

Parry Romberg Syndrome with its Oral Manifestations- Report of a Case

Arun George, Sankar Vinod Vichattu, Joy R Das, Ektah Khosla, Jinu Elizabeth James, Sarah Paul

ABSTRACT

Introduction: Parry Romberg syndrome (PRS) which is also called as progressive facial hemiatrophy is a development craniofacial anomaly. It is a very rare condition that slowly and progressively affects one side of the face as well as one side of the body with loss of soft and hard tissues

Case presentation: A 47 year old female reported to our Department of Oral and Maxillofacial Surgery with a chief complaint of pain in her lower left back tooth region and front tooth region since two weeks. On examination it was typical to classic Parry Romberg Syndrome.

Management and prognosis: Osteomyelitis was addressed with curettage and sequestrectomy of the affected site with antibiotic therapy. Treatment largely is based on cosmetic corrections as the patient exhibits immense psychological trauma as this happens over the course of the patients life.

Conclusion: The etiology is unknown although certain resources mention its relation to autoimmune conditions. It is associated with different systemic manifestations including craniofacial, ophthalmic and neurologic abnormalities. A multidisciplinary approach is mandated in such patients to enhance their quality of life.

Keywords: Parry Romberg Syndrome (PRS), hemifacial atrophy, hyperpigmented facial lesion, cosmetic correction.

INTRODUCTION

Syndromes are always a rare entity in maxillofacial practice. Here we present a case of Parry Romberg Syndrome (PRS) which is a rare condition where the published resources are also limited. It was first described by Parry in 1825 and Romberg in 1846. Eulenberg in 1871 gave the disease its current nomenclature, progressive hemifacial atrophy.¹ It is a rare clinical scenario of progressive hemifacial atrophy characterised by unilateral facial atrophy with gradual shrinkage of hard and soft tissues along with associated systemic manifestations. It is also associated with atrophy of contra lateral or ipsilateral arms and legs. The oral manifestations include atrophy of the tongue, wasting of masticatory muscles, deficiency of soft and hard palate, reduction in jaw growth on the affected side, delayed teeth eruption, absence of teeth, absence of salivary gland on the affected side^{6,7,8}. Early clinical features may be the presence of a depressed vertical scar along the centre of the forehead known as an "en coup de sabre".² Atrophy of the skin, fat, connective tissue, muscles and bones can result in restriction of growth in all dimensions. Intraoral abnormalities, include delayed dental eruption, root abnormalities and hypodontia.⁴

Areas of hyper or hypopigmented skin on the ipsilateral or contralateral side of the facial deformity is a characteristic finding. In this article we report a case with mostly all of these

Department of Oral and Maxillofacial Surgery, Mar Baselios Dental College, Nellikuzhi, Ernakulam, Kerala, India; Department of Pedodontics, Mar Baselios Dental College, Nellikuzhi, Ernakulam, Kerala, India.

Corresponding Author: Joy R Das, Department of Oral & Maxillofacial Surgery, Mar Baselios Dental College, Nellikuzhi, Ernakulam, Kerala, India. Email drjoy303@gmail.com

How to cite: George A, Vichattu SV, Das JR, Khosla E, Paul S, James JE. Parry Romberg Syndrome with its Oral Manifestations- Report of a Case. *Oral Maxillofac Pathol J* 2024; 15(2):286-288

Source of Support: Nil

Conflict of Interest: None

characteristic features of PRS. The aetiology of this disorder is unknown but thought to be of autoimmune nature due to the favourable response to immunosuppressive drugs.² The disease is self-limiting within 2 to 10 years of its diagnosis. It starts as a skin or subcutaneous disease, progresses to the underlying muscles, cartilage and hard tissues. Usually presented from middle of adolescence to fifth decade of life. Female predilection is noted in most instances.

There are also other systemic manifestations associated with the progressive hemifacial atrophy. Neurologic findings include migraine, epilepsy and trigeminal neuralgia. Ocular abnormalities are also noted.^{3,4,5}

CASE REPORT

A 47 year old female reported to our Department of Oral and Maxillofacial Surgery with a chief complaint of pain in her lower left back tooth region and front tooth region since two weeks. Patient noticed pain and tenderness while brushing. There was also a reddening of buccal vestibule on the left side posterior region with a draining sinus which upon investigation with an orthopantamogram revealed all characteristics of osteomyelitis (Figure 1). She discontinued the use of her denture since then. She had a medical history of hypertension for which she was under medication. Had a few episodes of seizures few years back in her life and was under medication for five years.

