Journal of Anesthesia & Clinical Research
ISSN: 2155-6148

Karkar and Hong, J Anesth Clin Res 2014, 5:3
DOI: 10.4172/2155-6148.1000397

Case Report Open Access

Anesthesia Management of a Patient with Parry-Romberg Syndrome: A Case Report

Ami M Karkar and Tao Hong*

Department of Anesthesiology, Georgia Regents University, USA

Introduction

Parry-Romberg Syndrome (PRS) is a rare condition manifesting in severe progressive hemifacial atrophy involving skin, soft tissue, and bone [1]. It is often found in the first decade of life, usually in females. It can cause severe facial pain and is associated with other autoimmune disorders and inflammatory changes noted on MRI [2]. Although many have studied its etiology and the complexities of surgical grafting, intraoperative anesthetic management of the multiple systems that are affected by PRS has not been studied [3].

Case Presentation

A 51-year-old female with Parry-Romberg Syndrome diagnosed at age 5 years, scleroderma, hypertension, supraventricular tachycardia, sciatica, and history of multiple facial and cervical reconstructions presented with complaints of esthetic dissatisfaction, difficulty breathing through nose, difficulty swallowing, and occasional reflux. She also reported numbness, tingling, and burning on the right side, including head and neck area. Previous surgeries included two abdominal flaps to the head and neck, three nasal surgeries and two blepharoplasties of her right eye, cervical liposuction, right nasolabial liposuction, three caserean sections, and hysterectomy. She has been on gabapentin, amitriptyline, tizanadine, meloxicam, alprazolam, metoprolol and lisinopril.

The patient weighs 125 pounds and is 5'3" tall with notable sinus tachycardia and a generalized right-sided atrophy (Figure 1). She had a Mallampati I classified airway, bilateral narrow nostrils and philtrum with right septal deviation and right nares patency less than left (Figure 2). There were several facial scars. Maxilla and mandible were edentulous with right-sided arch hypotrophy (Figure 3). Her neck was supple with limited range of motion. Her lung and cardiovascular exam are normal except the tachycardia noted previously. Bony and muscular hypotrophy in the right leg and foot making it shorter (Figure 4), right foot drop, and sclerodermal lesions on the left leg were present. Cranial nerve V2-3 hypoesthesia with tingling and burning sensation and right frontal temporal bone weakness were also present.

Figure 1: Patient with notable sinus tachycardia and a generalized right-sided atrophy.

The patient was cleared by cardiology for surgery and placed on metoprolol for supraventricular tachycardia, which was controlled. Echocardiogram showed an ejection fraction of 51% with normal left ventricular function. In the operating room holding area, she received Oxymetazoline nasal spray in the left nostril for vasoconstriction and perioperative intravascular (IV) metoprolol.

Patient was intubated with a 6.0 mm nasal RAE tube through the left nostril using a Glidescope directed with a McGill's forceps with grade 1 view. Patient was placed in prone position for the posterior iliac graft harvest then in the supine position for the Leforte I maxillary osteotomy with down fracture and right mandible and maxillary graft placement. She received 4 liters of crystalloid and 1 liter of colloid during the nine-hour case. Tachycardia on emergence, after paralytic reversal, required 3 doses of 2 mg metoprolol boluses. The patient was discharged from the recovery room with no complications.

Discussion

PRS has multisystem effects with associated disorders, which need to be considered for careful airway management, hemodynamic stability, and good neurologic outcomes. Consideration should be given to potentially difficult airways, connective tissue, cardiovascular, and neurologic aspects of PRS. Furthermore, the slow progression of PRS over 2-10 years before it stabilizes requires frequent and careful pre-operative evaluation in order to appropriately plan for new manifestations [4].

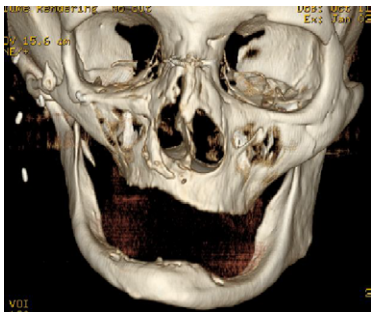

Figure 2: Mallampati I classified airway, bilateral narrow nostrils and philtrum with right septal deviation and right nares patency less than left.

