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An overlap of Parry Romberg syndrome and linear scleroderma: Report of a rare presentation.

Dr.Veenakrishnan[1], Dr Sreela L.S[2], Dr Philips Mathew[3], Dr Twinkle S Prasad[4], Dr Admaja K Nair[5]

[1]*(Junior Resident, Department of Oral and Medicine and Radiology, Government Dental College Kottayam, Kerala, India,)*
[2]*(Professor and Head of the Department, Department of Oral and Medicine and Radiology, Government Dental College Kottayam, Kerala, India)*
[3]*(Assistant Professor, Department of Oral and Medicine and Radiology, Government Dental College Kottayam, Kerala, India)*
[4]*(Associate Professor, Department of Oral and Medicine and Radiology, Government Dental College Kottayam, Kerala, India)*
[5]*(Assistant Professor, Department of Oral and Medicine and Radiology, Government Dental College Kottayam, Kerala, India)*

Abstract : *Parry Romberg Syndrome (PRS) is a relatively rare degenerative disorder of unknown etiology. It is characterized by a slow and progressive atrophy of one side of the face and frequently occurs in association with a linear form of localized scleroderma -en coup de sabre. The syndrome will have characteristic skeletal, dental, and soft tissue changes in the affected half of the face with or without neurological signs and symptoms. The exact etiology, and pathophysiology of PRS is not yet identified. Since there is no definitive treatment, management of such cases need prompt multidisciplinary approach, keeping in mind the aesthetics, restoring the masticatory function along with symptomatic treatment of neurological deficit. The present article reports one rare case of 34-year-old female patient with a progressive hemifacial atrophy of left side of the face with en coupe de sabre.*

Keywords: *Cone beam computed tomography, Facial asymmetry Facial hemi atrophy, localized scleroderma, Migraine with aura.*

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I. Introduction

Parry Romberg Syndrome (PRS) is a relatively rare degenerative disorder of unknown etiology. It is characterized by a slow and progressive atrophy of one side of the face and frequently occurs in association with a linear form of localized scleroderma -en coup de sabre. The syndrome will have characteristic skeletal, dental, and soft tissue changes in the affected half of the face with or without neurological signs and symptoms. The exact etiology, and pathophysiology of PRS is not yet identified. Since there is no definitive treatment, management of such cases need prompt multidisciplinary approach, keeping in mind the aesthetics, restoring the masticatory function along with symptomatic treatment of neurological deficit. The present article reports one rare case of 34-year-old female patient with a progressive hemifacial atrophy of left side of the face with en coupe de sabre.

Case Description

Parry Romberg Syndrome (PRS) also known as progressive hemifacial atrophy (PHA) of the face characterized by a slow progressive self-limiting atrophy of skin as well as the subcutaneous tissue on one side of the face and rarely the body. It was first described by Caleb Hillier Parry in 1825, detailed by Heinrich Romberg in 1846 [1] and this constellation of the craniofacial finding was later labelled as progressive hemifacial atrophy by Eulunberg in 1871. Other names used to describe this syndrome include progressive facial hemi atrophy, idiopathic hemifacial atrophy and Romberg syndrome.

PRS occurs insidiously during the first and second decade of life and progresses within 2-10 years following the onset and then stabilizes resulting permanent deformity of the affected side. The disorder generally involves the dermatomes supplied by the branches of trigeminal nerve affecting subcutaneous tissue, muscle, fat and osteocartilage, which results in a sunken hemiface appearance [2,3].

This report presents one rare case of a middle-aged female patient with Parry Romberg Syndrome associated with linear scleroderma and a history of neurovascular disease and aims to evaluate the clinical features and treatment part of this rare disease in detail.

II. Case Description

A 34-year-old female patient (Figure 1) presented to the Department of Oral Medicine and Radiology, with the complaint of multiple decayed teeth and root stumps.

