Journal of Advanced Medical and Dental Sciences Research

@Society of Scientific Research and Studies

Journal home page: www.jamdsr.com doi: 10.21276/jamdsr UGC approved journal no. 63854

(e) ISSN Online: 2321-9599; (p) ISSN Print: 2348-6805

CASE REPORT

Parry Romberg Syndrome: A Rare Case Report

Neha Verma[1], Swati Verma[2]

[1]Senior Resident, Unit Of Oral Medicine And Radiology, Faculty Of Dental Sciences, Institute Of Medical Sciences, Banaras Hindu University, Varanasi, India
[2] Senior Resident, Division Of Orthodontics and Dentofacial Deformities, Centre For Dental Education and Research, All India Institute Of Medical Sciences, Delhi, India

ABSTRACT:
Parry Romberg syndrome is a rare disorder leading to facial asymmetry. It affects subcutaneous tissue and fat resulting in their atrophy. Many etiologies have been proposed but it remains unclear. Apart from facial asymmetry it also involves nerves of the affected dermatome leading to neurological disorders. The condition is self-limiting and stabilizes after second decade , hence no treatment is required. However it is important to known the features of this syndrome for prompt diagnosis and management of these patients.
Key words: Hemifacial atrophy, Parry Romberg Syndrome.

Received: 8 February, 2019 Revised: 27 February, 2019 Accepted: 28 February, 2019

Corresponding author: Dr. Neha Verma, Senior Resident, Unit Of Oral Medicine And Radiology, Faculty Of Dental Sciences, Institute Of Medical Sciences, Banaras Hindu University, Varanasi, India

This article may be cited as: Verma N, Verma S. Parry Romberg Syndrome: A Rare Case Report. J Adv Med Dent Scie Res 2019;7(4): 23-25.

INTRODUCTION

Parry Romberg syndrome also known as Progressive Hemifacial Atrophy is a rare disorder causing facial and body hemi atrophy. it was first described in 1825 by Caleb Hillier Parry and later by Moritz Heinrich Romberg in 1846. In 1871 Eulenberg termed it as progressive facial hemiatrophy.[1] the disorder is not congenital but it progresses in first two decades of individual's life. It is unilateral, cause's atrophy of subcutaneous tissue and fat leading to deformity of face, limbs and trunk. It has also been associated with neurological disorders like neuralgia, epilepsy, paresthesia and headache. Though etiology remains unclear, chronic vascular disturbance or neurological pathosis leading to inflammatory process is the , proposed etiology. However genetic, autoimmunity, trauma, endocrine disorders, metabolic disorders, Lyme's disease and Borrelia Burgdorferi infection are the other causes for the condition.[2]Though it is generally self-limiting, but the deformities caused are enduring .[3]

Here we present case of a female pediatric patient who presented with the features of syndrome and was not associated with any neurological condition.

CASE REPORT

A 8 year old female patient came to department for her dental evaluation. She gave history of "shrinking" of left side of face and body since three years of age which has progressed to its present form. She reported difficulty in maintaining her balance since one year. She gave no history of such illness in family or any preceding trauma. There was no history of other systemic diseases, ocular or neurological involvement. On extraoral examination the left side of head was depressed, there was focal baldness with a scar in mid frontal region of forehead (coup de sabre).There was facial asymmetry on left side, right side of face was fuller as compared to left, left eye appeared shrunken due to periorbital loss of fat, the left malar prominence was depressed , and the lip and mouth were stretched to the left side of the face. [fg.1]

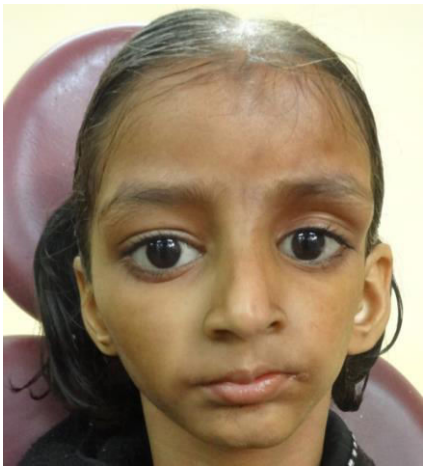

Figure 1 Extraoral Picture Showing Facial Hemiatrophy and Scar (Coup De Sabre) in Mid Forehead Region.

