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Case Report

Anesthetic implications in Poland syndrome

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ABSTRACT

Background: Poland Syndrome (PS) is a rare birth defect described as unilateral absence of pectoralis major, pectoralis minor, serratus anterior and external oblique muscles and upper limb anomalies. It is also associated with variable thoracic muscle, chest wall and lower limb deformities. Dextrocardia, diaphragmatic hernia, liver and GIT anomalies, renal agenesis have also been reported in literature. In addition, there is association of PS with patient's susceptibility to malignant hyperthermia (MH). For patients requiring general anesthesia, special preparation and execution are recommended.
Case Report: We share our anesthesia management in a child with PS posted for ear reconstruction and described that patients with PS requires special preparation and attention in intraoperative and post-operative period.
Conclusion: In patients of PS requiring general anesthesia adequate preparation must be made keeping in mind not only the anatomical changes but also the much higher possibility of MH in such patients.

1. Introduction

Poland Syndrome (PS) is a rare birth defect described as unilateral absence of pectoralis major, pectoralis minor, serratus anterior and external oblique muscles and upper limb anomalies. It is also associated with variable thoracic muscle, chest wall and lower limb deformities.[1] Dextrocardia, diaphragmatic hernia, liver and GIT anomalies, renal agenesis have also been reported in literature. In addition, there is association of PS with patient's susceptibility to malignant hyperthermia (MH). While most of the anatomical abnormalities can be detected during scanning for congenital anomalies in any syndromic patient, the likely association of PS with MH is difficult to diagnose and can have serious anesthetic implications. With this latter association in mind, we share our anesthesia management in a child with PS posted for ear reconstruction.

2. Case Report

A 12-year-45 kg, ASA-1 female, diagnosed case of PS was posted for stage-1 ear reconstruction for right microtia. The patient also had right facial nerve palsy, right thoracic cage deformity, underdeveloped right breast and depressed lower half of the sternum due to sternocostal cartilage defect. Upper limb examination showed shorter range of arm movement on left side compared to right (Figure 1). Spine examination revealed scoliosis in the upper thoracic region with convexity towards right (Figure 2).

Despite these anomalies her respiratory & cardiovascular functions were well-preserved. There was history of snoring

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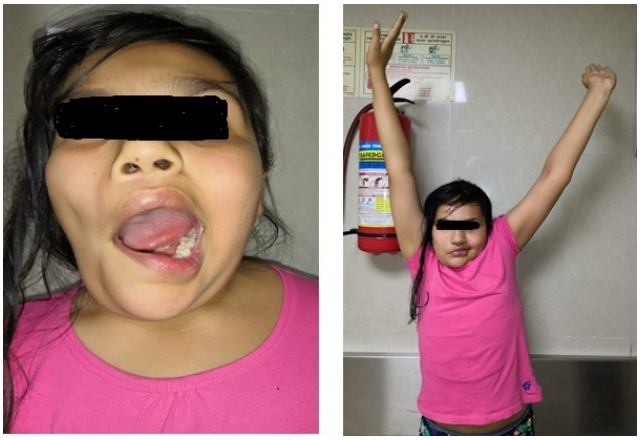

Fig. 1: Patient had right facial nerve palsy, right thoracic cage deformity. Upper limb examination showed shord arm sapn on left side compared to right