After obtaining informed consent from the patient, physical examination was carried out and a gross facial asymmetry with distinct hypoplasia of the left side of the face was noted (Figure 1). A linear scar like defect was present over the left side of the forehead passing downward along the lateral border of the nose terminating at the left nasolabial fold region (Figure 2()). Eyebrows, eyes, and ears were normal in size, shape and position. The left maxillary and mandibular bones appeared hypoplastic compared to right side. Deviation of the nose, angle of mouth and chin to the left side was evident. The upper and lower lip contracture and a slight commissural lift on the affected side with atrophy of masseter and buccinator muscle was noticed (Figure 2(b)). There was a diffuse hyperpigmentation present over the left perioral region (Figure 2(b)). Tenderness over left temporomandibular joint with deviation of mandible to the affected side on mouth opening was observed.

On further evaluation, patient gave a history of noticing brownish pigmentation and shrinkage of face at the age of 15 years. She visited a local physician who advised cosmetic correction for the same, but was not able to carry out the treatment due to financial problems. She was diagnosed with migraine at the age of 18 and was under the medication for past 16 years. There was no history of any eye or ear diseases or severe speech impairment. No family history of similar lesions, no specific history of trauma, no history of consanguineous marriage, and birth history of full-term normal delivery without any birth related trauma was reported.

Intraoral examination revealed partially bifid tongue with loss of papillae on the left side (Figure 3a). Hard tissue examination revealed multiple grossly decayed teeth and root stumps with no change in size and shape compared to the unaffected side. Canting of occlusal plane was evident on the affected side (Figure 4).

Following the diagnostic work up a provisional diagnosis of Parry Romberg Syndrome was made prior to investigations. Routine blood investigations were carried as a part of preliminary investigation which revealed all values within normal limits. Antibody profiling for antinuclear antibody was done and found to be negative.

Panoramic radiograph revealed thinning of body, ramus with prominent antegonial notch, hypoplastic slender condyle and coronoid process, and relatively shorter tooth roots on the left side (Figure 5). Paranasal sinus (PNS) view (open mouth) displayed hypoplastic frontal, maxillary and sphenoid sinuses on left side (Figure 6). Posteroanterior (PA) cephalogram confirms the clinical finding of asymmetry of the face with underdeveloped midface (Figure 7). Cone beam computed tomography (CBCT) demonstrated (Figure 8,Figure 9)marked osseous asymmetry and deviation of nasal septum to the left side with hypoplasia of all the air sinuses, maxilla condyle, coronoid and ramus and body of mandible on the affected side. Coronal sections demonstrated a prominent conchae bullosa on the left side.

Based on clinical and radiographic findings, the diagnosis of progressive hemifacial atrophy was made. As a part of the treatment all the root stumps and teeth with poor prognosis were extracted and advised prosthetical rehabilitation with a removable partial denture. In order to improve the facial disfigurement cosmetic surgical management using alloplastic implants were suggested and the patient was referred to the plastic surgery department.

We confirm that written consent was obtained from the patient for the publication of the report as well as the use of any photographs and agreed to withheld the identity unless needed.

III. Discussion

Parry Romberg Syndrome is an uncommon degenerative condition involving unilateral atrophy of the face, which rarely affects the limb and trunk. The prevalence rate was found to be 1 in 700,000(7 hundred thousand) in the general population [4]. The incidence and aetiology of this disease is poorly understood and various factors like trauma, autoimmunity, endocrine, metabolic and genetic involvement has been proposed [5]. A cerebral disturbance of fat metabolism has been suggested as a primary cause [6]. Some authors have proposed that a trophic malfunction of sympathetic nervous system can be a factor, since proper development of muscle, bone and skin requires trophic stimulation [5,9].