She had difficulty in standing without support, her left leg was thinner than the right.[fig.2]

Figure 2 Picture Showing Asymmetry in Lower Limbs

On intraoral examination she had mixed dentition with crowding, glossitis and angular chelitis.[fig.3]

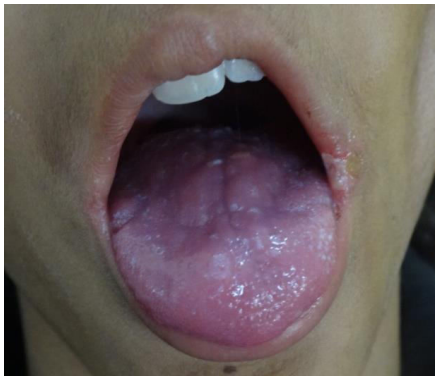

Figure 3 Intraoral Picture Showing Depapillation of Tongue and Angular Chelitis

On radiographic examination hypoplasia of left mandible and condyle were seen.[fig.4]

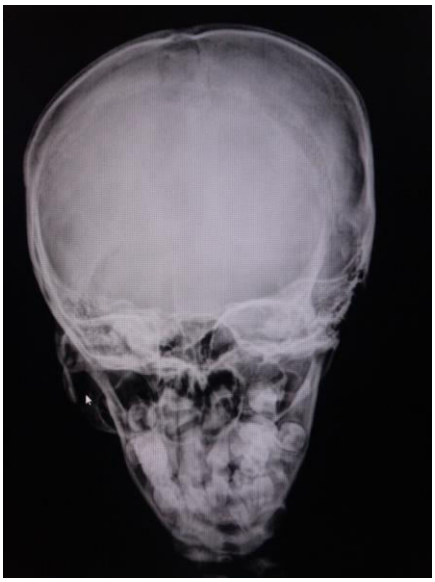

Figure 4 Anterioposterior Skull View Showing Hypoplasia of Left Mandible and Condyle.

Based on above clinical and radiographic findings she was diagnosed as having Parry Romberg syndrome and was referred to a Pediatric consultant.

DISCUSSION

Parry Romberg syndrome is a rare condition causing unilateral hemi atrophy of face, trunk and limbs. it is mostly seen affecting female population.[3] It commonly affects face, but in our case there was involvement left limbs as well. The skin of affected side is tensed like that in scleroderma with altered pigmentation, patches of hair loss on scalp, loss of peri orbital fat around affected eye, deviation of nose and mouth to the affected side is reported.[4] reported atrophy of lingual papillae.[5]These all features were present in our case, however there was no neurological involvement as is reported in many cases.[6]

The differential diagnosis includes Hemifacial Microsomia, Goldenhar syndrome which are congenital and non-progressive conditions and Barraquer-Simon Syndrome which is bilateral and involves the adipose tissue.[5]

This condition is self-limiting and usually ceases after second decade of life. Though corticosteroids and immunomodulators have played role is constraining the progression. As the deformities are permanent in nature, reconstruction and orthodontic procedures are helpful in restoring function and esthetics post stabilization.[3]

CONCLUSION

Parry Romberg syndrome is a disfiguring condition with associated loss of function. Its prompt diagnosis and

multidisciplinary approach in treatment can lead to better quality of life in these patients.

REFERENCES

1. Regezi JA, Scuibba JJ, Jordan RC. Clinical pathological correlations. 4th ed. USA: Saunders: Elsevier Science; 2003. Oral pathology.
2. Kumar AA, Kumar RA, Shantha GP, Aloogopinathan G. Progressive hemi facial atrophy - Parry Romberg syndrome presenting as severe facial pain in a young man: A case report. Cases J. 2009;2:6776.
3. Madasamy R, Jayanandan M, Adhavan UR, Gopalakrishnan S, Mahendra L. Parry Romberg syndrome: A case report and discussion. J Oral Maxillofac Pathol. 2012;16(3):406–410. doi:10.4103/0973-029X.102498
4. Ong K, Billson FA, Pathirana DS, Clifton Bligh P. A case of progressive hemifacial atrophy with uveitis and retinal vasculitis. Aust N Z J Ophthalmol. 1991;19:295–8.
5. Deshingkar SA, Barpande SR, Bhavthankar JD, Humbe JG. Progressive hemifacial atrophy (Parry-Romberg Syndrome). Contemp Clin Dent. 2012;3(Suppl 1):S78–S81. doi:10.4103/0976-237X.95111
6. Padmavathy J, Rao LL. Unilateral linear pansclerotic morphea affecting face and limbs. Indian J Dermatol Venereol Leprol. 2005;71:192–4.

Source of support: Nil **Conflict of interest:** None declared