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Original Research Paper

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A CASE OF OSLER WEBER RENDU SYNDROME AND ITS ANAESTHETIC IMPLICATIONS

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ABSTRACT Osler-Weber-Rendu disease or hereditary haemorrhagic telangiectasia (HHT) is an autosomal dominant genetic disorder. First recognised in the 19th century, this rare often undiagnosed familial disorder with abnormal vascular structures, causes bleeding from the nose and gastrointestinal tract. This condition is characterised by lack of communicating capillaries connecting arteries and veins resulting in multiple arteriovenous malformations (AVMs) and telangiectasia. We describe a case report of anaesthesia management of a patient with Osler-Weber-Rendu disease.

KEYWORDS : Osler-Weber-Rendu disease, hereditary haemorrhagic telangiectasia, Anaesthesia

INTRODUCTION

Hereditary hemorrhagic telangiectasia (HHT) and Osler-Weber-Rendu Syndrome are uncommon disease.[1,2] These are present in many people but remains undiagnosed. It is an autosomal dominant disorder characterized by multiple arterio-venous malformations and telangiectases that affect multiple organ systems. Hereditary hemorrhagic telangiectasia patients have a propensity for bleeding. Especially from the oropharynx, nasopharynx and gastrointestinal tract, as well as from rupture of AVMs of other organ systems.

Children and adults share the same manifestations, with epistaxis and skin telangiectases being the most common. Treatments for HHT include embolization, surgery, laser and hormone therapy. Anaesthetic care of patients with HHT involves very specific interventions with regard to control of bleeding, maintaining adequate oxygenation, and balancing hemodynamic values to optimize perfusion without compromising anaesthetic depth.

Case report

A 5-year-old female child weighing 12 kg presented with supracondylar fracture of right humerus in emergency. Past history of the patient revealed multiple episodes of epistaxis. There was no history of bleeding gums, hematemesis, black coloured stools, red colored urine etc. There was no history of headache, seizures, stroke, shortness of breath, receiving blood transfusion in past. Family history of recurrent epistaxis and telangiectasias on lips and tongue was present in father.

On examination, vitals of patient were stable. On airway examination multiple hemangiomas were present on tongue. No cardiovascular abnormalities were found on examination and rest systemic examination was also normal. After history and examination, a possible diagnosis of Osler Weber Rendu syndrome was made. Hence, the patient was investigated and managed keeping in mind the bleeding complications of syndrome.

Complete haemogram of the patient was normal with hemoglobin of 9.7 g/dl and platelet count of 397000/mcl. Bleeding time was 2 minutes and clotting time was 4.5 minutes. Coagulation profile of the patient was normal with INR of 1.1. Rest of investigations including liver and kidney function tests were within normal limits. ECHO was normal. Detailed airway examination was done. Neck movements were adequate.

Patient was posted for closed reduction with K – wire under GA. Shifted for surgery in emergency OT with 20G intravenous cannula in right hand. ECG leads and Oxygen Saturation Probe was attached. SPO2 was 97%

Intravenous fluids were started after ensuring that there was no air bubble at any place in intravenous drip set assembly. Pre-anesthetic medications; inj midazolam 0.25 mg and inj glycopyrolate 0.1 mg were given. Patient was pre-oxygenated with 100% oxygen for 3 minutes. Induction with inj fentanyl 20+5 mcg, inj propofol 25 mg (15+5+5), and inj atracurium 6 mg loading dose was done. Patient was ventilated for 4 minutes after which check laryngoscopy was done gently. During check laryngoscopy, no hemangioma like lesions were found in pharynx and epiglottic region.

Considering complications of intubation such as hemodynamic instability, hypertension, raised intracranial pressure and also shorter duration of surgery we decided to secure the airway with a supraglottic airway device i.e. i-gel size 2 after proper lubrication and gentle manipulation.

Anaesthesia was maintained with N2O 60%, Oxygen 40%, Isoflurane 0.2- 1.2% and inj atracurium 1 mg SOS was given. Adequate depth of anaesthesia was maintained. Surgery lasted for 1 hour 20 minutes. Reversal was done with inj neostigmine 0.6 mg and inj glycopylorate 0.12 mg. Recovery was smooth and spontaneous. Regular respiration was achieved. I-gel was removed and patient was monitored for one hour in PACU.

DISCUSSION

Patients with HHT may present for embolization procedures of AVMs, as well as for non–HHT-related procedures. Regardless of the procedure or underlying cause, the diagnosis of HHT will have an impact on many aspects of the anaesthetic plan of care.

Patients with HHT require as first priority of anaesthetic management the prevention of bleeding from telangiectases in the oropharynx, trachea and lungs.3 We assessed the

patient's hemoglobin and hematocrit, platelet count, and coagulation studies to evaluate the propensity for bleeding.

Since, the telangiectases are frequently located in the oropharynx, we did thorough airway assessment and check laryngoscopic examination of airway after induction. No telangiectasias were found in the oropharynx. The intubation plan should include a gentle direct laryngoscopy with minimal manipulation of the airway. Considering complications of intubation and shorter duration of surgery we decided to secure the airway with i-gel. Proper lubrication and gentle manipulation done.

It was also considered that intravenous line to be free of any bubble to prevent micro air emboli as patient could have had pulmonary arterio-venous malformation.

Suction catheter was lubricated, inserted and was removed after gentle suctioning. Insertion of devices in the nasopharynx was avoided. During positive pressure ventilation airway pressures were maintained on lower side. Adequate depth of anaesthesia was maintained so as to avoid pain and raised intra cranial pressure as these patients can have cerebral A-V malformations.

CONCLUSION

Be aware of the diagnosis of Osler Weber Rendu syndrome and its various bleeding manifestations. A basic knowledge of the challenges that can present in these patients is first step in providing a safe and effective anaesthetic technique. So, in scenario of possible bleeding manifestation soft gel like cuffless supraglotic airway device i.e. i-gel is better suited, as it prevents compression trauma to surrounding structures, avoids hemodynamic instability and sympathetic stimulation.

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