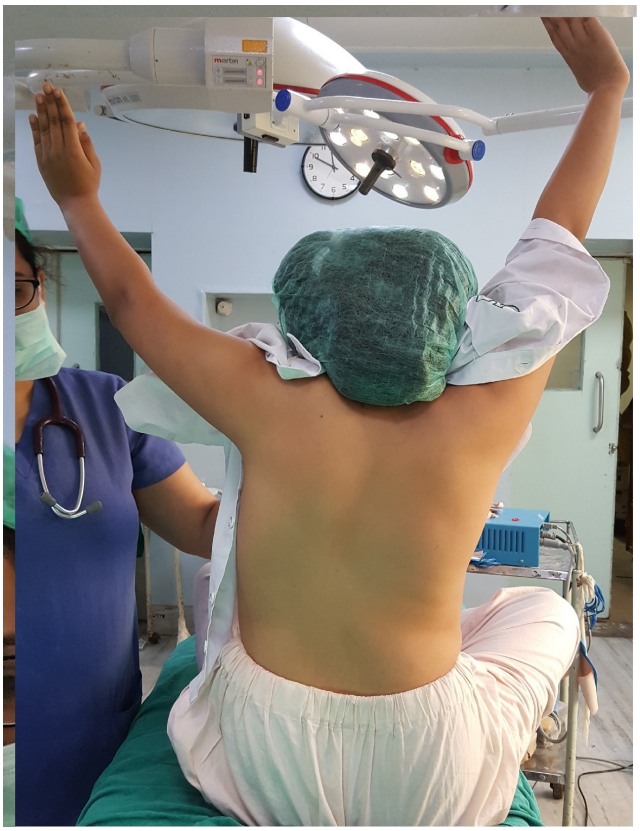

Fig. 2: Spine examination revealed scoliosis in the upper thoracic region with convexity towards right

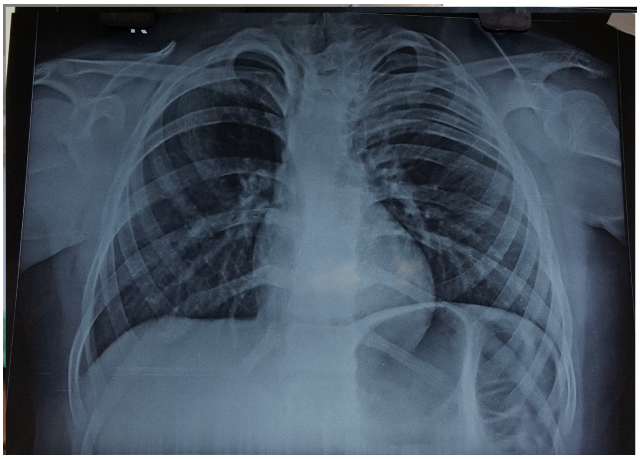

Fig. 3: Chest X-ray (PA) revealed unequal number of ribs (7 on right & 11 ribs on left side), left upper ribs crowding, elevation of left hemidiaphragm, hyperlucency in right upper zone, upper thoracic scoliosis and a normal cardiac silhouette

without interruptions in sleep. Airway examination revealed gross facial asymmetry with angle of mouth deviated towards left on smiling. Rest of the airway examination was unremarkable.

Blood investigations were within normal limits. ECG showed normal sinus rhythm. Chest X-ray (PA) revealed unequal number of ribs (7 on right & 11 ribs on left side), left upper ribs crowding, elevation of left hemidiaphragm, hyperlucency in right upper zone, upper thoracic scoliosis and a normal cardiac silhouette (Figure 3). 2D Echo reported normal study. CT Scan (face & PNS) revealed small sized right external ear with relatively small right maxilla and mandible and atrophy of right facial muscles.

2.1. Anesthetic plan & preparations

Written informed consent was taken from parents. Patient was pre-medicated with tab alprazolam (0.25mg) two hours pre-operatively. We planned for total intravenous anaesthesia (TIVA) with propofol, dexmedetomidine and non-depolarising muscle relaxants. Anesthesia machine was prepared by flushing for 2 hours with new Bain circuit attached and vaporizers removed. Airway cart along with emergency drugs including injection dantrolene sodium and provisions for bladder and gastric wash were kept ready.

