodysplasia ossificans progressiva. Recently, a correlation between TMJ ankylosis with EHPVO has been described. Hypercoagulability or reduced fibrinolytic activity in EHPVO patients may have predisposed them to joint ankylosis.[2] The possible mechanism could be protein C deficiency or activated protein C resistance in EHPVO patients. Protein C is found to be responsible for inactivation of factor V and factor VIII and it also stimulates fibrinolysis by stimulating the release of plasminogen activator from the endothelial cells.[3] It is possible that protein C deficiency may lead to delayed lysis of fibrin network, formed in exposed medullary spaces post-condylar fracture. Hence, the increased neoangiogenesis in patients with protein C deficiency together with inflammation and wider penetration of osteogenic cells and new vessels from mandibular stump may lead to ankylosis of the joint.[4]

The difficult airway was especially challenging in this child because of the small mouth opening, near total trismus, overcrowding of the soft tissues and associated mandibular/maxillary hypoplasia. In children with TMJ ankylosis, the position of larynx may be altered due to facial asymmetry, complicating the airway.[5] The problems related to difficult airway of TMJ ankylosis are superimposed on already difficult paediatric airway.[6] Paediatric patients with TMJ ankylosis are commonly associated with OSA, which further presents as a challenge to anaesthesiologist as awake intubation is often not a feasible option in them.

Haematemesis from bleeding oesophageal varices and epistaxis from EHPVO with portal hypertension added to the problem of airway management in this patient with TMJ ankylosis. Splenomegaly further aggravated the situation by increasing the risk of aspiration because of abdominal distension and increased risk of bleeding during airway management.

Blind nasal intubation, retrograde intubation over a wire, fibreoptic-guided intubation or tracheostomy are various alternatives described to secure the airway.[5] Fibreoptic intubation is the ideal technique to secure the airway. However, even it is not devoid of complications. Standby tracheostomy or other equipment to secure surgical airway should be kept ready.

In our case, child with TMJ ankylosis associated with EHPVO with portal hypertension, huge splenomegaly, hypersplenism and OSA posed a great challenge for anaesthesiologist to secure the airway. The low platelet count due to hypersplenism can lead to epistaxis during airway manipulation and blur the vision during fibreoptic intubation. However, fibreoptic bronchoscopy still remains the best possible alternative to secure the airway while maintaining spontaneous ventilation in such kind of high-risk 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.

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Conflicts of interest

There are no conflicts of interest.

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