



Anesthetic Considerations of Patient with Parry Romberg Syndrome

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ABSTRACT

Parry Romberg syndrome is a rare progressive degenerative disease characterized by unilateral atrophy affecting the skin, connective tissue, muscle, and bone, typically occurs in children and young adults. The end result is facial asymmetry associated with other skin, dental, visual, cardiovascular, and neurological disorders that ceases without apparent cause after a highly variable period. Inconsistency in the pattern of atrophy and multisystem involvement make intraoperative anesthetic management of these patients a challenge for anesthesiologists. We present a case of Parry Romberg syndrome and her associated clinical findings with specific attention to the anesthetic consideration of this disease.

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Introduction

Parry Romberg syndrome (PRS), or progressive facial hemiatrophy (PHA), was first described by Caleb Hillier Parry and Moritz Heinrich Romberg in 1825 and 1846, respectively (1, 2).

It is a degenerative disease characterized by unilateral atrophy of body with variable involvement of the skin and subcutaneous tissue, bone, muscles, and also connective tissue. It may occasionally extend to the neck

and sometimes the body. The disease usually begins with slow progression during the early decades of life until it stabilizes. The end result is facial asymmetry, which may present associated with other skin, dental, visual, and neurological disorders. It usually involves the left side of the face, and is more frequent in women; the prevalence is estimated to be at least 1/700,000 individuals (3-7).

The definitive etiology is not known yet, and many theories about PRS have emerged

throughout the years, such as sympathetic nervous system dysfunction, trauma, infection, and inflammatory conditions. The most recent and reliable theory is a genetic alteration during the embryogenesis of the central nervous system, together with cerebral sympathetic nervous system hyperactivity with autoimmune origin (7-9).

Case Report

A 50-year-old woman with PRS, diagnosed at the age of 14 years by progressive hemifacial atrophy, presented with complaints of esthetic dissatisfaction programmed for autologous facial fat injection. Past surgical history was two craniofacial reconstruction procedures twenty years ago, due to bone atrophy, and two cesarean sections under general anesthesia. She had no drug history. The patient was 75 kg in weight and 170 cm in height. She had no history of cardiac and respiratory disturbances, and was cleared by cardiology for surgery. The patient had obvious thinning of the subcutaneous skin and fat on the left side of face and neck. In neurologic examination, left eye ptosis and mydriasis with visual disturbance were detected, and was cleared by ophthalmologist with no intracranial tumor. The airway evaluation resulted in Mallampati of grade I, a 6-cm thyromental distance, a 5-cm interdental distance, and left sided atrophy of the tongue and uvula; no mobile teeth were identified (Figures 1 and 2).



Figure 1. Patient with atrophy of skin and subcutaneous fat predominantly on the left side of the face and tongue



Figure 2. Airway evaluation revealed mallampati class I and obvious atrophy of uvula and left side of the tongue

Reviewing past anesthesia records, a Cormack-Lehane II classification was established. She had a right narrow nostril and right nasal septal deviation. She had also a focal scleroderma on her left side low back region (Figure 3).

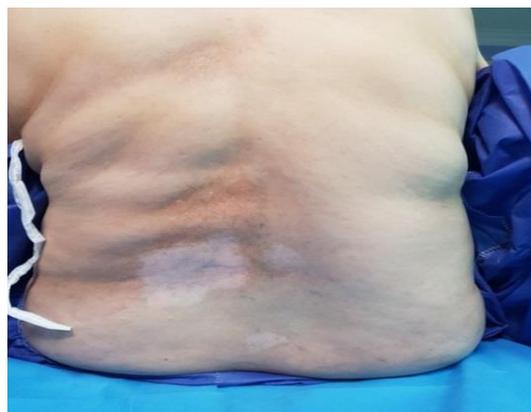


Figure 3. Focal scleroderma with telangiectasia on left side of low back region

On arrival the operating room, the patient was under standard non-invasive blood pressure monitoring, pulse oximeter, and electrocardiography. A peripheral venous catheter was placed, and the patient was oxygenated by simple face mask. The anesthetic technique used was intravenous (IV) sedation with 100 microgram fentanyl, 2 mg midazolam, and IV infusion of 25 microgram/kg/minute propofol. Patient was placed in supine position for abdominal liposuction. After tumescent anesthesia of abdomen by surgeon with subcutaneous infiltration of tumescent fluid containing lidocaine 0.1%, saline, and epinephrine