Inside the operation theatre standard monitors were attached including ECG, NIBP, SpO2 and temperature probe. IV line was secured with 18G cannula and IV fentanyl 2mcg/kg injected. After pre-oxygenation with 100% oxygen anesthesia was induced with propofol 2mg/kg and muscle relaxation was achieved with vecuronium 0.1 mg/kg. Airway was easily secured with size 7 cuffed orotracheal tube that was connected to Bain circuit through heat and moisture exchanger (HME). Anesthesia was

maintained with N2O in O2 (70:30) at 10lpm along with infusions of dexmedetomidine 0.5mcg/kg/hour and propofol 80-100mcg/kg/hour, titrated to keep BIS value between 40-60. Fluid requirement was met with IV fluids given at room temperature. Analgesia was achieved with IV diclofenac, fentanyl supplements and paracetamol. Intercostal nerve block with bupivacaine (0.25%) was given for post-operative analgesia for the site of costal cartilage harvest for constructing the ear. Intraoperative period was uneventful. Anesthesia was reversed after completion of surgery. Post-operative period was uneventful, and the patient was discharged on 4^{th} post-operative day.

3. Discussion

Patients with PS require frequent cosmetic and corrective surgeries. Although the cause of PS is still unknown, the disruption of blood in the subclavian arteries and its branches during 6^{th} week of gestation is the prevailing hypothesis.[2] As the subclavian artery usually supplies blood to embryonic tissue that form chest wall and upper limb, the site of its disruption explains the range of defects.[3–5] Its occurrence is sporadic in nature with incidence of 1:30000 to 1:80000 in live births[6] with preponderance for males (2:1 to 3:1 as compared to females)[7,8] and the right side of the body (60% to 75%).

Patients with PS are susceptible to malignant hyperthermia as PS and malignant hyperthermia share the same genetic loci of chromosomes 5p,7q11.22.[9] Although there is no case reported wherein these patients have developed MH following general anesthesia even when inciting agents were used,[10] still it is advocated to avoid anesthetic agents which trigger MH, for instance depolarising muscle relaxant and inhalational anesthetic. This recommendation is based on the knowledge that the incidence of MH in normal population is 1 per 62000 when triggering agents are not used, 1 per 4500 when triggering agents are used and 1:2000 in MH susceptible patients, like these PS patients.[9]

Regional anesthesia if indicated should be the anesthesia of choice but was not applicable to our patient. For patients requiring general anesthesia, special preparation and execution are recommended. The recommendation is to flush the anesthesia machine to achieve volatile anesthetic concentration of less than 5 part per million (ppm), which may take 10 to 104 minutes with different machines.[9] Other measures include removing the vaporizers from anesthesia workstation, renewing the CO2 absorbent, using a new disposable breathing circuit and, if possible, a fresh gas hose. Fresh gas flow should be kept at least 10 litres per minute to avoid the rebound. Application of charcoal filters on inspiratory and expiratory limbs has been shown to accelerate the cleansing process, such filters should be replaced with a new set every 60 minutes.[9] Intraoperatively, stringent temperature and ETCO2 monitoring intraoperatively. Anesthesiologists should maintain constant vigil for signs of MH. Cold IV fluids should be readily available.

We adhered to most of these recommendations in our case. Anesthesia workstation was prepared by removing the vaporizers, changing the soda lime, using new Bain circuit with HME near the patient and flow rates of 10lpm. We flushed the circuit for two hours at 10lpm flow. Before beginning anesthesia we checked that the expired gas did not have any trace of any volatile anesthetic for five minutes at O2 flow of 5lpm, a surrogate for volatile anesthetic concentration of less than 5ppm, as our gas monitors do not give values in ppm. Ideally there should be a dedicated anesthesia machine for patient who is MH, but we did not have such provision. We used TIVA and non-depolarizing muscle relaxants only. Temperature and EtCO2 were monitored continuously and the alarm limits were set in a narrow range of 35 to 37 °C for temperature and 30 to 40 mmHg for EtCO2.

4. Conclusion

In patients of PS requiring general anesthesia adequate preparation must be made keeping in mind not only the anatomical changes but also the much higher possibility of MH in such patients.

5. Source of Funding

None.

6. Conflict of Interest

The authors declare no conflict of interest.

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