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Parry Romberg Syndrome: A case report and review

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Abstract
Parry-Romberg Syndrome (PRS) or idiopathic hemifacial atrophy is a rare developmental craniofacial syndrome. It is characterized by slowly progressive atrophy of one side of the face, primarily involving the skin, fat and connective tissue. It is an acquired condition of unknown aetiology presenting usually in childhood or adolescence, with gradual progression over several years. A multidisciplinary approach is effective in the management of this disorder. We present a case report of PRS in a15 year old girl with the review of aetiology, clinical features and treatment of PRS. The diagnosis was established mainly based on clinical and radiological findings. Clinicians must be aware of PRS to identify the invalidating disorder.

Keywords: PRS (Parry Romberg syndrome), Romberg sign, partial lipodystrophy

1. Introduction
Parry Romberg Syndrome (PRS) is a rare and remarkable disease entity which is also called as idiopathic progressive hemifacial atrophy, as it is characterised by atrophy of the skin, fat, connective tissue and muscles of one side of the face and is of unknown aetiology [1]. It is an acquired condition presenting usually in childhood or adolescence with gradual progression over several years and has higher prevalence towards female. Parry Romberg syndrome was named so, as it is first reported by Caleb Parry in 1815 and described by Moritz Romberg in 1846 [2]. Exact pathogenesis was unknown but contributory factors were disturbance of fat metabolism, trauma, viral infection, heredity, endocrine disturbances and autoimmunity. As very few case report were reported in literature, the prevalence of the disease has not been determined precisely, although it is estimated that it affects at least 1 in 700,000 people [3].
On the basis of the severity of disease and the areas affected by this syndrome; PRS has been classified into the following forms:

1.1 Mild form: Skin and subcutaneous tissue atrophy limited to the area supplied by one of the sensory branches of the trigeminal nerve, without bone involvement;

1.2 Moderate: Involvement of the area supplied by 2 branches of the trigeminal nerve, without bone involvement;

1.3 Severe: Involvement of the area supplied by all 3 branches of the trigeminal nerve and/or involvement of the bone structure [4].
PRS can also be classified according severity of facial soft tissue atrophy and the extent of involvement of the osseous framework work which include; (1) Mild: the facial atrophic area is confined to a small region and is located in the lateral face, the nasal ala and upper lip are normal, and there is no deviation of the oral commissure, and the occlusal plane is horizontal. (2) Moderate: there are large areas of soft tissue atrophy, and the nasal ala and upper lip are also affected; the oral commissure is deviated; the bony framework is nearly normal; and the occlusal plane is nearly horizontal or slightly deviated. (3) Severe: moderate form of soft tissue atrophy and serious bone framework atrophy, involving the zygoma, maxilla, and mandible; the chin and occlusal plane have deviated extensively to the affected side [5].
Parry Romberg syndrome appears to overlap with “en coup sabre” which is a type of linear scleroderma affecting head [6] Extra orally, there is presence of typical features like wasting of facial muscles, localized area of alopecia, loss of the eyelash and eyebrow, and blanching of the hair. Extracutaneous manifestations of the disease includes neurological, auricular, ocular, autoimmune and dental abnormalities as described by other authors.

The most common complications are: trigeminal neuritis, facial paresthesia, severe headache, and epilepsy [3]
Parry Romberg syndrome is a self-limited condition with no specific line of treatment. To provide better aesthetics, multidisciplinary attendance of plastic surgeons, physicians, dental surgeons, phono-audiologists and psychologists were required. Now a days, cosmetic surgeries with autogenous fat graft, injection of silicone or bovine collagen and inorganic implants are some alternatives to correct deformities. Besides, aesthetic improvement, symptomatic treatment for neurological disorders is indicated [7].

2. Case Report

A 15 year old girl reported to our department with a chief complaint of forwardly placed teeth. Patient revealed that she had progressive shrinking of the left side of the face which started approximately 9 years ago with depigmentation on the back side of the neck for which she was undergoing treatment. No history of trauma was given by the patient. Medical and family history were non-contributory. Patient appeared normal at her younger age which is evident in a photograph which was taken at the age of 4 years. (Fig 1)

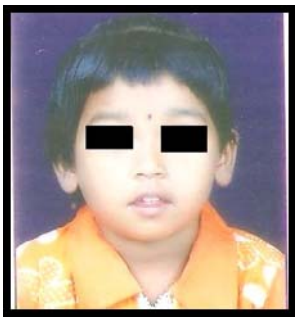

Fig 1: Photograph at the age of 4 year

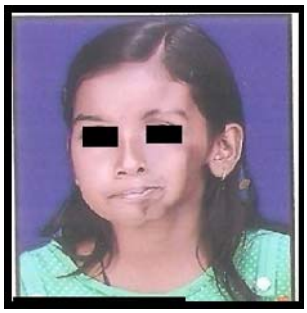

Fig 2: Photograph at the age of 10 year.

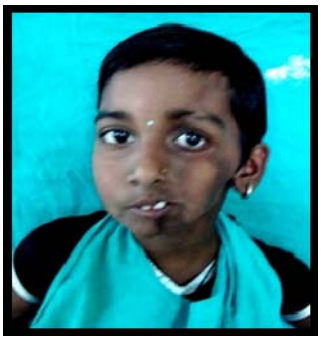

Fig 3: Photograph at the age of 15 year.

