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Case Report

Anaesthesia Section

Anaesthetic Management in a Patient of Goldenhar Syndrome Posted for Drainage of Brain Abscess with External Ventricular Drain Insertion

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ABSTRACT

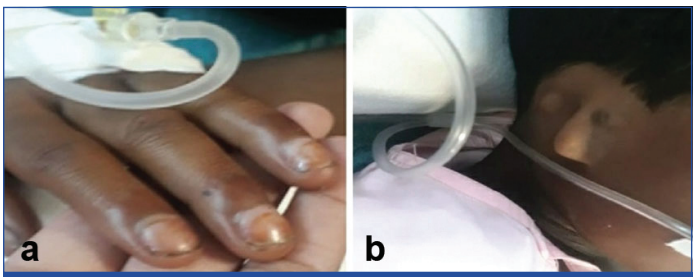
Goldenhar syndrome or Oculo-auricular Vertebral Dysplasia (OAVD) is characterised by a wide range of congenital anomalies including ocular, auricular, facial, cranial, vertebral and cardiac abnormalities. Facial and oral abnormalities especially mandibular hypoplasia, micrognathia, high arch palate and limitation of neck movements resulting from vertebral anomalies, the difficult intubation, laryngoscopy and mask ventilation were expected. Hereby, presenting the case report of a 6-year-old male child, known to have goldenhar syndrome, who underwent drainage of left sided brain abscess with external ventricular drain insertion. In view of the anticipated difficult airway and cardiac anomalies, careful preoperative evaluation, preparation and well formulated contingency plans for airway maintenance, endotracheal intubation and intraoperative haemodynamic is required to combat the perioperative anaesthetic challenges in all cases of goldenhar syndrome.

Keywords: Anaesthetic challenges, Cardiac, Congenital, Difficult airway, Oculo auricular vertebral dysplasia, Peroperative

CASE REPORT

A 6-year-old male child presented with sudden onset fever 103°F for five days, four episodes of non projectile vomiting, headache for two days, and two episodes of generalised tonic clonic seizure. The child was diagnosed four years back with Tetralogy Of Fallot (TOF) with Ventricular Septal Defect (VSD) and severe pulmonary stenosis diagnosed on 2D Echocardiogram (2D Echo). Blalock Taussig (BT) shunt surgery was done at 4 years of age. He was known to have Goldenhar syndrome and weighed 14 kg at the time of presentation. Thus, he belonged to American society of Anaesthesiologist (ASA) grading class III. Patient was fully immunised and milestones achieved as per age. The child was not on any anticoagulants.

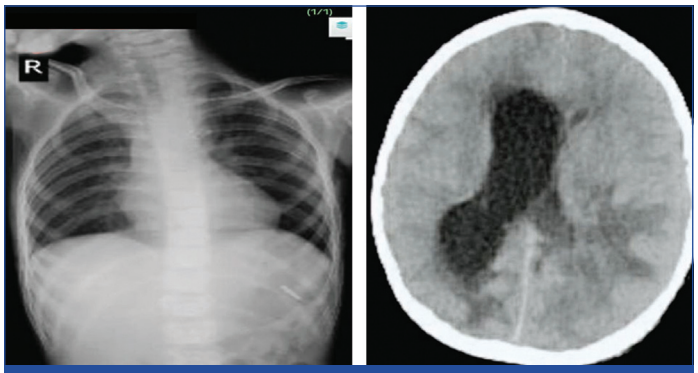
On clinical examination, the child had features of microtia, partially deaf (could hear only loud sounds), coloboma of both eyes, high arched palate and grade 3 clubbing [Table/Fig-1]. Cardiac examination revealed grade 3 pan systolic murmur (TOF with VSD).



[Table/Fig-1]: a): Grade 3 clubbing; b): Microtia in patient.