The syndrome initially develops in the first 20 years of life as in this case and characteristically burns out in 2 to 10 years before being stationary [8]. The disorder shows a female predilection (3:2) and more often occurs over the left side of the face(4)]. The unilateral atrophy of the skin, subcutaneous tissue, muscles and bone results in facial asymmetry with a distinct demarcating line between the affected and unaffected tissue resembling a large linear scar called en coup de sabre (ECDS), which represents a localized form of linear scleroderma; however, many authors consider it as a separate entity [5]. Mayo clinic has classified linear scleroderma to be a subtype of localised scleroderma and results in sclerosis confined to skin. Differentiating the

two is difficult as some patient initially presented with linear scleroderma transforms to progressive hemi facial atrophy and both will have a similar age of onset and progressive course (9). In case of localised scleroderma cutaneous sclerosis manifested as en-coupe de sabre will be localised to the fronto-parietal region involving the forehead, and in PRS a widespread unilateral tissue atrophy with a thin skin will be evident. In our case co-existence of both the condition is seen. Enophthalmos, misshapen ears, alopecia as well as psychological and relational disorders (9) has also been seen with the syndrome but were not observed in this case. Skin will be tense with loss or gain of pigmentation. The common neurological and neurovascular symptoms associated with PRS include trigeminal neuralgia, migraine type headache (which was noted in our case), and focal epilepsy). Teeth abnormalities, masticatory muscle hypoplasia and multiple facial bone atrophy were also reported. Intraoral soft tissues of the involved side also get affected which will indirectly impair chewing, smiling and speech. Pain secondary to masticatory muscle spasm, locking of the jaw and temporomandibular joint disorders has also been reported [8].

Differential diagnosis include Bell's palsy (where weakening of facial muscles occurs rather than thinning), Hemifacial microsomia and its clinical variants such as Goldenhar syndrome (these are non-progressive and congenital in nature), Post-traumatic atrophy, acquired partial progressive cephalothoracic lipodystrophies - Baraquer Simons Syndrome (bilateral with systemic involvement, involves primarily adipose tissue) fat necrosis, congenital deformities (such as wry neck) and Rasmussen encephalitis [8].

Since this disease is self-limiting and there is no definite cure, management is mainly directed in treating the active state with immunosuppressive agents and corticosteroids. Surgical treatment requires a multidisciplinary approach with frequently repeated procedures based on the degree of involvement and are mostly advised only after the burn out phase and should involve expertise involvement of dermatologist, dentists and psychologists [10]. Guerrerosantos et al. divided patients with PRS into four categories based on the intensity of the defect to plan treatment. Type 1 patients had a mild form of the disease which can be noticed easily only by patient's family or close acquaintances. There is no bony involvement. In Type 2, the defect is more obvious but do not involve hard tissue. Type 3 patients show both soft tissue and hard tissue involvement with severe deformity. Type 4 patients have more severe deformity with the skin almost attached to the bone and patients have functional problem of their lip and nose [7]. For mild to moderate cases lipoinjection, fat grafting along with other soft tissue fillers can be employed [8]. For severe cases combined soft tissue and skeletal augmentation is advised. Lip repair, lip augmentation, Z-plasty, face-lift, nasal reconstruction and other available appropriate adjuvant techniques can be used to generate a better result [8]. In dental point of view, correction of eruption problem and associated difficulties should be considered if the lesion is in the active state. Preservation or the replacement of occlusion possess a great challenge to the dentist. Correction of malocclusion by orthodontic treatment and myofunctional therapies have greater role during the growth phase .Once the burn out stage has reached and the growth phase has been completed as in the present case, focus should be given in correcting the functional aspect by fabricating prosthetic appliance to correct occlusal irregularities and modifying the aesthetic appearance with the help of plastic surgery(10).Instructions regarding the need of oral hygiene maintenance and mouth dilating exercises such as tongue blades and ice cream stick exercises should be advised in patients with decreased mouth opening[11]. TMDs should be treated with muscle relaxants ,dental appliances and physiotherapy .Oral sensory disorders such as dysgeusia can occur in some instances and care should be taken to identify such conditions and provide necessary treatment[11].

IV. Figures

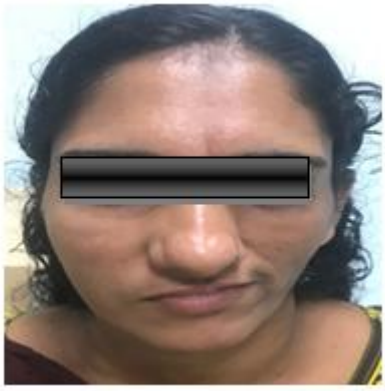

Figure 1 . Frontal view showing gross facial asymmetry, hypoplasia on the left side.