***Corresponding author:** Tao Hong, Department of Anesthesiology, Gerogia Regents University, Augusta, Georgia 30912, USA, Tel: 570-854-7974; E-mail: thong@gru.edu

Received January 27, 2014; **Accepted** March 28, 2014; **Published** March 31, 2014

Citation: Karkar AM, Hong T (2014) Anesthesia Management of a Patient with Parry-Romberg Syndrome: A Case Report. J Anesth Clin Res 5: 397. doi:10.4172/2155-6148.1000397

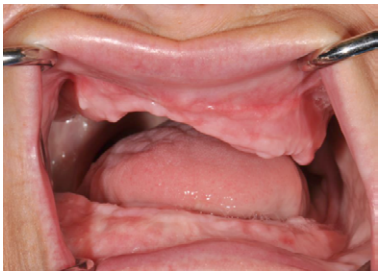

Figure 3: Maxilla and mandible were edentulous with right-sided arch hypotrophy.

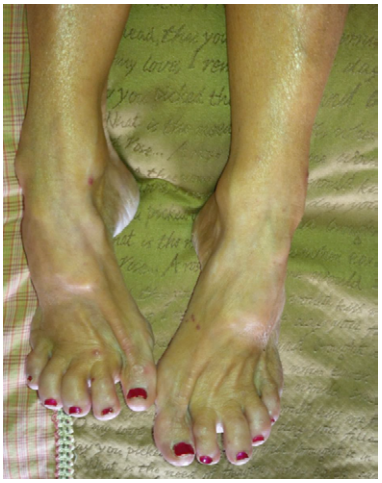

Figure 4: Bony and muscular hypotrophy in the right leg.

Preoperative Management and Anesthesia Induction

Patients with PRS have severe hemi facial atrophy and dental anomalies. There is often deviation of the mouth and nose toward the affected side [5]. Although external asymmetry and edentulism can be obvious, deviation of the tongue and uvula should be assessed prior to intubation. In younger patients, roots of teeth are often poorly developed or resorbed so careful diagnostic laryngoscopy is critical to prevent freeing any loose teeth. Also, dermatologic skin fibrosis and tense muscles should be evaluated during the neck exam. Glidescope and fiberoptic laryngoscopy should be available. As with scleroderma, oral and nasal telangiectasias may bleed profusely if traumatized during tracheal intubation. Preoperatively, the patient should receive Oxymetazoline nasal spray in the nostril planned to be nasally intubated to vasoconstrict the vessel and minimize traumatic bleeding. Furthermore, the relaxation of the lower esophageal sphincter makes these patients more susceptible to aspiration of gastric contents so preoperative antacids may be useful.

The cardiovascular system is often affected by an inflammatory process. The arterial line is recommended for those patients for better hemodynamic management. Arterial catheterization can be difficult as it may be in Raynaud's phenomenon and scleroderma from dermal thickening. Thus, ultrasound guidance may be helpful. Furthermore, hypertrophic cardiomyopathy (HOCM) has been associated with PRS so an electrocardiogram and echocardiogram should be performed preoperatively and preoperative beta-blocker administered to optimize ventricular filling [6].

PRS may be caused by trophic malfunction of the sympathetic system since normal development of skin, muscle, and bone requires trophic stimulation [7]. It is associated with neurologic disorders such as trigeminal neuralgia, facial paresthesia, headache and focal epilepsy [8]. Some patients with autonomic dysfunction have ipsilateral Horner's syndrome [6]. The trigeminal pathway has been indicated as the cause of the trigeminal neuralgia and paresthesia, likely from nerve compression by thickened connective tissues surrounding nerve sheaths. Only Falla et al. has studied this at a microscopic level and found that after examining biopsied intraepidermal nerve fibers, the disabling condition is not associated with trigeminal system damage but probably arises from musculoskeletal abnormalities [1]. Autonomic dysfunction should also be expected. Experimental studies involving unilateral sympathectomy have produced similar affects as with PRS [4]. Focal epilepsy is the most common neurologic manifestation corresponding to frontoparietal lesions found in the brain of PRS patients [9]. This is often diagnosed at a young age [10]. Thus, appropriate seizure prophylaxis is indicated.

Anesthesia Maintenance

Also as in scleroderma, decreased pulmonary compliance from diffuse interstitial pulmonary fibrosis may require increased positive airway pressure for adequate ventilation. These patients may also have decreased diffusion capacity, so it is important to make appropriate changes for adequate ventilation. With vascular changes, they often have some degree of pulmonary hypertension.

Cardiac arrest and refractory shock occurred in 10 of 69 reported cases in patients with hypertrophic cardiomyopathy. It is important to maintain blood pressure (preload and systemic vascular resistance) and heart rate to prevent worsening of the outflow obstruction by administering adequate crystalloid for a lengthy surgery as well as colloid as needed depending on blood loss. Avoid dehydration, tachycardia and increased myocardial contractility. As in scleroderma, these patients could have myocardial changes such as sclerosis of small coronary arteries and of the conduction system, which can lead to cardiac dysrhythmias and congestive heart failure. Therefore, dysrhythmias should be treated accordingly if they occur. Chronic hypertension is often commonly seen.

