ANAESTHETIC MANAGEMENT FOR PATIENT OF LENS DISLOCATION WITH MARFAN SYNDROME

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INTRODUCTION

Marfan syndrome is an autosomal dominant, multisystem disease with a reported incidence of 1 in 3000 to 5000 individuals. Classic manifestations involve ocular (lens dislocation, myopia), cardiovascular (aortic root dilatation with aortic regurgitation, mitral valve prolapse with mitral regurgitation), and musculoskeletal abnormalities (long bone over growth, scoliosis, kyphosis, joint hypermobility).

CASE HISTORY

A 20-year-old female, 47 kg with a history of Marfan syndrome presented to Sumatera Eye Centre (SMEC) reporting a progressive decrease in vision and worsening glare in both eyes during the last few months. Baseline BP (130/80 mmHg), pulse (80 x/min) and SpO2 (100%) were noted. Chest x-ray showed scoliosis thoracalis dextra. Other examinations were within normal limits.

Anaesthetic management: Patient was preoxygenated for 5 minutes. Premedications are 100 mcg Fentanyl, IV, 2.5 mg Midazolam IV and 40 mg lidocaine IV. Patient was induced with 100 mg Propofol IV to induce deep sleep. After epiglottis could be visualized, 50 mg atracurium IV was injected, no. 7 cuffed endotracheal tube was placed.

Anaesthesia was maintained by Oxygen : Air (50% : 50%), sevoflurane (1% - 2%).

Intraoperatively, patient hemodynamic remain stable with minimal blood loss was 10 ml. Surgery lasted for 1 hour 15 minutes and in the end patient was extubated in operating room. Coughing or gagging is minimized by extubating the patient at a moderately deep level of anaesthesia. As the end of the surgical procedure approaches, muscle relaxant reversal is used and spontaneous respiration is allowed to return. Anesthetic agents like 40mg lidocaine IV were continuously given to blunt cough reflexes temporarily during gentle suction of the airway. Extubation proceed there after during spontaneous respiration with 100% oxygen. Patient transferred to recovery room and hemodynamic was closely monitored.

REFERENCES


DISCUSSION

• Patient is relatively tall (168 cm), arachnodactyly, with arm span more than height.
• The patient's father was known to have musculoskeletal abnormalities with height 188 cm.
• Chest x-ray showed scoliosis thoracalis dextra.
• Examination shown a high arched palate, grade II mallampati with pointy chin indicating possible intubation problem.
• Visualization of larynx by Laryngoscopy was difficult due to high arched palate and pointy chin.

CONCLUSION

General anesthesia with endotracheal tube is the chosen technique for this patient. It is prudent to avoid excessive elevation of intracocular pressure as can occur during laryngoscopy, intubation, or response to painful surgical stimulation. Thus, intubation should be performed gently and smoothly with deep extubation. The pre-existing intubation problem is increase in intraocular pressure and respiratory complication in patients with Marfan's syndrome demand careful preoperative assessment and the use of skillful anaesthesia technique to avoid fatal complications.