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Case Report

Difficult Ventilation due to Upper Airway Bleeding During Dental Surgery in a Homocystinuria Patient – a Case Report

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Abstract

Aim and background: Homocystinuria is a rare metabolic disorder with characteristic involvement of connective and vascular tissue associated with increased bleeding risk. This article reports a rare and clinically significant case of difficult ventilation caused by upper airway bleeding in a case of homocystinuria during dental rehabilitation surgery, emphasising the challenges faced in airway management and the tailored anaesthetic care given.

Case report: A 9-year-old female with homocystinuria was taken up for dental rehabilitation surgery. Patient sustained profuse upper airway bleeding after attempted nasal intubation and sudden blockage of both bronchi, which led to difficulty in securing the airway and further ventilation difficulties, which were managed with oral intubation, bronchial suctioning, and tailored care to clear the airways and improve the ventilation.

Conclusion: In a case of homocystinuria with increased bleeding tendency due to coexisting Aspirin administration, the anesthesiologist should understand the pathogenesis, complications, and tailor the anaesthetic care with minimal airway manipulation and trauma, and manage all difficult ventilation that may arise as a complication of the same.

Abbreviations

ASA: American Society of Anaesthesiologists; BURP: Backwards Upwards Rightward Pressure manoeuvre; COX: Cyclo-Oxygenase pathway; ET: Endotracheal Tube; MPS: Mallampati Score; NPO: Nil Per Oral; OD: Once Daily; PEEP: Positive End-Expiratory Pressure

Introduction

Homocystinuria is an autosomal recessive genetic disease resulting from a deficiency of the cystathionine-beta-synthase enzyme. The reported incidence is 1:200,000, with involvement of the central nervous system, eyes, cardiac, respiratory, skeletal, hepatic, skin, and hair [1]. Airway management in these patients is often challenging, as attributed to a high-arched palate, abnormal dentition, and marfanoid habitus, which affect positioning [2]. Accumulation of plasma homocysteine can also lead to vascular injury, predisposing the patients to bleeding and thromboembolic risks associated with vascular proliferation, platelet aggregation and activation defects, and an abnormal procoagulant anticoagulant balance [1]. We present here a case report of a 9-year-old homocystinuria patient whose dental rehabilitation surgery had to be deferred in view of difficult ventilation owing to profuse upper airway bleeding.

Case presentation

A 9-year-old female weighing 20 kg presented to the pedodontics department with extensive dental caries and abnormal dentition. After a detailed evaluation, the patient

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was posted for a dental rehabilitation surgery under general anaesthesia. Patient has a normal birth, development, and an immunisation history with a diagnosis of homocystinuria 1 year back as an incidental finding on further evaluation of persisting anaemia. The patient was henceforth started on Vitamin B12 and Tablet Aspirin 75 mg OD for thromboprophylaxis. On examination, the child was found to have poor nourishment with marfanoid habitus, like tall, lean stature with elongated fingers, a high arched palate with abnormally crowded dentition, along with buck teeth and pectus excavatum. No other relevant genetic history in the family or any other past interventions in the patient. No other diagnostic challenges were faced.

With an MPS of 2, a large tongue, and the above-mentioned airway findings patient was prepared as a case of difficult intubation. All other systems were found to be normal. Routine investigations came out to be normal, with special emphasis on normal coagulation parameters. Preoperatively patient was well hydrated following the NPO norms, and Tab. Aspirin 75 mg was continued till the previous day of surgery. After ensuring stable vitals and a pre-anaesthetic checkup child was pre-medicated with 100 mg Ketamine and 0.1 mg glycopyrrolate intramuscularly 30 minutes before and shifted to the theatre. After attaching all ASA standard monitors child was induced with 100% oxygen given via paediatric circuit (weight of the child being 20 kg), ensuring adequate chest rise and 100% saturation along with 30 mcg Inj.Fentanyl and Sevoflurane 2% dial setting. After ensuring proper ventilation child was paralysed with 10 mg of Inj.Atracurium. Following all standard protocols of difficult intubation, the child was first intubated with nasal intubation using a flexometallic cuffed tube of size 5.0. However, on further airway visualisation using normal laryngoscopy by an experienced anesthesiologist, it was found that there was profuse bleeding seen in the oral cavity requiring continuous suctioning, making vocal cord visualisation difficult. Immediately nose was pressed continuously with a gauze piece, which helped in reducing the bleeding. During the attempt, there was a significant saturation drop to 10% following which the attempt was abandoned and mask ventilation was continued. Despite adequate mask ventilation techniques, it was noted that there was minimal chest rise on both sides, hinting at further ventilation difficulties. A small sterile suction catheter was advanced to the right and left bronchi, and sterile suctioning was done. Slowly, the saturation started rising, and after attaining a satisfactory buildup of saturation to 90% following retrieval of clots from the bronchus, another attempt was made at oral intubation with a normal ET tube size 5.0, CL grade was 2b, and the airway was secured with the BURP technique after proper positioning and simultaneous continuous suction in view of the persistent bleeding. After proper confirmation of the ET tube positioning, aided by a normal capnography waveform tube was fixed at 15 cm lip margin. On auscultation, minimal air entry on both sides of the chest with no chest rise was noted, further exacerbated by the decreasing saturation despite proper ET placement. An immediate sterile suctioning of the ET tube was done along with other supportive measures like Hydrocortisone injection and nebulisation with Levosalbutamol and Budecortisone, and continuous pressing of the nose was done. Multiple thick clots were suctioned out from the ET tube, following which high PEEP ventilation was initiated, leading to improved chest rise and increasing saturation. All throughout the event, apart from the reactionary tachycardia patient retained stable hemodynamics. After attaining a saturation of 100% with hand ventilation patient was reversed from the effects of muscle paralysis by appropriate doses of neostigmine and glycopyrrolate, respectively. After ensuring adequate spontaneous breaths and airway reflexes patient was extubated, with the surgery deferred on further discussion with the operating team. Patient maintained a saturation of 98% on room air throughout her post-extubation phase in the recovery room. The patient was monitored using the Aldred score in recovery and attained satisfactory recovery of all systems. No extra neurological monitors were required post-reversal. No other adverse events apart from occasional coughing were reported in the recovery room, with stable hemodynamics. A paediatric consult was sought, where the patient was advised to undergo 24-hour observation and monitoring. After a period of 12 hours on satisfactory recovery child was restarted on oral feeds and Tablet Aspirin 75 mg for further thromboprophylaxis after ensuring a complete cessation of further airway bleeding and normal coagulation parameters. The child was discharged with a follow-up advised after 2 weeks.