Neurovasculitis should be considered [7,11]. Some patients reveals vessel caliber changes within the symptomatic hemisphere in cerebral angiography [12,13]. There is a case report of cerebral subarachnoid hemorrhage [14]. Thus, hypertension should be avoided, but cerebral perfusion maintained.

Postoperative Management

An asymmetric upper airway can cause obstruction and difficult ventilation. We need to maintain patent airway to prevent atelectasis or negative pressure pulmonary edema. Additionally, tachycardia caused by pain or hypovolemia caused by blood loss or dehydration can worsen hypertrophic cardiomyopathy. Therefore, good pain control and maintaining intravascular volume are important [15]. Because the length of the surgery is long, electrolytes and glucose often are needed to be monitored to prevent arrhythmias and hypoglycemia.

Other auto-immune manifestations to consider are Hashimoto thyroiditis, Grave's disease, multiple sclerosis, autoimmune hemolytic anemia, lupus, and torticollis [16].

References

1. Falla M, Biasiotta A, Fabbrini G, Cruccu G, Truini A (2012) Cutaneous

innervation and trigeminal pathway function in a patient with facial pain associated with Parry-Romberg syndrome. J Headache Pain 13: 497-499.

2. Moseley BD, Burrus TM, Mason TG, Shin C (2010) Neurological picture. Contralateral cutaneous and MRI findings in a patient with Parry-Romberg syndrome. J Neurol Neurosurg Psychiatry 81: 1400-1401.
3. Liu CC, Chang CS, Wu RS (2005) Anesthetic approach to the Dyke-Davidoff-Masson syndrome--a case report. Acta Anaesthesiol Taiwan 43: 55-58.
4. Mazzeo N, Fisher JG, Mayer MH, Mathieu GP (1995) Progressive hemifacial atrophy (Parry-Romberg syndrome). Case report. Oral Surg Oral Med Oral Pathol Oral Radiol Endod 79: 30-35.
5. Madasamy R, Jayanandan M, Adhavan UR, Gopalakrishnan S, Mahendra L (2012) Parry Romberg syndrome: A case report and discussion. J Oral Maxillofac Pathol 16: 406-410.
6. Behera M, Nimkhedkar K, Gupta RS, Hassadi MF, Mousa ME (1988) Facial hemiatrophy (Parry-Romberg syndrome) and hypertrophic cardiomyopathy. J Assoc Physicians India 36: 394-395.
7. Blitstein MK, Tung GA (2007) MRI of cerebral microhemorrhages. AJR Am J Roentgenol 189: 720-725.
8. Buonaccorsi S, Leonardi A, Covelli E, Indrizzi E, Perdicchi A, et al. (2005) Parry-Romberg syndrome. J Craniofac Surg 16: 1132-1135.
9. Yano T, Sawaishi Y, Toyono M, Takaku I, Takada G (2000) Progressive facial hemiatrophy after epileptic seizures. Pediatr Neurol 23: 164-166.
10. Aktekin B, Oguz Y, Aydin H, Senol U (2005) Cortical silent period in a patient with focal epilepsy and Parry-Romberg syndrome. Epilepsy Behav 6: 270-273.
11. Auvinet C, Glacet-Bernard A, Coscas G, Cornelis P, Cadot M, et al. (1989) [Parry-Romberg progressive facial hemiatrophy and localized scleroderma. Nosologic and pathogenic problems]. J Fr Ophtalmol 12: 169-173.
12. Moko SB, Mistry Y, Blandin de Chalain TM (2003) Parry-Romberg syndrome: intracranial MRI appearances. J Craniomaxillofac Surg 31: 321-324.
13. Woolfenden AR, Tong DC, Norbash AM, Albers GW (1998) Progressive facial hemiatrophy: abnormality of intracranial vasculature. Neurology 50: 1915-1917.
14. Kuechler D, Kaliaperumal C, Hassan A, Fanning N, Wyse G, et al. (2011) Aneurysmal subarachnoid haemorrhage in Parry-Rhomberg syndrome. BMJ Case Rep 2011.
15. Chang KH, Sano E, Saitoh Y, Hanaoka K (2004) [Anesthetic management of patients with hypertrophic obstructive cardiomyopathy undergoing non-cardiac surgery]. Masui 53: 934-942.
16. El-Kehdy J, Abbas O, Rubeiz N (2012) A review of Parry-Romberg syndrome. J Am Acad Dermatol 67: 769-784.