

Undiagnosed bleeding disorder and pediatric airway: Prudence of an anesthetist prevails!

Kumbhar Prathamesh Gurudas¹, Suruchi Richhariya², T Jayasree³

From ¹Senior resident, Department of Anaesthesiology, AIIMS Bhopal, ²Assistant Professor, Department of Anaesthesiology, Mansarovar Medical College and MGU Hospital, Madhya Pradesh, Bhopal, ³Senior resident, Department of Anaesthesiology, All India Institute of Medical Sciences, Mangalagiri, Andhra Pradesh, India

ABSTRACT

A 7-year-old, 28-kg male child with cerebral palsy and intellectual disability was diagnosed to have left-sided pelvi-ureteric junction obstruction and left hydronephrosis, for which he was planned for left-sided pyeloplasty under general anesthesia. The patient had developed a spontaneous oro-nasal bleed, which was managed successfully in the perioperative period without any hemodynamic instability and perioperative respiratory adverse events (PRAE). The patient was extubated on the table without any airway complications after confirming complete neuromuscular blockade recovery.

Key words: Hemophilia A, Laryngoscopy, Oro-nasal bleeding, Pediatric airway, Perioperative respiratory adverse events

Pediatric airway management morbidity is a common problem in pediatric anesthesia [1]. Knowledge of the functional anatomy of the airway in children forms the basis of understanding the pathological conditions that may occur [2]. Various developmental aspects of respiratory physiology put infants and young children at an increased risk of respiratory failure, which is associated with a higher rate of critical incidents during anesthesia [3]. Surgical otolaryngologic emergencies such as foreign body aspiration, post-tonsillectomy bleeding, obstructive laryngeal papillomatosis, peritonsillar abscess, and laryngeal trauma can be life-threatening [4].

In the following case report, we elaborate on spontaneous upper airway bleed and its successful management in a 7-year-old child being operated for pyeloplasty.

CASE DESCRIPTION

A 7-year-old, 28-kg male child with cerebral palsy presented to the pediatric emergency department with vague pain in the abdomen and hematuria. The patient had spastic paraparesis with right spastic upper limb mono-paresis along with mental retardation, though bulbar reflexes and vocalization were well maintained without any evidence of stridor.

On further evaluation, the patient was diagnosed with left pelvi-ureteric junction obstruction with left hydronephrosis.

The patient was planned for left open pyeloplasty under general anesthesia. A pre-anesthetic checkup was done. The patient was fit for the procedure with no anticipated difficult airway and no obvious congenital malformations, and all routine investigations were within normal limits (Platelet count: 3 lacs/dL, prothrombin time -international normalised ratio. [PT-INR]: 1.2, done 4 days before the surgery).

For the surgery, the plan of anesthesia was general anesthesia with endotracheal intubation along with controlled mechanical ventilation. An ultrasonography-guided unilateral transversus abdominis plane (TAP) block administration after closure is planned for perioperative analgesia. Standard intravenous anesthesia induction was done, and the patient was intubated with a 5.5 mm internal diameter cuffed endotracheal tube with direct laryngoscopy. A 12 Fr Ryle tube was placed nasally for aspiration of gastric contents. Both procedures were done gently without any trauma.

The patient was given the right lateral position, and the surgeons proceeded with the incision after taking care of all aseptic precautions. Over the period, it was observed that there was more than usual oozing from the surgical site and the handled tissue. On careful examination, we have observed blood-tinged secretions oozing out from the nose and oral cavity. Due to lateral positioning, all the secretions were got accumulated in the right cheek fossa. Temporarily, we placed dry gauze pieces for soakage of secretions. Tranexamic acid 10 mg/kg was also given IV. The perioperative timeline is given in Table 1.

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Correspondence to: Dr. Kumbhar Prathamesh Gurudas, Senior resident, Swami Vivekanand Boys PG hostel, AIIMS Bhopal, India. E-mail: prathameshk37@gmail.com

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Table 1: Perioperative timeline table

Time from surgical incision	Event
15–20 min	Observing bloody secretions oozing from the nose and mouth.
25 th min	Injection of tranexamic acid 10 mg/kg given IV.
60 min	Continuous bloody discharge from the oral cavity and nose.
110–120 min	Supine positioning, wiping and suctioning of clots.
125 th min	IV injection of propofol+injection of loxicaid given, direct laryngoscopy done and visible clots removed with pediatric Magill's forceps.
130 th min	Saline-soaked gauze placement as a throat pack, administration of xylometazoline nasal drops.
135 th min	Extubation under deeper planes of anesthesia in right lateral decubitus with complete neuromuscular blockade recovery.

Surgery concluded in 2 h. Throughout the surgical time, there was continuous bloody discharge from the nasal and oral cavity. We changed the gauze piece for the soakage 3 times. After giving an ultrasound-guided TAP block on the left side, the patient was made supine, covered adequately, and all monitors and intravenous fluids were connected. We observed some clots near the external nares. These clots were removed by simply wiping the nares. We tried to do suctioning of the oropharynx, which led to aspiration of frank blood and blood clots.