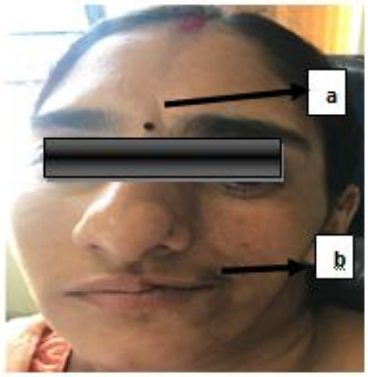

Figure 2. Left side of the face revealed a) prominent cleft: en coup de sabre extending from the forehead to the nasolabial fold, b) perioral hyperpigmentation in relation to the nasolabial fold, hypoplasia of the maxilla and the mandible.

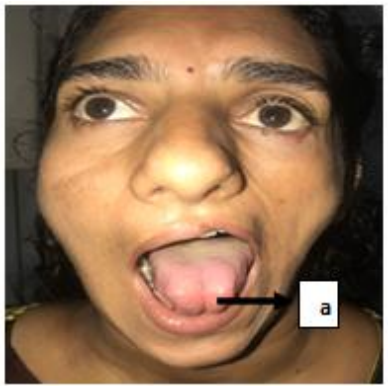

Figure 3. Intraoral view showing a) partially plicated tongue anteriorly, with loss of papilla on the left side of the dorsum of tongue.

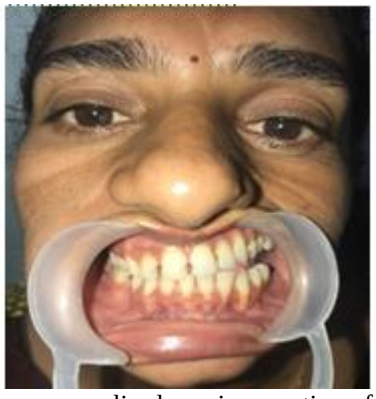

Figure 4. Clinical evaluation of dentition: generalised spacing, canting of the occlusal plane on the left side was noted.

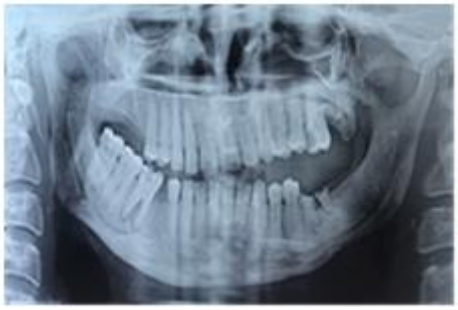

Figure 5. Panoramic radiograph showing hypoplastic mandible with slender condyle, coronoid, widened sigmoid notch, deepened antegonial notch and shorter tooth roots on the left side.

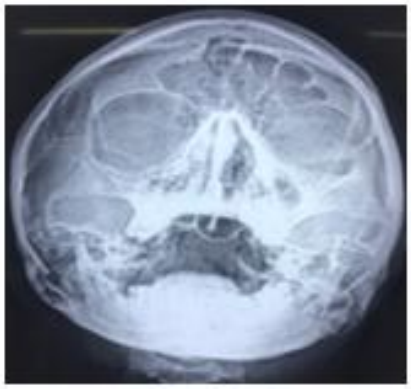

Figure 6. Paranasal sinus (PNS) view, open mouth: showing hypoplasia of frontal, ethmoid and sphenoid sinus on left side.

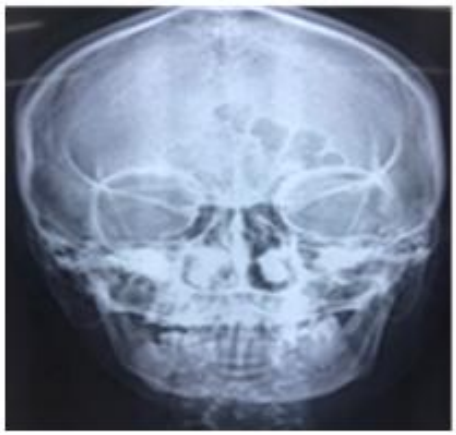

Figure 7. Posteroanterior (PA) cephalogram: showing evident facial skeletal asymmetry in relation to the left side.