General examination revealed facial asymmetry with atrophy of the left side of the face, atrophy of the left hand and leg (Figure 2,3). Bilateral TMJ clicking was present There was hyperpigmentation on all these atrophied areas. There was a

scar tissue along the left side symphyseal region of the mandible from where the hyperpigmentation started in the face. The atrophy and the hyperpigmentation extended superiorly upto her tragus region. The left hand was hypertrophied upto her elbow with hyperpigmentation. The skin over the atrophied region was sort of contracted like a scar. Similar condition was noted in the left leg with atrophy on the lower leg extending to the angle upto the metatarsals. Disturbance in gait was not noted.

Intraoral examination revealed an edentulous maxilla and mandible. There was hypertrophy of the tongue on the left side. The edentulous mandibular arch was hypertrophied extending from the anterior premolar region to the retromolar region (Figure 4). The mouth opening was well within normal limits. There was no hyperpigmentation. Diffuse erythmatic swollen alveolar region was noted towards the posterior region with

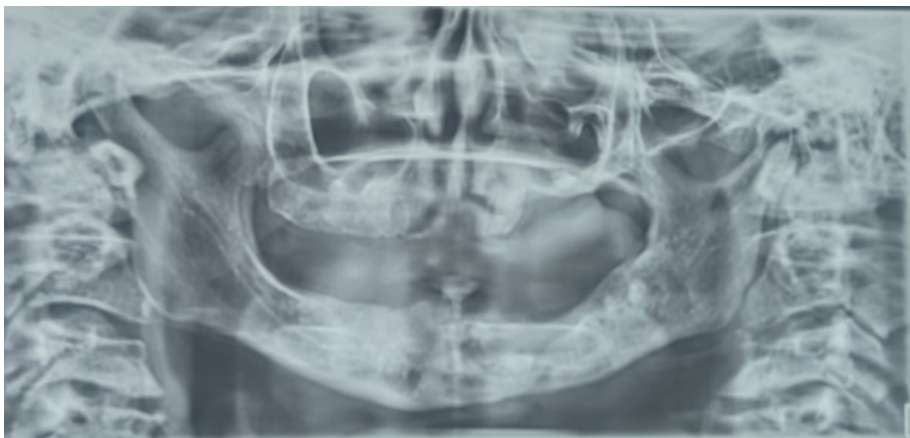


Fig. 1: Atrophic mandible on the left side



Fig. 2 Left side atrophy of the mandible and visible colour change of the skin overlying the region



Fig. 3: Atrophy of the left hand and limb



Fig. 4 Intra orally atrophied area of the left side of mandible extending from anterior region to the retromolar region

the presence of a draining sinus. The area was tender and soft in consistency. Orthopantomogram (OPG) revealed multiple radio opacities with patches of radiolucencies suggestive of osteomyelitis.

The patient also showed reduction in salivary flow on the left side, the saliva present was of a thick consistency. The patient mentioned the same was present since few years and patient was not willing for further examination of salivary glands. Neurologic symptoms were minimal although the patient reported random headaches.

All these anomalies in the patient were noted and it was typical to classic Parry Romberg Syndrome. Osteomyelitis was addressed with curettage and sequestrectomy of the affected site with antibiotic therapy. It is not known whether osteomyelitis happened as a sequelae to PRS and no literature were found supporting the same although there was a case where osteomyelitis presented in a patient with PRS which exacerbated to trigeminal neuralgia which is a neurologic sequelae to PRS.

DISCUSSION

The patient discussed above showed most of the clinical presentations pathognomonic to PRS. This is also known as progressive hemifacial atrophy with a prevalence of 1: 70000002. Usually there is no sex predilection although some literature shows a greater propensity to females¹. Some literatures support the fact that PRS is an autoimmune disorder which regresses with immunosuppressants.² There is no known etiology; although could be triggered by a trauma or a viral infection. It progresses slowly, stabilizes after a period of 2 to 10 years. The diagnosis is made upon clinical findings and imaging.

The first symptom usually presents as “en coup de sabre” which is a unilateral frontal or frontoparietal impression of skin and underlying tissues, with scarring of the dermis. The overlying skin can be hyperpigmented which is however not found in most patients. Our patient exhibited this classic feature. There is hypertrophy of the skin, fat, connective tissues, muscles and bone, which over a period of time progresses slowly and exhibits as atrophy over the facial skeleton and limbs associated with the typical hyperpigmentation. There are neurologic and ophthalmic manifestations associated with PRS. Migraine, trigeminal neuralgia and epilepsy were noted as neurologic manifestations. Enophthalmos was seen as an ophthalmologic manifestation. Our patient had history of seizures for which she was under medication and unexplained headaches similar to migraine.