Discussion

Homocystinuria is an inborn error of metabolism that affects the transsulfuration metabolic pathway due to a deficiency of the cystathionine beta-synthase enzyme [1]. There will be gross involvement of the connective tissues in patients with homocystinuria. Affected individuals often manifest with Marfanoid features like high arched palate, pectus excavatum, and crowded teeth, making intubation difficult and ventilation problems owing to the chest deformities [3]. General anaesthesia in these patients warrants the need for difficult intubation preparation and proper airway positioning. However, no data regarding difficult airway management in homocystinuria has been published to date [4]. All the necessary equipment and precautions for a difficult intubation were taken in our case, along with the execution of intubation by a skilled anesthesiologist as demanded by the situation.

The association between homocystinuria and vascular events was first reported in 1976, with vascular complications leading as a major cause of mortality and morbidity in these patients. Patients are often at risk of thromboembolic events, which may be attributed to the endothelial injury, procoagulant-anticoagulant imbalance, and platelet dysfunction induced by the high levels of homocysteine in the blood [5]. Literature backed by studies suggests good preoperative hydration to maintain blood viscosity and low-dose aspirin as prophylaxis, and vitamin supplementation linked to vitamin B12 deficiency [6]. Our patient was put on a prophylactic dose of 75mg/day of Aspirin, continued the night before the surgery, which may have predisposed the patient to an event of profuse airway bleeding despite normal coagulation parameters. Any minimal trauma to the fragile connective tissue of the upper airways associated with the effects of Aspirin in preventing platelet aggregation

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by COX inhibition and increased vascular damage by increased homocysteine levels may cause profuse bleeding in the upper airways. In our case, an attempted nasal intubation, added with the effect of the prophylactic dose of Aspirin, might have been the cause of the profuse bleeding, which hindered the direct laryngoscopy and intubation in the first attempt. Hence, we advise minimal trauma during intubation and preference of oral intubation over nasal intubation, even for dental surgeries with intraoperative tube positioning, depending upon the side to be operated, considering the risks vs benefits in these scenarios.

Difficult ventilation is an associated complication of homocystinuria attributed to pectus excavatum [3]. Reports of associated Mounier-Kuhn syndrome in connective tissue disorders causing tracheobronchomegaly associated with air loss during ventilation, despite an adequately sized cuff size can also add to the ventilation difficulties [7]. In our case, difficult ventilation seemed to be attributed to all these factors combined with significant bleeding and clot formation, which might have led to blockage of both bronchi, which is dangerous. Efforts to relieve the bronchial blockage with small-sized catheters advanced into the bronchus have been successful, which led to improved ventilation despite the preexisting ventilation difficulties. Also, a smaller tube has been used for intubation in this case to avoid further trauma and bleeding. Sterile suctioning after intubation has also shown significant ventilation improvement and better clot clearance from the lower airways. Further nebulisations and a steroid bolus have significantly relieved the airway hyperreactivity, which may have been induced by the excessive and prolonged airway manipulation in this case.

Other supportive measures, like proper positioning, keeping in mind the increased risk of osteoporosis, avoidance of nitrous to prevent the inhibition of the enzyme methionine synthase and further aggravation of the condition, and proper maintenance of euvolemia to decrease the risks of thrombosis, were all followed in this case, as backed by relevant literature and studies [8].

The strength of this study was the efficient management of the blocked airway by a team of experienced anesthesiologists, which was essential for the further resuscitation of the patient. The limitation of the study is the use of a cuffed tube and the attempt to do a nasal intubation, which could have been avoided by keeping in mind the necessity to avoid airway trauma in such cases.

Conclusion

Understanding the pathogenesis and complications and tailoring the anaesthetic management is of prime importance in cases of homocystinuria. Minimal airway manipulation and trauma should be the key, considering the increased bleeding tendency with the coexisting Aspirin administration. Also, the coexisting connective tissue disorder can add to the difficult ventilation, which was aggravated by the blockage of the bronchus caused by the profuse upper airway bleeding in this case.

Patient perspective

Perspective couldn't be acquired in view of the patient being a minor. Parents were properly informed about the intraoperative events.

Informed consent

An informed high-risk consent for carrying out the procedure under general anaesthesia, explaining all the relevant complications and necessary interventions, was taken from the father of the patient.

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All the authors declare that they have: Substantial contributions to the conception or design of the work; or the acquisition, analysis, or interpretation of data for the work; AND Drafting the work or revising it critically for important intellectual content AND Final approval of the version to be published; AND Agreement to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved. We would like to thank the Department of Anaesthesia, Christian Medical College, for the institutional support. We certify that Dr Jithin J will correspond on behalf of all the authors with the Editorial board. We also grant him the right to carry out the necessary changes as per the recommendations of the journal and act as a guarantor for the manuscript on our behalf. We also certify that no external funding/grants were received.

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