REV COLOMB ANESTESIOL. 2016;xxx(xx):xxx–xxx

Revista Colombiana de Anestesiología
Colombian Journal of Anesthesiology
www.revcolanest.com.co

Case report

Anesthetic implications of Parry Romberg Syndrome: A case report☆

Guiomar Fernández-Castellano[a,*], *Rosana Guerrero-Domínguez*[b], *Daniel López-Herrera-Rodríguez*[b], *Ignacio Jiménez*[c]

[a] *MD Internist, Resident of Anesthesiology and Resuscitation, Hospitales Universitarios Virgen del Rocío, Seville, Spain*
[b] *Specialist in Anesthesiology and Resuscitation, Hospitales Universitarios Virgen del Rocío, Seville, Spain*
[c] *Director of the Unidad de Gestión Clínica de Anestesiología y Reanimación Hospital de Traumatología y Rehabilitación, Hospitales Universitarios Virgen del Rocío, Seville, Spain*

ARTICLE INFO

Article history:
Received 27 June 2015
Accepted 6 April 2016
Available online xxx

Keywords:
Laryngoscopy
Anesthesia
Airway management
Facial hemiatrophy
Craniofacial dysostosis

ABSTRACT

Introduction: Parry–Romberg Syndrome is a rare degenerative disease characterized by unilateral atrophy affecting the skin, connective tissue, muscle and bone. The end result is facial asymmetry associated with other skin, dental, visual, cardiovascular, and neurological disorders.

Clinical findings, diagnostic evaluation and interventions: The case of a patient with Parry–Romberg Syndrome programmed for frontonasal flap remodeling is discussed. The patient's history includes trigeminal neuralgia, epilepsy, and two previous surgical interventions. Uneventful endotracheal intubation with the Glideoscope® video laryngoscope was performed, upon adequate pre-oxygenation followed by anesthetic induction.

Conclusion: The phenotypical characteristics of Parry Romberg Syndrome are severe facial hemiatrophy and craniofacial anomalies that require careful preoperative evaluation and management of a potentially difficult airway. Consequently, the use of video laryngoscopes is a first-line approach. Due to the syndrome's associated disorders, it is essential to maintain hemodynamic stability and prevent any potential seizures.

☆ Please cite this article as: Fernández-Castellano G, Guerrero-Domínguez R, López-Herrera-Rodríguez D, Jiménez I. Implicaciones anestésicas del Síndrome de Parry-Romberg; reporte de un caso. Rev Colomb Anestesiol. 2016. http://dx.doi.org/10.1016/j.rca.2016.04.004

* *Corresponding author at*: Calle Genaro Parladé, N°3, 4°D, 41013, Seville, Spain.
E-mail address: guio012@hotmail.com (G. Fernández-Castellano).

2256-2087/

Implicaciones anestésicas del Síndrome de Parry-Romberg; reporte de un caso

Palabras clave:
Laringoscopía
Anestesia
Manejo de la vía aérea
Hemiatrofia facial
Disostosis craneofacil

RESUMEN

Introducción: El Síndrome de Parry-Romberg es una enfermedad degenerativa poco común, caracterizada por una atrofia unilateral que afecta la piel, el tejido conjuntivo, el músculo y el hueso. El resultado final es una asimetría facial cursando con otras alteraciones cutáneas, dentales, oculares, cardiovasculares y neurológicas.

Hallazgos clínicos, evaluación diagnóstica e intervenciones: Presentamos un caso de un paciente con Síndrome de Parry-Romberg programado para remodelación de colgajo frontonasal. Entre sus antecedentes destacan neuralgia del trigémino, epilepsia y dos intervenciones quirúrgicas previas. Tras una adecuada preoxigenación y posterior inducción anestésica, se realiza una intubación endotraqueal sin incidencias mediante el videolaringoscopio Glideoscope®.

Conclusión: El Síndrome de Parry Romberg presenta como características fenotípicas hemiatrofia facial grave y anomalías craneofaciales, que requieren una cuidadosa evaluación preoperatoria y el manejo de una vía aérea potencialmente difícil. Es por esto que los videolaringoscopios resultan una alternativa de primera línea. Debido a sus trastornos asociados, es esencial mantener la estabilidad hemodinámica y la prevención de posibles crisis convulsivas.

Introduction

Parry–Romberg Syndrome (PRS), also known as progressive facial hemiatrophy, is a rare degenerative condition characterized by unilateral atrophy with variable involvement of the skin, the subcutaneous tissue, the muscles, the connective tissue and the bones.[1,2] It may occasionally extend to the neck and even the body as a whole.[3] Typically, the onset of the atrophy is during the early decades of life,[1,3] with a slow progression until the disease stabilizes.[3] The end result is facial asymmetry, which may present associated with other skin, dental, visual, and neurological disorders. It is usually more frequent in females and generally involves the left side of the face.[2] The incidence and precise etiology is jet unknown,[2,3] although the most recent and reliable theory is a genetic alteration during the embryogenesis of the central nervous system,[2] together with cerebral sympathetic nervous system hyperactivity, probably of autoimmune origin.[4]

The treatment goal in PRS is remodeling of the facial contour, minimizing any atrophy-related complications,[2,3] including a difficult airway. We would like to discuss the case of a PRS patient with difficult airway predictors and a successful orotracheal intubation using the Glideoscope® video laryngoscope (VL).

Clinical case

33-year old patient diagnosed with PRS since childhood, programmed for surgical remodeling of a frontonasal flap (Fig. 1). The patient's history includes trigeminal neuralgia and epilepsy treated with carbamazepine and valproic acid. No relevant cardiac has been identified and the patient underwent two facial reconstruction procedures ten years ago.

Clinical findings, diagnostic evaluation and interventions

A comprehensive physical examination was performed during the pre-anesthesia visit and complementary tests were reviewed. The findings include evidence of a marked facial asymmetry, atrophy of the left side of the face, left alar retraction, nasal septum deviation, hypoplasia of the frontal, malar, and maxillary bones and of the left hemimandible. There is also evidence of a temporal bone and cheek overgrowth resulting from the hyper development of the temporal muscle, the masseter and the left parotid gland (Fig. 2). The patient has an eye prosthesis (following the loss of his left eye due to endophthalmus) and his bodyweight is 80 kg.

The airway evaluation resulted in Mallampati grade III, a 5 cm thyromental distance, and a 4 cm interdental distance, deviation of the tongue and uvula and discrete limitation of the cervical extension. No mobile teeth were identified. Reviewing past anesthesia records, a Cormack–Lehane IIb classification was established that required intubation with the Eschmann guide.

When admitted to the OR, the patient was under standard non-invasive blood pressure monitoring, pulse oximeter, and electrocardiography. A peripheral venous catheter was placed and the patient was properly pre-oxygenated. The anesthetic technique used was IV induction with 150 μg of fentanyl and 180 mg of Propofol. There was no airway obstruction during manual face mask ventilation. Using the Glideoscope® VL proper visualization of the glottis was obtained, prior to administering a neuromuscular block (Rocuronium 50 mg) for uneventful orotracheal intubation at the first attempt. Upon