(1:1,000,000), the procedure of facial fat transfer began, and the surgery was completed after three hours. She received three liters of crystalloid during the surgery. There was no problem during anesthesia and surgery, and she was discharged from the recovery room with no complications. Postoperative pain relief was achieved by intravenous pethidine in the ward, and oral nonsteroidal anti-inflammatory drugs (NSAIDs) at home.

Discussion

PRS is a disease with associated disorders involving many systems with slow progression over two to ten years before it stabilizes that requires frequent and careful preoperative evaluation in order to appropriately plan for new manifestations. Consideration should be given to potentially difficult airways, connective tissue, cardiovascular, and neurologic aspects of PRS.

Patients with PRS have severe hemifacial atrophy and dental anomalies. There is often deviation of the mouth and nose toward the affected side. 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. Moreover, dermatologic skin fibrosis and tense muscles should be evaluated during the neck examination. Glidescope and fiberoptic laryngoscopy should be available. As with scleroderma, oral and nasal telangiectasia 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 should be administered to optimize ventricular filling (5, 6).

PRS may be caused by trophic malfunction of the sympathetic system; since normal development of skin, muscle, and bone requires trophic stimulation. It is associated with neurologic disorders such as trigeminal neuralgia, facial paresthesia, headache, and focal epilepsy. Some patients with autonomic dysfunction have ipsilateral Horner's syndrome. 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. Assessment of this at a microscopic level showed that after examining biopsied intra-epidermal nerve fibers, the disabling condition is not associated with trigeminal system damage; but probably arises from musculoskeletal abnormalities (4-7). Autonomic dysfunction should also be expected. Experimental studies involving unilateral sympathectomy have produced similar affects as with PRS. Focal epilepsy is the most common neurologic manifestation corresponding to frontoparietal lesions found in the brain of patients with PRS. This is often diagnosed at a young age. Thus, appropriate seizure prophylaxis is indicated (4-8).

Although our case was diagnosed previously at age 14, many differences and similarities are between our patient and other syndromes that should be included for differential diagnosis.

PRS shares many clinical features, such as age of onset, associated neurologic

symptoms, and cutaneous presentations with en coup de sabre. It is a linear scleroderma that usually involves the anterior segment of the scalp. These two diseases have some overlapping features; for example, some patients with PRS have typical scleroderma localized outside the face. Deeper head and neck structures, such as the oral or pharyngeal musculature, are not commonly involved in en coup de sabre. The elevated titers of autoantibodies in en coup de sabre is typical; that is not elevated in PRS (9, 10).

Other diseases in which facial asymmetry is prominent clinical features include Barraque-Simons syndrome and hemifacial microsomia (or first and second brachial arch syndrome); but unlike PRS, these conditions are typically congenital and nonprogressive (11).

Hemifacial hyperplasia causes asymmetry and overgrowth, but not atrophy of the face. Partial lipodystrophy (Barraque-Simons syndrome) has bilateral manifestations rather than unilateral manifestation that is common in PRS (11, 12).

Silent sinus syndrome may also be considered, but these patients present later in life with imaging findings of maxillary sinus atelectasis and evidence of ostiomeatal obstruction at the same side (13).

Postoperative considerations: An asymmetric upper airway can cause obstruction and difficult ventilation that causes atelectasis or negative pressure pulmonary edema. Additionally, tachycardia caused by pain or hypovolemia caused by blood loss or dehydration can worsen hypertrophic cardiomyopathy in patients with cardiac involvement. Therefore, good pain control and maintaining intravascular volume are important (6).

Conflict of Interests

Authors have no conflict of interests.

Acknowledgments

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