Patient had a glasgow coma scale score of E3V3M4 with normal tone and power of grade 4/5 in all the 4 limbs. Deep tendon reflexes were present with plantar extensor response bilaterally. Fundoscopy showed papilledema. Haemogram and coagulation profile within normal limits. Blood culture showed no growth. Lumbar puncture and Cerebrospinal Fluid (CSF) study did not reveal any abnormalities. The 2D Echo findings revealed continuous flow across the BT shunt, large VSD with overriding of aorta, stenotic pulmonary valve, severe hypoplasia of pulmonary arteries (6 mm), pulmonary artery hypertension and ejection fraction of 60%. There was no evidence suggestive of infective endocarditis.

X-ray neck Anteroposterior (AP) and lateral view showed no abnormality, while chest radiograph showed a boot-shaped heart [Table/Fig-2]. Magnetic Resonance Imaging (MRI) brain showed cerebral abscess in left temporoparietal region extending into left ventricle with perilesional oedema [Table/Fig-3].



[Table/Fig-2]: Chest X-ray showing mild cardiomegaly (boot shaped heart).
[Table/Fig-3]: Magnetic Resonance Imaging (MRI) brain showing cerebral abscess left temporoparietal region. (Images from left to right)

Anaesthetic Management in Operation Theatre

Nil by mouth status was confirmed and informed written consent was taken. Antibiotic prophylaxis (inj. ceftriaxone 50 mg/kg intravenously) was given 30 minutes prior to surgery. Multipara monitors were attached and peripheral venous line 22 gauge secured. De-airing of intravenous line was done and ensured that no air bubbles were present in the i.v. line. Baseline values were recorded as pulse rate of 126 bpm, non invasive Blood Pressure (BP) of 100/60 mmHg, SpO_2 was 92% with 6 litre O_2 and end tidal carbon dioxide concentration value of 22 mm Hg. Preoperatively SpO_2 maintained in 80-85% range (off O_2). The patient was given inj. glycopyrrolate 0.004 mg/kg i.v., inj. midazolam 0.05 mg/kg i.v. and inj. fentanyl 2 mcg/kg i.v. Preoxygenation was done with 100% oxygen. Induction was achieved with inj. ketamine 2 mg/kg i.v.

After confirmation of ventilation, neuromuscular blocking agent inj. atracurium 2 mg/kg i.v. was given. After 3 min of oxygenation, the child was intubated with uncuffed endo tracheal tube number 5.5. High up anterior larynx with Cormack Leanne grade 4 was visualised on laryngoscopy. Vocal cords were not visible even with optimum positioning and assistance. The intubation was difficult and was done on second attempt with the help of paediatric bougie using Backward Upward Rightward Pressure [BURP] manoeuvre. Anaesthesia was maintained with sevoflurane, oxygen and air. Correct placement of endotracheal tube was confirmed by auscultation and capnogram. Following this, femoral artery cannulation was done using 22 G catheter. Right subclavian vein cannulation was done using 5.5 Fr triple lumen catheter. Electrocardiogram (ECG), capnography and arterial blood pressure monitoring, 2 hourly monitoring of random blood sugar and Arterial Blood Gas (ABG) analysis and hourly urine output monitoring were done throughout the surgery. Baseline arterial BP was 90/64 mmHg. Intraoperatively, the patient developed hypotension (70/40 mmHg). Inj. Phenylephrine 10 mcg/kg i.v. bolus was given and it was repeated after 5 minutes. Eventually, the patient was started on inj. noradrenaline (2/50) infusion 0.05-0.1 mcg/kg/min titrated according to the blood pressure values.

Postoperatively, noradrenaline infusion was discontinued as the child was haemodynamically stable. The child was shifted to Paediatric Intensive Care Unit (ICU) postoperatively and kept for 2 days. No episodes of convulsions were there in the next 48 hours and he was haemodynamically stable. The child was extubated on post operative day 2. The arterial and central lines were removed, nasogastric tube feed was started and was shifted out to ward on post operative day 3. The child was discharged on postoperative day 7.

DISCUSSION

Goldenhar syndrome or Oculo-auricular Vertebral Dysplasia (OAVD) is characterised by a wide range of congenital anomalies including ocular, auricular, facial, cranial, vertebral and cardiac abnormalities. Aetiology is not fully established [1]. Chromosomal abnormalities, abnormal development of neural crest cells, ingestion of drugs cocaine, thalidomide, retinoic acid, alcohol during pregnancy factors leading to development of this disease [2,3]. Autosomal recessive or dominant inheritance possible. Male predominance with male:female ratio 2:1 [4].