Intraoral soft tissue changes include unilateral tongue atrophy, wasting of the muscles of mastication, soft palate deficiency and absence of salivary glands on the affected side.³ Our patient showed most of these features. She had reduction of salivary flow on that particular side where she exhibited thick mucous sticky saliva. She was not willing for any investigations regarding salivary glands.

Intraoral hard tissue findings included hypoplasia of the mandible and maxilla, abnormality in tooth and root morphology, delayed eruption, crossbite and malocclusion³. Our patient exhibited hypoplasia of the mandible and premature loss of tooth. OPG clearly reveals the atrophied mandible extending upto the condyle region.

In literature a case of osteomyelitis was presented in a patient with PRS, which further progressed to trigeminal neuralgia which is a neurologic sequelae of PRS, considering the fact that occurrence of osteomyelitis in our patient could be a reason for trigeminal neuralgia in the future.⁹

Treatment for PRS is usually symptomatic, although immunosuppressants were found to have beneficial effects in reducing the neurological findings associated with this syndrome. It is also considered to be a clinical variant to linear scleroderma which is an autoimmune disorder. Immunosuppressants like azathioprine and hydroxychloroquine (HCQ) were found to be useful in neurologic manifestations of PRS.¹⁰

Cosmetic procedures are usually helpful in enhancing the psycho social well being of the patient. Recent researches have added the option of stem cell enriched fat grafts, which should reduce fat absorption. Fat grafts in general have gained importance because it is a less invasive technique with good aesthetic results, and can be added even during disease progression. Orthodontic and orthognathic interventions can also be considered. Unilateral distractions followed by jaw corrections are also treatment options. Autologous or allogenic bone grafts are also needed to correct the bony deformities.

CONCLUSION

Parry Romberg Syndrome is a rare entity in day to day clinical practice, characterized by unilateral atrophy and hyperpigmentation of face and body parts. A multidisciplinary approach is mandated in such patients to enhance their quality of life.

REFERENCES

1. Tk Mittal, H Safaei, L Carter and Cj Bates. (2020). Parry Romberg Syndrome. An unusual case with dental features. *Journal of Oral Care and Dentistry* 2(1). DOI: 10.5281/zenodo.4178244
2. Stone J. Parry Romberg syndrome. *Practical Neurology* 2006; 6: 185-188
3. Oral manifestations of Parry-Romberg syndrome: A review of literature Nader A. Al-Aizari, Saleh Nasser Azzeghaiby, *Avicenna J Med.* 2015 Apr-Jun; 5(2): 25-28
4. Parry Romberg Syndrome: Literature Review and Report of Three Cases N Girish Kumar 1, Brig S Maurya 2, Col S Sudeep J *Maxillofac Oral Surg.* 2019 Jun;18(2):210-216. doi: 10.1007/s12663-018-1147-7. Epub 2018 Aug 18
5. Parry-Romberg syndrome with rare maxillofacial deformities: A report on two cases Author links open overlay panel Xiao-Jun Tang, Wei Liu, Bin Yang *Journal of Cranio-Maxillofacial Surgery* Volume 42, Issue 6, September 2014, Pages 780-783
6. Fornwalt BE, Altman JS. Parry-Romberg syndrome in an adult: Report of a case. *Ear Nose Throat J.* 2013;92:E1-3.
7. Blaszczyk M, Królicki L, Krasu M, Glinska O, Jablonska S. Progressive facial hemiatrophy: Central nervous system involvement and relationship with scleroderma en coup de sabre. *J Rheumatol.* 2003;30:1997-2004.
8. Khan M, Khan M, Negi R, Gupta N. Parry Romberg syndrome with localized scleroderma: A case report. *J Clin Exp Dent.* 2014;6:e313-6.
9. Popovici, Florin AU Mergeani, Athena Exacerbation of secondary trigeminal neuralgia due to osteomyelitis in a case of Romberg facial hemiatrophy *Romanian Journal of Neurology,* March 2011;12;3-8
10. Korkmaz C, Adapinar B, Uysal S. Beneficial effect of immunosuppressive drugs on Parry-Romberg syndrome: a case report and review of the literature. *South Med J.* 2005 Sep;98(9):940-2.

