NATURALISTA CAMPANO
ISSN: 1827-7160
Volume 28 Issue 1, 2024

Anaesthetic Considerations in A Case of Parry Romberg Syndrome with Hemi Facial Atrophy Posted for Facial Remodeling Surgery

Dr. Anand. S[1*], Dr. Navaneetha Krishnan[2]

[1]Post Graduate, Department of Anesthesiology, Saveetha Medical College, Chennai, Tamilnadu, India
[2]Professor, Department of Anesthesiology, Saveetha Medical College, Chennai, Tamilnadu, India

Email: [1]dranandanesthesia@gmail.com, [2]navanee.priya@gmail.com

Corresponding author: Dr. Anand. S
[1*]Post Graduate, Department of Anesthesiology, Saveetha Medical College, Chennai, Tamilnadu, India

Abstract: Managing anesthesia in patients with Parry Romberg syndrome is challenging due to limited mouth opening, requiring careful planning and a deep understanding of the condition's pathophysiology. This report discusses anesthetic management for a 33-year-old female diagnosed as Parry Romberg Syndrome with hemi facial atrophy posted for surgical remodeling of face with ALT flap.
This case report highlights the necessity for a tailored anesthetic plan With the posibility to perform awake fiber-optic intubation due to patient age and cooperation level, a combination of general anesthesia with preserved spontaneous respiration was implemented. The report details the use of fiberoptic bronchoscopy for intubation, the intraoperative pharmacological strategy involving sevoflurane and propofol, and the postoperative care that resulted in improved mouth opening. The case underscores the importance of multidisciplinary collaboration and advances in surgical and anesthetic techniques for managing complex airways . The outcomes contribute to the literature on anesthetic approaches in such intricate scenarios, suggesting that with appropriate techniques, successful management is possible.

1. Introduction:

Parry Romberg Syndrome, also known as Progressive Hemi facial Atrophy, is a rare disease of unknown etiology . It is characterized by the progressive and self-limited painless loss of soft tissue and in some cases hard tissues of one side of the face, usually the left. The disease starts with the atrophy of the subcutaneous tissue with thinning of the dermis and darkening of the skin. When the onset is before the second decade, underlying muscle and bone will also be involved. This progresses for a period of 2–10 years and then enters a stable phase. The loss of soft and hard tissue leads to aesthetic and functional deficits which are compounded by the presence of associated symptoms like neuralgia, migraine, epilepsy and ocular involvement. The degree of deformity depends on the age at which the disease manifests first; the younger the age the more severe the deformity. These patients undergo severe psychological trauma and social problems.

Case report:
33-year old female presented with complaints of right hemi facial atrophy, drippling of saliva, restricted mouth opening diagnosed with PRS couple of years back, programmed for surgical remodeling of face with ALT flap. The patient is a known case of epilepsy on Sodium valporate. No relevant cardiac condition identified.

Pt was diagnosed with T2DM 1 year backwas on OHA's. INVESTIGATIONS : Hb 9.4, platelets 2.53, PT 11.8/INR 1.02 and aPTT 25.6,, CBG 184 mg/dl. 2D echo and ECG WNL/ Airway MMP couldn't be assessed due to restricted mouth opening of <1cm

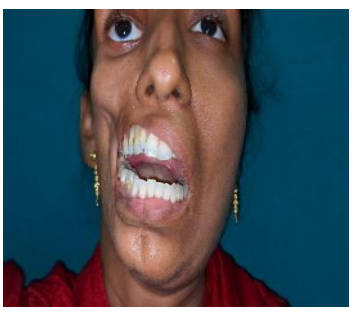
Pt came with Hb Level of 5.4 after which 2 units PRBC transfusion was done before being posted for surgery. Pre operative high risk consent and ICU bed with ventilator standby reserved in view of prolonged surgery.

PRE OP PREPARATION: Nasal pack with 2% lignocaine with adrenaline followed by 4% lignocaine nebulization done.

Pt was explained about awake Fiberoptic intubation procedure and consent obtained**.** The treatment goal in PRS is remodeling of the facial contour, minimizing any atrophy-related complications including a difficult airway

INTRA OPERATIVE EVENTS:

- Patient was shifted into the OR, routine monitors connected, two 16G venflon secured , under aseptic precautions intra arterial line for IBP monitoring and central line with right IJV secured
- Patient was pre oxygenated using nasal prongs with 4 liters oxygen .
- Followed by topical orotracheal anesthesia with 10% lignocaine solution spray, superior laryngeal nerve block and transtracheal nerve block was given. Dexmeditomidine infusion was started @1 mcg/kg.
- Awake FOB intubation was proceeded while the pt was being oxygenated through the opposite nostril & spray as you go techinique. After visualization of vocal cords 6.5 size flexometallic ETT was secured in place and position confirmed with capnography and five point auscultation.
- Pt induced with thiopentone and paralyzed with Atracurium as muscle relaxant. General anesthesia was maintained with inhalational isofluorane. Blood sugar was controlled by Alberti regime.
- Blood transfusion was started when blood loss was 100ml. Duration of surgery lasted for 9 and half hours. Four units PRBC,FFP and platelets transfused in ratio of 1:1:1. injection calcium gluconate administered. Intra op ABG was taken.
- After surgery pt shifted to ICU for observation where she tolerated CPAP with PS ventilation.
- Pt was extubated next day following T-piece trial and leak test performed.



2. Discussion:

- In case of an expected difficult intubation, airway blocks followed by nasotracheal intubation with a flexible fiberopticbronchoscope is an effective and safe technique which is considered as the gold standard.
- The phenotypical characterstics of PRS like severe hemifacial atrophy, dental and cranieo facial abnormalities in order to prevent any lesions during airway manipulation, as well as any nasal or oral telangiectasias (in cases associated with scleroderma) that may bleed during the procedure.
- PRS has been associated with hypertrophic cardiomyopathy. So an ECG and an echocardiography should be made prior to surgery, considering the administration of a perioperative beta blocker to optimize ventricular filling

3. Conclusion:

This poster highlights the importance of a continuous and careful pre-operative evaluation due to the progressive facial changes, proper difficult airway management, and the prevention of any potential cardiovascular and neurological complications that may arise, in order to provide a safe anesthetic technique to PRS patients. The role of FOB for excellent visualization of cords.

4. References:

1. Karkar AM, Hong T. Anesthesia management of a patient with Parry-Romberg Syndrome: a case report. J Anesth Clin Res. 2014;5:391. [Links]
2. 2. Nair PA, Patel NH, Diwan NG. Parry-Romberg Syndrome: a linear variant of scleroderma with discoid lupus erythematosus on scalp - an association. Int J Trichol. 2014;6:714. [Links]
3. 3. Kumar S, Kumar D, Kumar R, Rajkumar, Prabhakar R. Parry-Romberg Syndrome: a case of late onset with rapid progression. J Clin Diagn Res. 2014;8:27-8. [Links]
4. Lonchampt P, Emile J, Pélier-Cady MC, Cadou B, Barthelaix A. Central sympatheti dysregulation and immunological abnormalitie in a case of progressive facial hemiatrophy (Parry-Romberg disease). Clin Auton Res. 1995;5:199-204.