Whatever might have been the cause of bleeding, it was mandatory to control upper airway bleeding, protect the airway and prevent perioperative respiratory adverse events (PRAE). First of all, we deepened the plane of anesthesia with the help of sevoflurane, targeting a minimum alveolar concentration (MAC) above one. We removed the Ryle tube gently after attaching the Yankauer suction tip. We decided to do a laryngoscopy and remove all the visible clots and blood. We observed that, the source of bleeding was the nasopharyngeal area, hence it could be called posterior epistaxis. We administered an injection of propofol 1 mg/kg and an injection of lidocaine 1.5 mg/kg to blunt the sympathetic response, and then proceeded with laryngoscopy. We observed multiple clots of around 1–2 cm in diameter, completely obscuring the view of the glottis. We used pediatric Magill's forceps for intraoral manipulation. All the clots were removed under vision without causing any trauma. After clearing out the oropharyngeal and supraglottic areas, we kept small saline-soaked gauze pieces for the purpose of tamponade effect and to check for any post-nasal drip.

We put xylometazoline nasal spray in both nares to decongest the nasal mucosa. We cleared out any oozing, residual collection of blood or post-nasal drip by throat

packing 2–3 times. We planned to extubate in a deep plane of anesthesia after ensuring complete neuromuscular recovery, as awake extubation in children could have led to agitation, restlessness, and more chances of PRAE, such as laryngospasm, bronchospasm, and sympathetic stimulation, which could have further aggravated the bleeding. After confirming spontaneous breathing efforts and stoppage of bleeding, we administered an injection of neostigmine 50–80 mcg/Kg + injection of glycopyrrolate 10 mcg/Kg slowly, keeping MAC around 0.2–0.4. We waited for regular and adequate breathing efforts, kept the child in the right lateral decubitus, and then extubated. To maintain airway patency, we applied airway maneuvers as needed.

DISCUSSION

The pediatric airway is substantially different from the adult airway, and obstruction can lead to rapid desaturation in infants and young children [5]. Spontaneous epistaxis under general anesthesia is a rare untoward event [6]. The most common causes of epistaxis are inflammatory, environmental, and traumatic causes and medication misuse, but rarely, children may have predisposing anatomic, hematologic, or vascular abnormalities or even sinonasal tumors [7].

Inherited bleeding disorders (IBDs) are caused by quantitative and qualitative alterations of either platelets or plasma proteins involved in coagulation and fibrinolysis [8]. In the reported case scenario, posterior epistaxis under general anesthesia with a normal laboratory profile points toward underlying undiagnosed coagulopathy. Anesthetic management has played a crucial role in airway safety, as there was upper airway bleeding coupled with a challenging pediatric airway.

The pediatric airway is more prone to PRAE, as even the smallest clot, any trickle of secretions or blood, or any degree of airway edema can lead to the same. PRAE can substantially increase the morbidity and mortality [9]. As an anesthesiologist, we must be vigilant and calm, remembering that we should do no harm.

In the case of similar clinical situation, it demands amalgamation of conventional anesthetic practices and novel airway management approaches, which involves: (a) Do not panic; (b) call for help; (c) maintain normothermia, euglycemia, adequate ventilation and oxygenation; (d) maintain depth of anesthesia using intravenous or inhalational anesthetic agents to blunt sympathetic response and airway reactivity; (e) use of direct laryngoscopy or video laryngoscopy for glottic visualization; (f) do no harm: Gentle atraumatic manipulation; (g) use of oxymetazoline or xylometazoline as nasal decongestant [10]; (h) use of saline-soaked gauze pieces and pediatric Magill's forceps for manual clot evacuation; (i) use of well-functioning suction apparatus for clearing out upper

airway; (j) well-equipped pediatric airway trolley; (k) Help of otolaryngologist for epistaxis control if not managed conservatively.

When epistaxis does not respond to simple measures, the source of the bleeding should be located and treated appropriately. Treatments to be considered include topical vasoconstriction, chemical cautery, electrocautery, nasal packing (nasal tampon or gauze impregnated with petroleum jelly), posterior gauze packing, use of balloon system (including a modified Foley catheter), and arterial ligation or embolization [11].

In the post-operative period (post-op day 1), further hematological evaluation drew attention to the presence of factor VIII deficiency in the child. Hemophilia A is a genetic disorder of hemostasis associated with a deficiency or reduced activity of clotting factor VIII (FVIII), which remains unacceptably underdiagnosed in India [12]. Strategies for conserving factor concentrates include educating doctors and patients, prenatal diagnosis, increasing the use of antifibrinolytic agents, physiotherapy, the use of fibrin glue, and simple orthotic and prosthetic measures [13]. Genetic testing, including next-generation sequencing, enables family planning, carrier testing, and prenatal diagnosis [14]. This patient has been registered with the Central Haemophilia Federation of India. Hemophilia care in Asia has evolved to a great extent; however, some challenges remain for which a strategic approach, along with multi-stakeholder involvement, is needed [15].

CONCLUSION

In our case, simple measures taken in a well-organized fashion helped a lot to control the bleeding and to make the upper airway uncomplicated. The more invasive and extensive treatment modalities stated above can be considered in torrential bleeding episodes. Knowledge of pediatric airway anatomy and physiology, PRAE preventive strategies, and a holistic approach for the management of upper airway bleed and underlying coagulopathy is of utmost importance for improved perioperative outcomes.

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