Anaesthetic Management of Turner's Syndrome posted for Modified Radical Mastoidectomy

Barkha Vaishnav, Ankita Patel**, Heena Chhanwal***, Suktara Sharma*****

Abstract

A 25 year old female, known case of Turner's syndrome was posted for elective Radical Modified Mastoidectomy. Patient had tachycardia, high BP and ECG changes along with all signs and symptoms of Turner's syndrome. During surgery, the aims of anaesthetist were: 1.) To maintain hemodynamic stability, 2.) To provide optimum analgesia intraoperatively and post operatively and 3.) To maintain Peak Airway Pressure within normal limits. Patient was managed intra-operatively by monitoring vitals and maintaining airway pressures. Also airway was secured with blockbuster LMA in order to minimize haemodynamic fluctuations. Surgery was well managed and lasted for 3.5 hours. Patient was managed well intraoperatively with newer supraglottic device and all parameters were served. Patient was smoothly extubated and shifted to recovery with Aldrete score 8 out of 10. Thus, surgery was managed well both intraoperatively and postoperatively and also throughout surgery, patient remained stable. Use of LMA blockbuster was found to be useful and effective in such case for securing airway.

Keywords : Blockbuster LMA, Supraglottic, Turner's syndrome

Introduction:

Turner's syndrome is one of the most common sex chromosomal abnormalities seen in females, caused by Monosomy (XO). It is a condition that results when one of the X chromosomes (sex chromosomes) is missing or partially missing. Turner's syndrome can cause a variety of medical and developmental problems, including short height, failure of ovaries to develop and heart defects.[1] In this syndrome, the chromosome change happens randomly before birth.

Case report:

A 25 year old female patient, weighing 45 kg, known case of Turner's syndrome was posted for Modified Radical Mastoidectomy on 28/10/2021. Patient was kept nil by mouth overnight before surgery and was brought on OT table.On table, monitors were attached and IV line was secured with two large bore canula. Right sided arterial line was taken by Seldinger's technique and CVP line was kept standby. IV fluids (crystalloids) were administered. Patient had sinus tachycardia (P=130), high BP(160/100mmHg), missed beats (3-4/min) on palpation and ECG changes (v2-v3 T inversion). Expert Echo showed mild degree of coarctation of aorta. Medical fitness was obtained. Rest all blood investigations and other routine investigations were normal. Written and informed consent of patient's relative was taken.

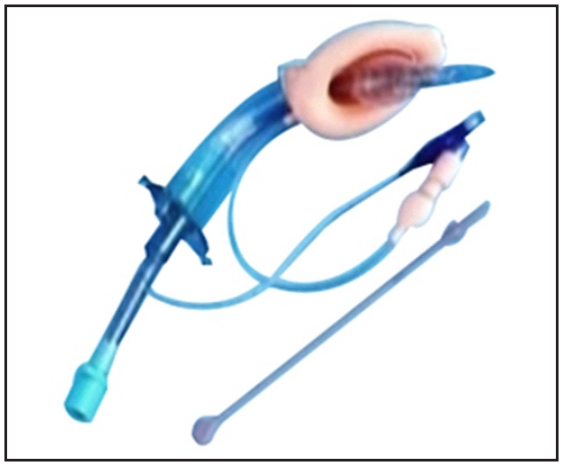
Premedication was given with Inj. Glycopyrolate 0.2mg IV, Inj. Midazolam 1mg IV, Inj. Fentanyl 100mcg IV. Induction was done with Inj. Propofol 100mg IV. Airway was secured by inserting LMA blockbuster "size 3" and post insertion, no vital fluctuations were observed. Blockbuster LMA is a newer advanced supraglottic device which has good seal and convenient for long time ventilation.[2] It is easy to insert and fix as well as all complications associated with intubation can be avoided. It has two passages- one for inserting endotracheal tube and the other one for gastric tube so that chances of gastric reflux and aspiration are minimized. There is a guide wire along with LMA by which the tube can be pushed into the

* Resident
** Assistant Professor
*** Professor & Head, Department of Anaesthesia, GCS Medical College Hospital & Research Centre, Ahmedabad, Gujarat, India
**** Associate Professor, Department of Otorhinolaryngology, GCS Medical College Hospital & Research Centre, Ahmedabad, Gujarat, India
Correspondence : Dr. Ankita Patel
E-mail : drankitapatel30@gmail.com

airway and LMA can be removed subsequently. Thus, LMA aids easy and smooth intubation. Muscle relaxation was given by Inj. Atracurium 25 mg IV and maintenance was done with Oxygen+N_2O+ Sevoflurane+ Atracurium 5mg SOS. Surgery lasted for 3.5 hours without any significant events. Patient was smoothly extubated and was able to follow verbal commands with adequate tone and power. Patient was shifted to recovery room for observation and no post-op complications were found. Aldecrete score was noted which was 8 out of 10 while shifting.

Thus, surgery was managed well both intraoperative and postoperative and also throughout surgery, patient remained stable. Use of LMA blockbuster was found to be useful and effective in such case for securing airway.[3]

Figure 1: LMA (Laryngeal Mask Airway) blockbuster



Discussion:

Turner syndrome is a frequent and complex genetic abnormality affecting women, being associated with a wide variety of and anatomical physiological changes especially related with the airways and cardiovascular system. The challenges for Anaesthesiologist include a short neck and maxillary and mandibular hipoplasia which might be responsible for difficult airway.[4,5] The shorter length of trachea as well as the higher location of its bifurcation can predispose to bronchial intubation and accidental endotracheal extubation when the tracheal cannula is under traction.

In this patient we are using LMA Blockbuster[6] size 3 for securing airway. This newer supraglottic airway device has good airway seal and convenient for long time ventilation. It is easy to insert and fix as well as complication associated with intubation can be avoided and no need for laryngoscopy.

Mastoidectomy is a procedure usually done for patients with Middle ear infections, chronic long standing infections (like otitis media) affecting Middle Ear and frequently accompanied by Cholesteatoma (a destructive non- cancerous skin cyst) or an unhealed ear drum perforation. Hence this procedure was mandatory in order to preserve long term hearing.[7]

Conclusion:

Turner's syndrome is a genetic abnormality with significant anatomical and physiological abnormalities, therefore knowledge of this disorder allows for a safer anaesthetic management. It can be achieved with usage of LMA blockbuster for securing airway that may reduce Peri-operative morbidity and mortality.

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