Anaesthetic Management of a Patient with Complete AV Block Associated with Congenital Corrected TGA with Difficult Airway

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Congenital corrected Transposition of Great Arteries (CCTGA) is a rare anomaly and comprises less than 1 % of all forms of congenital heart diseases. The AV conduction tissues may be congenitally abnormal in terms of both their location and function in specific forms of CHD, most notably L-TGA and endocardial cushion defects.[1,2] Patients with complete heart block coming for non-cardiac surgeries pose a challenge to anaesthesiologist. Further a difficult airway associated with such a case is an additional challenge.

CASE REPORT

A twenty six year old male patient with alleged history of fall from a two-wheeler (RTA) with complaint of nasal and oral bleed and periorbital edema one month back presented to the maxillofacial department. He had no history of loss of consciousness, vomiting or seizures. The patient was posted for maxillofacial surgery.

During pre-anaesthetic evaluation, a past history of dizziness and dyspnoea on exertion (grade II) was elicited. His pulse rate was 45/mt and blood pressure was 100/80 mm/hg.

Cardiovascular examination revealed -- S1, S2 and a Loud S2 (A2 component), with no murmurs. Respiratory system revealed normovescicular breath sounds; no added sounds.

Oral cavity examination revealed limited mouth opening of only one finger. There was gross irregularity of dentition due to fracture. Thyromental distance was 5.5 cms. Patient had nasal bone fractures. Cervical spine examination was normal with no restriction of neck movements. Investigations like complete haemogram, BT/CT, RBS, blood urea, Sr creatinine, and ECG were done. Complete heart block was detected. The patient was evaluated further with Echo-cardiography and Treadmill test.

Echo-cardiography showed congenitally corrected Transposition of Great Arteries with Ejection fraction of 42 % with mild aortic regurgitation and mild right ventricular (systemic ventricle) dysfunction. Tread mill test showed increase in heart rate from 50 to 86 beats per minute at which patient became symptomatic. Case was accepted under ASA Gr III physical status and risks were explained to the patient and informed risk consent was taken.

A 18G vasofix cannula was inserted in upper limb and connected with extension line with bi-way for administration of drugs. In view of complete heart block with significant history of dizziness, prophylactic transvenous temporary pacemaker was implanted via femoral vein on the previous day, before the proposed surgery. The heart was paced at the rate of 55 beats per minute. The patient was kept under observation in cardiac ICU and maintained good hemodynamic stability with that paced rate.

As the patient had a predicted difficult airway and it was decided to go ahead with awake Fibre-optic nasal intubation. Consent for Tracheostomy was taken in view of difficult airway. Endo-carditis prophylaxis was given with inj amoxycilin 2 gm and inj gentimicin 80 mg iv.

Nasal packing was done with 4% lignocaine which was removed after 20 minutes.

In the operation theatre patient was premedicated with Injection glycopyrolate 0.2mg i.v Injection midazolam 1mg i.v Injection fentanyl 50 micrograms i.v., Bilateral superior laryngeal nerve block was administered with 2% lignocaine. Then oral cavity was sprayed with 10% lignocaine spray. A trans-tracheal lignocaine spray with 4% lignocaine was administered. The dose of local anaesthetic given was calculated and kept much below the toxic dose for the patient.

Invasive blood pressure monitoring was planned and radial artery was cannulated with 20G vasofix and connected to transducer and zeroed. Inj. Isoprenaline was kept ready on table.

Awake nasal fibre optic intubation was performed and airway was secured with no 7.5 mm nasal endotracheal tube and after confirming proper placement, inj. thiopentone 250 mg i.v and injection vecuronium 5mg i.v was given. Inj tramadol 100mg was given intravenously. Monitoring included pulse oximetry, ECG, Invasive blood pressure, capnography and temperature. Maintenance was done with oxygen, nitrous oxide and 0.6% isoflurane with intermittent boluses of injection vecuronium 1mg intravenously.

Intraoperative hemodynamic parameters were within

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normal limits. At the end of the surgery on table extubation was carried out with all reflexes intact and good muscle power. Patient was shifted to Cardiac Intensive care unit with the temporary pacemaker in situ and monitored for 48 hours. Patient was maintained on inj. fentanyl infusion for postoperative pain relief.

There was good hemodynamic stability throughout the postoperative period.Temporary pacemaker was removed when patient's vitals were stable after 48 hours.Patient was advised to seek further cardiac consultation regarding implantation of a permanent pacemaker.

DISCUSSION

Few patients with CCTGA live past 50 years, with the median age of death at 40 years.[3] The cause of death is usually heart failure, resulting from the inadequacy of right ventricle to support the systemic circulation.[3] Patients traditionally present with heart failure or heart block[4] but other common presenting characteristics include single loud second heart sound, heart murmur, cyanosis, bradycardia and tachyarrhythmia.

Congenital corrected Transposition of Great Arteries with complete heart block is a disorder of cardiac conduction system. The adult congenital heart disease patient at baseline or in need of intervention presents unique challenges to anaesthesiologists. Anaesthetic complications encountered in such patients with complete heart block include bradycardia, hypotension, arrthymias, cardiac arrest or even sudden death. Permanent pacemaker is the therapy of choice in symptomatic patients, hence in this patient temporary pacemaker insertion was carried out prior to surgery.

The AV-Node and the bundle of His are usually anterior and the long penetrating bundle is vulnerable to fibrosis with advancing age . In L-TGA, it is estimated that 3% to 5% of patients will have complete AV block at birth, and an additional 20% will develop spontaneous complete block by adulthood.[5] This makes the conduction system somewhat tenuous with a progressive incidence of complete AV block occuring approximately at the rate of 2% per year.[7,8]

Katherine W et al have reported successful management of three cases for cesarean section with TGA associated with varying degrees of conduction defects using continuous lumbar epidural catheter technique.[6] In our case the patient had complete heart block with symptoms, thus necessitating the use of transvenous pacemaker during perioperative period. Further our patient had difficult airway, hence a decision to go ahead with awake fibreoptic at the first instance was taken to avoid any trauma to airway and ensure haemodynamic stability.

A case of Congenital corrected Transposition of Great Arteries with complete heart block with difficult airway can be managed with a well planned multidisciplinary team approach under general anaesthesia by awake fibreoptic endotracheal intubation and prophylactic temporary pacemaker in situ with continuous ECG monitoring. Though permanent pacemaker is the treatment of choice in such cases, a temporary trasvenous pacemaker is a must in these cases coming for non-cardiac surgeries until permanent pacemaker implantation. Meticulous perioperative care of such a patient leads to favourable outcome without complications.

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