

Parry–Romberg syndrome affecting one half of the body

Jugajyoti Pathi, Pallavi Mishra¹, Harish Kumar¹, Abikshyeet Panda¹

Departments of Oral and Maxillofacial Surgery and ¹Oral and Maxillofacial Pathology, Kalinga Institute of Dental Sciences, KIIT, Bhubaneswar, Odisha, India

Corresponding author (email: <abikshyeet@yahoo.com>)

Dr. Abikshyeet Panda, Department of Oral and Maxillofacial Pathology, Campus 5, Kalinga Institute of Dental sciences, KIIT, Bhubaneswar, Odisha, India.

Received: 14-04-16

Accepted: 21-06-16

Published: 25-07-16

Abstract

Parry-Romberg syndrome, which is also known as progressive hemifacial atrophy, is a poorly understood rare condition. In this condition, the face shows unilateral, slowly progressive atrophy. Disturbance in fat metabolism, viral infection, trauma, heredity, endocrinal disturbances, and autoimmunity are few possible factors in its pathogenesis. Rarely, only this syndrome progresses and involves one half of the body. Our attempt is to present a case of Parry–Romberg syndrome involving one half of the body, which is a rarity in itself.

Key words: *Coup de Sabre, Parry–Romberg syndrome, progressive hemifacial atrophy*

INTRODUCTION

Parry–Romberg syndrome is a condition characterized by a slowly progressive shrinkage of the soft tissues, and sometimes bones of the face, unilaterally and occasionally bilaterally. Atrophy of the facial tissues including muscles, bones, and skin of one side are usually noticed in this condition.^[1] This condition, otherwise known as progressive hemifacial atrophy, as labelled by Eulenberg (1871), is very rare and was first described by Caleb Parry in 1815 followed by Maritz Romberg in 1846.^[2-4] This condition results in several aesthetic, functional, and psychological problems when the symmetry of the face is lost. The incidence and cause of this condition is unknown, however, a cerebral disturbance on fat metabolism is blamed. Trauma, viral infection, endocrine disturbances, autoimmunity, and heredity have also

been implicated in the pathogenesis.^[5] It manifests in the first and second decade of life, beginning as atrophy of the skin and subcutaneous structure of the face, and progresses at a variable rate affecting the dermatomes of one or more branches of the trigeminal nerve, and then becomes stable. Hypoplasia of the underlying bone may also occur if the condition starts during the first decade.^[5,6] Other associated conditions are ocular changes, neurologic disturbances, such as focal epileptiform seizures, and trigeminal neuralgia.^[7] Many times the affected skin shows discolouration due to hypopigmentation or hyperpigmentation before the atrophy sets in. Patchy alopecia may also be observed.^[8] Bilateral facial atrophy or unilateral involvements of entire body are rare presentations. There is a high incidence noticed in females than males.^[6] Here, we present a case of Parry–Romberg syndrome with unilateral progressive atrophy of one

Access this article online	
Quick Response Code:	Website: www.jispcd.org
	DOI: 10.4103/2231-0762.186792

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How to cite this article: Pathi J, Mishra P, Kumar H, Panda A. Parry–Romberg syndrome affecting one half of the body. J Int Soc Prevent Communit Dent 2016;6:387-90.

side of the face along with unilateral involvement of the entire body which is rarely encountered.

CASE REPORT

A 21-year-old male patient reported to the outpatient department of Kalinga Institute of Dental Sciences with a complaint of progressive deformity of the right side of the face since last 9 years. On examination, the patient had asymmetry of face on the right side with marked hypoplasia and hyperpigmentation of the skin. Lip was deviated to the right side. Right eyebrow showed loss of hair [Figure 1]. There was a linear scar (coup de sabre) extending from the forehead to mandibular menton dividing the affected side from the unaffected. On intraoral examination, right side soft palate and faucial area showed atrophy [Figure 2]. Right buccal mucosa was stiff to palpate. General examination revealed that the entire right side of the body was atrophied when compared with the left side [Figure 3]. Cone beam computed tomography (CBCT) three-dimensional (3D) reconstruction showed decreased vertical height of the right side mandible compared to the left side [Figure 4]. Clinical and radiographic findings led to a diagnosis of Parry–Romberg syndrome. Surgical intervention and alloplastic fat graft was suggested to the patient but he refused any such intervention due to financial constraints. The patient was recalled after 6 months. There was no further progression of the disease.