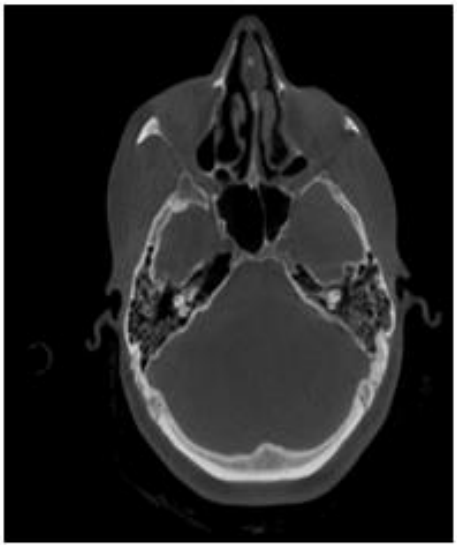

Figure 8. Cone beam computed tomography (CBCT)
a)axial:atrophy of left ethmoidal sinus

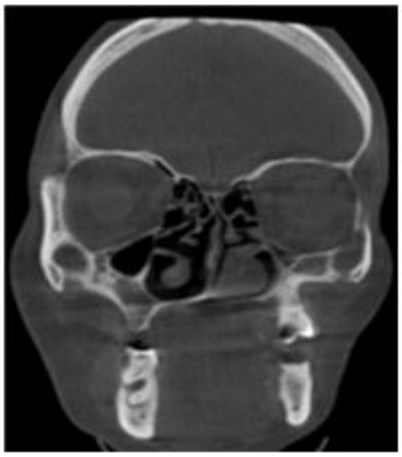

Figure 8b) sagittal sections -displays deviation of nasal bone to the left side with a prominent concha bullosa within the nasal cavity. Hypoplasia is observed in relation to left frontotemporal region.

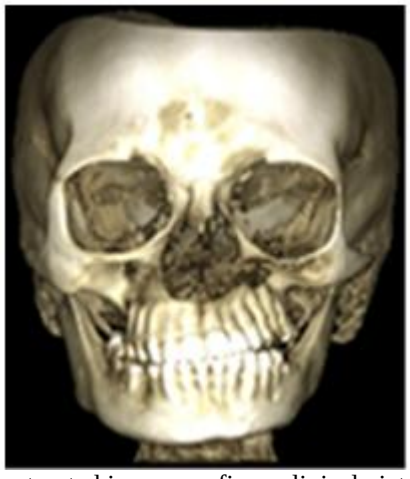

Figure 9. a) Three-dimensional reconstructed image confirms clinical picture of gross facial asymmetry visible upward cant of occlusal plane on left side, hypoplasia of zygoma and zygomatic process

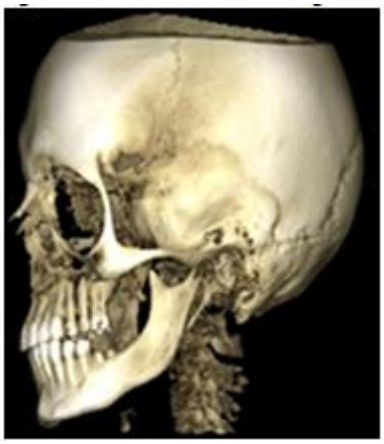

Figure 9 (b) Hypoplasia of ramus, angle, body of mandible,coronoid and condylar process in relation to the left side. (There are two figures which needs explanation. Please separate as a and b).

V. Conclusion

In brief, a case of progressive hemifacial atrophy with its archetypical feature has been discussed. PRS is considered to be a disfiguring degenerative disorder of unknown origin where early accurate diagnosis and prompt management is necessary for enabling the patient to improve the quality of life. Appropriate diagnostic work-up and a multidisciplinary therapeutic approach is the key in the rehabilitation of the patient.

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