GOLDENHAR SYNDROME- ANAESTHETIC MANAGEMENT

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HOW TO CITE THIS ARTICLE: Bashir R, Qureshi FM. Goldenhar syndrome- anaesthetic management. J. Evid. Based Med. Healthc. 2018; 5(18), 1501-1502. DOI: 10.18410/jebmh/2018/315

PRESENTATION OF THE CASE

A 6-month-old male child presented to Ophthalmology Outpatient Department of our hospital with decreased vision and painless swelling near left eye since birth. On ophthalmic examination, the child had hypertelorism, partial coloboma of both eyes, and bilateral congenital cataract. There was inferolateral limbal dermoid of about 2.5*2.5 mm size with convergent squint of left eye. The physical examination showed facial asymmetry due to underdevelopment of left maxilla and normally appearing mandible. Airway evaluation showed Mallampatti III with restricted movements of head and neck. On ear examination, both ears were small and low set with stenosed bilateral external auditory canals. Cardiovascular and central nervous system examinations were normal with no associated congenital anomaly. X-ray chest was normal. Computed tomography of head showed bilateral stenosed external auditory canals, hypoplastic left maxilla and cleft palate. The patients had undergone repair of cleft lip left side at age of 3 months. The cleft palate was extending almost upto premaxilla from uvula. Haemoglobin, bleeding and clotting time, plate counts, blood urea, serum creatinine, blood glucose, liver enzymes were within normal limit.

Figure 1 **Figure 2**

Figure 1. Picture of the Child Showing, Hypertelorism, Coloboma and Operative Scar of Cleft Lip.
Figure 2. Picture of the Child Showing Cleft Palate.

CLINICAL DIAGNOSIS

Goldenhar syndrome, also known as oculo-auriculo-vertebral dysplasis, is a rare congenital disorder.[1,2]

Financial or Other, Competing Interest: None.
Submission 10-04-2018, Peer Review 16-04-2018, Acceptance 28-04-2018, Published 30-04-2018.
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DOI: 10.18410/jebmh/2018/315

DIFFERENTIAL DIAGNOSIS

1. Nager acrofacial dysostosis syndrome.
2. Treacher-Collins Syndrome.
3. Duane retraction Syndrome.
4. Pierre Robin Syndrome.
5. Fraser Syndrome.
6. Van der Woude Syndrome.

PATHOLOGICAL DISCUSSION

Goldenhar syndrome, also known as oculo-auriculo-vertebral dysplasias, is a rare congenital disorder, first described by Dr. Maurice Goldenhar in 1952. The syndrome occurs randomly, with no apparent cause. Most cases are not inherited; however, few families show unclear inheritance pattern. The male to female ratio is 2:1 and its occurrence is general 1 in every 5000 live birth.[1,2] This syndrome only involves face, eyes, ears and vertebrae and has wide spectrum of symptoms and physical features that range from mild to severe and is often associated with difficult intubation. Usually children with Goldenhar syndrome have normal life span.

Goldenhar syndrome is a rare congenital disorder and consists of ocular, auricular and skeletal anomalies with variable presentations. In most cases one side of body is affected but in 10-33% cases it may be bilateral.[2,3] Ocular abnormalities include epibulbar dermoids, coloboma, microphthalmia, palpebral fissures, strabismus, and vision defects. Amongst auricular features, preauricular skin tags and accessory tags are common.[1] The spine bifida is commonest and least severe of all anomalies; craniofacial abnormalities may include maxillary, mandibular and temporal hypoplasia, macrostomia, cleft lip and palate. Many affected individuals have additional skeletal, neurologic, cardiac, pulmonary, renal, and/or gastrointestinal abnormalities. Feingold and Baun listed criteria for Goldenhar syndrome, of which at least two are require for the diagnosis.[4,5,6]

DISCUSSION OF MANAGEMENT

Pre-anaesthetic assessment of airway was made by month opening, Mallampatti classification, thyromental distance and neck movement. A written informed parental consent was take after discussing risks and management with parents and ophthalmic surgeon. The difficult airway cart with percutaneous and surgical tracheostomy set was kept ready. The patient was premedicated with glycopyrrolate (0.1 mg/kg) and fentanyl (1 mcg/kg).

The patient was put to sleep using inhalational induction with sevoflurane 1-3% using oxygen 100% as carrier gas. A 24 G I/V cannula was secured and intravenous infusion of

ringers lactate was started at 6-8 ml/kg[4] and inj. Propofol 1% 05 mg/kg was given intravenously to achieve adequate induction with very little haemodynamic changes. The standard monitors for heart rate (ECG), systemic blood pressure (NIBP), pulse oximetry (SPO_2) were attached. The patient was premedicated with glycopyrrolate (0.1 mg/kg) and fentanyl (1 mcg/kg).

After induction, classic LMA size 1½ was put in and secured in place. The patient was put on oxygen, nitrous oxide and isoflurane (1-2%) for maintenance of anaesthesia with patient breathing spontaneously using paediatric circuit (J-R modification of Ayre's T-piece). Left side lens aspiration was done. The surgical procedure was smooth and lasted 30 minutes. Ondansetron (0.5 mg/kg) was given to prevent postoperative nausea and vomiting 15 minutes before completion of surgery. The heart rate, blood pressure and oxygen saturation were observed intraoperatively and postoperatively for 30 min.

As soon as the surgical procedure was finished, and eye bandaging done, LMA was taken out in still deeper planes of anaesthesia to prevent haemodynamic response to extubation. N_2O and isoflurane were put off and patient kept on 100% oxygen till he regained consciousness. The intraoperative SPO_2 and haemodynamics remained stable. The patient was shifted to the recovery room and kept under observation where he had a smooth and uneventful early recovery and was sent to postoperative ward

The heart rate, blood pressure and SpO_2 were monitored every 3 minutes. The heat rate remained intraoperatively between 100-120 bpm, the blood pressure was between 80-100 mmHg systolic and diastolic between 40-60 mmHg. The SpO_2 remained between 97-100% intraoperatively.

After extubation these vitals were monitored for 30 min. which remained within 10% of baseline which was statistically insignificant (p value >0.05). After 30 minutes the patient was pain free, conscious and comfortable in his mother lap was kept on IV fluids Ringers lactate for 3 hours. After 3 hours the patients were given oral sips of apple juice which he tolerated. He had no episode of postoperative nausea and vomiting.

FINAL DIAGNOSIS

Goldenhar Syndrome with familial tendency.

Awareness of this congenital disorder will help to plan an anaesthetic management depending on the type and severity of anomalies. Fiber optic intubation, Light wand/stylet intubation or LMA are various options depending on the type of surgery to be formed. Our anaesthetic plan was a safe and an alternate one for patients with this disorder with early recovery.

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