DISCUSSION

Parry–Romberg syndrome is a degenerative condition which is uncommon in nature. A slowly progressive unilateral atrophy involving the facial tissue, including muscles bones and skin, is characteristic of this

condition. The etiology and incidence are unknown. The primary cause is proposed to be a cerebral disturbance of fat metabolism. Viral infection, trauma, endocrine imbalance, autoimmunity, and heredity are considered to be pathogenetic.^[8,9] The condition has a high female predilection and presents mostly on the left side of the face.^[4,9,10] However, in our case the patient was male and had presentation on the right side. The skin can be dry and can show pigmentation. A demarcation line can be present between the normal and abnormal skin, mimicking a big linear scar called coup de sabre.^[4,11,12] The present case also showed a similar hyperpigmentation and coup de sabre. Enophthalmous is the most common ocular manifestation resultant of fat loss around the orbit. Other common complications are eyelid atrophy, uveitis, retinal vasculitis, oculomotor defects, and glaucoma.^[12] Some patients present with smaller ear on the affected side due to atrophy.^[11] The present case did not present any ocular manifestations or ear anomalies. Alopecia is also reported on the affected side, as seen in the present case on the right parietal region and right eyebrow.^[9] Cases may be associated with neurological complications such as trigeminal neuralgia, movement disorders secondary to brain lesions, hemiplegic migraines, facial palsy, headache, epilepsy, behavioral changes, sympathetic hyperactivity, progressive intellectual deterioration due to cerebral hemiatrophy (Rasmussen encephalitis), and oculomotor nerve palsy.^[5,13,14] The present case did not show any such complications. Frank elevation of the mouth, nose, facial midline, and dental midline is characteristic. Upper lip atrophy leads to exposure of maxillary anterior teeth. There can be atrophy of the tongue.^[5] The present case showed deviation of mouth and nose to the affected side.



Figure 1: Facial profile showing marked hypoplasia of the right side of the face resulting in facial asymmetry



Figure 2: Intraoral image showing atrophy of the right side soft palate and faucial area

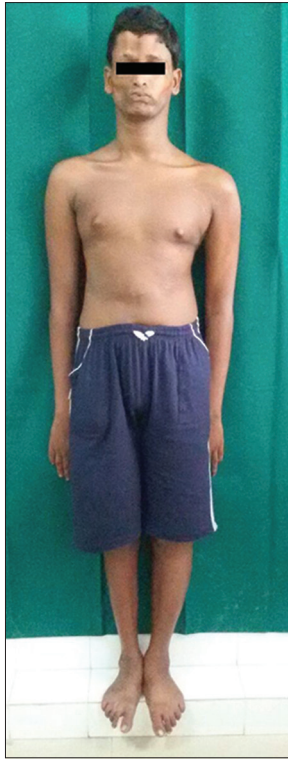


Figure 3: Right side of the body shows atrophy in comparison to the left side

Hemifacial microsomia (first and second branchial arch syndrome) and its variants such as Goldenhar syndrome, post-traumatic atrophy, and partial lipodystrophy (Barraquer-Simon Syndrome) can be considered in the differential diagnosis.^[7] Treatment is limited and unsatisfactory. Transfer of autologous fat tissue to the affected side, silicon injection, inorganic implants, and bovine collagen are few modalities that have been explored.^[15,16] However, these methods can never completely restore the appearance of the patient. Our patient was suggested a surgical alloplastic fat graft but he refused the treatment.

There is a controversial relationship between Parry–Romberg syndrome and localized scleroderma. The term Parry–Romberg syndrome is advocated to be used for progressive hemifacial atrophy without features of cutaneous scleroderma. However, patients with Parry–Romberg syndrome often report cutaneous changes. Hence, it can be suggested that localized scleroderma and Parry–Romberg syndrome may represent the same disease process.^[7]

CONCLUSION

Parry–Romberg syndrome or progressive hemifacial atrophy is an uncommon condition whose pathophysiology is poorly understood. Involvement



Figure 4: Cone beam computed tomography image showing decreased vertical height of the right side mandible

of one side of the body is still a rarity, which is seldom ever encountered. Such a rare case with involvement of entire half of the body is reported in the present case. Treatment of such cases becomes a challenge as we do not have treatment options to offer. Poorly understood conditions like this should be highlighted in the literature for enhancing the clinical acumen.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

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