

## Anaesthetic management of a patient with Osler-Weber-Rendu's syndrome posted for Young's procedure

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Sir,

Osler-Weber-Rendu disease or hereditary haemorrhagic telangiectasia (HHT) is an autosomal dominant genetic disorder. First recognised in the 19<sup>th</sup> 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.

A 65-year-old female with HHT was admitted with a history of repeated attacks of epistaxis over last 15 years, breathlessness, easy fatigability and repeated blood transfusions. Her haemoglobin was 2.5 g%, with a hypochromic microcytic picture. After transfusion of eight units of packed cells over 20 days, it increased to 7.7 g%. Nasal endoscopy and oesophago-gastro duodenoscopy revealed multiple telangiectasias. Other haematological, coagulation and routine investigations were normal. Chest X-ray showed cardiomegaly. Echocardiography and magnetic resonance imaging (brain) were grossly normal. To control epistaxis, the otorhinologist posted her for Young's procedure. There was a history of tonsillectomy and hysterectomy 15 years ago, both uneventful. Family history revealed history of epistaxis in her brother, father and son. A thorough preoperative evaluation was done and written informed consent was taken. The night before surgery, patient was given tablet ranitidine 150 mg and tablet diazepam 10 mg. On the day of surgery, after confirmation of nil by mouth status an intravenous (IV) line was secured, 1 g ampicillin was administered an hour prior, and Ringer's lactate started. Preinduction injection midazolam 1 mg, injection fentanyl 50 µg, injection ondansetron 4 mg were administered IV. After preoxygenation with 100% oxygen for 3 min anaesthesia was induced with injection thiopentone 5 mg/kg, injection lignocaine 2% 60 mg and intubation was aided by injection suxamethonium 100 mg IV. Patient was intubated with number 7.5 cuffed, lubricated endotracheal tube, cuff inflated, throat packed gently and maintained on O<sub>2</sub> + N<sub>2</sub>O (50%:50%), propofol infusion (4 mg/kg/h) and injection

vecuronium 4 mg IV on controlled ventilation with tidal volume of 6 ml/kg. Intraoperatively pulse, blood pressure, respiration, oxygen saturation and electrocardiogram were well maintained throughout. At the end of surgery, after reversal with injection glycopyrrolate 0.5 mg and injection neostigmine 2.5 mg IV and trachea extubated under vision after removal of throat pack, avoiding coughing and straining and was asked to breathe through mouth as nasal packing was done. She was observed for adequacy of ventilation and any nasal bleed thereafter.

Spontaneous and recurrent epistaxis, multiple mucocutaneous telangiectasias (lips, oral cavity, fingers and nose), visceral involvement (gastrointestinal, pulmonary, cerebral, or hepatic AVMs) and a first-degree relative with the condition are the criteria for diagnosis of HHT (presence of three or four criteria-definite, two criteria-suspected, none-unlikely).[1,2] The diagnosis may be confirmed by genetic testing for mutations involving endoglin, ALK-1 or SMAD4.[2]

Onset of symptoms may be delayed until the fourth decade of life. In patients who do not present before the age of 60 years, mortality is minimal. Significant morbidity and mortality in younger patients is attributed to the consequences of visceral involvement. Our patient was 65 years old, with recurrent epistaxis over last 15 years, strong family history, and oral, nasopharyngeal and gastrointestinal telangiectasia. She thus definitely had HHT as per the Curacao criteria.[3] To control epistaxis, nostril closure (Young's procedure) was carried out. This safe and efficacious procedure causes complete cessation of epistaxis in most patients with HHT.[4] To prevent bleeding during instrumentation gentle laryngoscopy and intubation with minimisation of pressor response is important. We prevented the later by use of fentanyl and lignocaine. Intraoperatively rise in airway pressures is to be avoided to prevent rupture of AV malformations[5] and increase in shunt and hypoxia.[6] Thus, the use of lower tidal volumes. AVMs cause loss of capillary filter. Air, thrombi and bacteria are not filtered. A filter at the hub for IV infusions is hence advocated, and endocarditis prophylaxis is required. Rise in blood pressure can be detrimental, so maintaining it within 20% of baseline is important. Blunting of the pressor response and intraoperative propofol infusion helped maintain the blood pressure. However, since patients with AV malformations have a right to left shunt and decrease in systemic vascular resistance (SVR), anaesthesia induced drop in SVR is a concern. An unpredictable response to induced hypotension is seen due to the absence of tone in the AV malformation and so is the response to vasopressors. Preoperative optimisation of fluid status helps maintain intraoperative blood pressure. Postoperative retching and vomiting can cause bleeding due to breaking of the telangiectatic lesions. Use of ondansetron and intraoperative propofol infusion prevented this problem. Similarly, gentle airway suctioning and smooth extubation avoided any mucosal trauma. Nasal packing is placed postoperatively, an

awake and co-operative patient is required. Anaesthetic care of patients with HHT involves very specific interventions with regard to control of bleeding, maintaining adequate oxygenation and balancing haemodynamic values to optimize the perfusion, with optimal anaesthetic depth.

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