Incidence rate 1 in 5600. Facial and oral abnormalities especially mandibular hypoplasia, micrognathia, high arch palate and limitation of neck movements resulting from vertebral anomalies-difficult intubation, laryngoscopy and mask ventilation expected [5]. Goldenhar syndrome was first described by Dr. Maurice Goldenhar in 1952 [6]. Feingold and Baum criteria is used to diagnose Goldenhar syndrome-eye abnormality associated with ear, mandibular, vertebral or cardiac anomalies (at least 2 of the above should be present) [7].

Thorough preoperative evaluation and preparation for anaesthetic and airway management as per the clinical situation along with adequate monitoring is required. The airway management is challenging and can be complicated by the presence of retrognathia, micrognathia, vertebral anomalies, mandibular hypoplasia, high arched palate cleft palate [8].

Decision was made to proceed with laryngoscopy and oral endotracheal intubation as this patient had adequate mouth opening and no vertebral anomalies/gross facial deformities. Difficult intubation facilities such as curved and straight laryngoscope blade sizes 1 and 2, Laryngeal Mask Airway (LMA) size 1, 1.5 and 2, paediatric stylet, smaller size endotracheal tubes and standby fibre optic paediatric bronchoscope was kept ready [9].

In 2005, Grewal A et al., published a similar case report of goldenhar syndrome in a 2-year-old child, who was posted for limbal dermoid excision. The child had marked micrognathia which hampered the insertion of a small macintosh laryngoscope blade along with mandibular hypoplasia, high arched palate and large tongue in relation to small jaw which made glottis visualisation impossible [10]. But blind oral intubation could be done on 2nd attempt. In 2017, Choudhury M and Malhotra Kapoor P, published a case series which reported a similar case with high up larynx with adequate mouth opening and another case of goldenhar syndrome with unanticipated difficult airway without the presence of any pre disposing factors [11].

Recurrent desaturation and bradycardia was reported in both these cases and tracheostomy had to be done. TOF and VSD are the most commonly reported heart defects in goldenhar syndrome.

Presence of cardiovascular anomalies {in this patient tetralogy of fallot with ventricular septal defect, severe pulmonary stenosis and status post Blalock Tausig (BT) shunt surgery} was an additional challenge for anaesthetic management. Arterial line cannulation is important for blood gas analysis and arterial BP monitoring. Anaesthetic induction, airway manipulation and surgical blood loss can cause potential unstable hemodynamic in these patients [12]. There is risk of diastolic run off and coronary and cerebral steal associated with BT shunt repair. Diastolic BP should be maintained slightly above the range in operated case of BT shunt patients to maintain the vital organ perfusion [13]. This patient developed hypotension after anaesthetic induction and was managed with inj. phenylephrine i.v. boluses and inj. noradrenaline infusion. Phenylephrine increases systemic vascular resistance and maintains BP. Phenylephrine was preferred over ephedrine and Mephentermine as the latter drugs have a positive ionotropic effect, increases the myocardial work load and oxygen consumption [14]. Also, ephedrine increases the pulmonary artery pressure. Inhalational anaesthetic of choice was sevoflurane because of its better haemodynamic stability and quick recovery time and also, it maintains cardiac output by stabilising heart rate and preventing bradycardia [15].

Prophylaxis for infective endocarditis should be considered in cases with associated with cardiac anomalies. Ideal protocol of management should also include intraoperative transoesophageal echocardiography and Bispectral index monitoring for assessment of depth of anaesthesia to regulate the anaesthetic drug dosing and to detect cerebral perfusion pressure [16].

CONCLUSION(S)

Anaesthetic techniques for patients with goldenhar syndrome depend on the type, extent and severity of craniofacial vertebral anomalies, associated cardiovascular problems, and nature of surgery. Careful preoperative evaluation, preparation and well formulated contingency plans for airway maintenance and endotracheal intubation and intraoperative haemodynamic is required in all cases of goldenhar syndrome.

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