On general examination, the patient was found to be lean with a short stature. Romberg sign was negative. On extra oral examination, there was a marked asymmetry of the face (fig 3). There was hypoplasia involving the left side of the face due to atrophy of subcutaneous tissue and muscles. Discrete areas of hyper pigmentation were noted on the left side of the face and neck. Notching was noted on the forehead extending from the left mid supraorbital region to the parietal region (en coup de sabre). There was enophthalmos on the left side with an area of alopecia on the left eye brow. Deviation of the lips to the affected side was appreciated. There was atrophy of masseter, buccinator and temporalis muscles. Vitligo was evident on the left sub occipital region.
On intra oral examination, atrophy of the left side of the tongue was observed with slight deviation to the left side. Generalized enamel hypoplasia was seen with proclination of 11, 21 and mobility of 32.
Based on the history and clinical findings a provisional diagnosis of severe form of progressive hemifacial atrophy (Parry Romberg syndrome) was given and the patient was subjected to investigations. Partial Lipodystrophy (Barraque-Simons syndrome) and Rasmussen encephalitis were considered for differential diagnosis.
The radiological investigations carried out included CT scan of brain, orthopantomography, PA view of skull and X-ray of spine. CT scan of the brain was found to be normal. Digital OPG(fig 4) and PA view (fig 5) of skull revealed asymmetry of the face, hypoplasia of maxillary sinus, nasal turbinates and conchae on the left side, hypoplasia of the left condyle, ramus and body of the mandible. Styloid process could not be traced on the left side. Ophthalmologic and ENT consultations were sought to rule out any syndromic association. Audiogram did not show any hearing deficit and significant visual defects were not detected except myopia. Hematologic investigations and blood sugar levels were within normal limits. Considering the clinical findings and investigative reports, a final diagnosis of severe form of Parry Romberg syndrome was arrived at. The patient was then referred to the department of paediatric dentistry for needful.

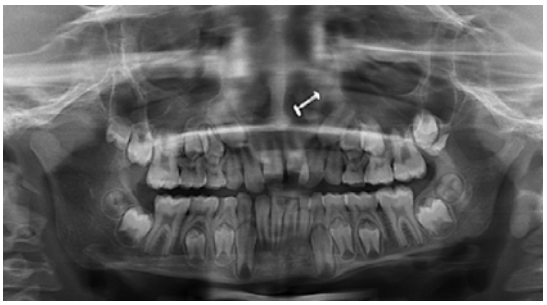

Fig 4: Panoramic view revealed hypoplasia of the left condyle, ramus and body of the mandible.

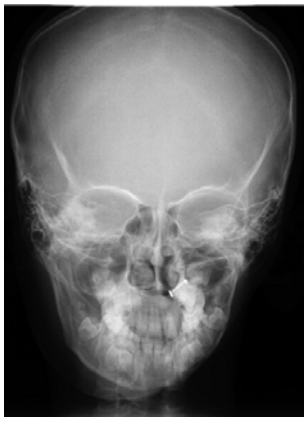

Fig 5: PA view revealed asymmetry of the face, hypoplasia of maxillary sinus.

3. Discussion

Parry Romberg syndrome is a rare condition which is sporadic. It has been reported that it is more frequent in females, without apparent geographic or ethnic predilection. This disease usually manifest during the first and second decades of life, resulting in an initially insidious but progressive hemiatrophy of the face [3]. As the disease progresses, patients starts experiencing atrophy of the skin and subcutaneous tissues. This with duration may involve the underlying muscular, cartilaginous, osseous, and glandular structures causing severe atrophy. The most commonly involved areas reported are maxillary or periorbital region which may with time expand to involve the forehead, teeth, jaw, and neck to varying degrees [8]. A linear depression generally located on the paramedian forehead, also known as en coup de sabre may be associated with this syndrome. Earlier onset and longer duration of PRS increases the severity of this disease and hence causing severe facial deformity. PRS is generally associated with skin discoloration, alopecia and is typically restricted to one side of the head and neck, but bilateral disease also has been reported in the past. Teeth abnormalities such as shorter crowns and roots along with crowding of teeth has been reported. The frequent involvement of masticatory muscles is frequently seen in patients with PRS [3, 15]. Hemiatrophy in PRS in severe cases also causes. Sometimes it may be associated with cranial neuropathies involving cranial nerves III, V, VI, and VII. Neurologic symptoms are seen in 15%– 20% of patients affected by PRS, with most common being ipsilateral headaches, facial pain, and seizures. Other neurologic symptoms associated with PRS include trigeminal neuritis, facial paresthesia and cognitive impairment [11]. Romberg sign is observed in patients with neurological and vestibular dysfunction. Romberg test is performed by having the patient's feet together and eyes closed, which eliminates the visual clues that help to maintain posture. Patients with positive Romberg's sign show diminished proprioception [12]. In 10%–35% of patients ophthalmologic symptoms involving the ipsilateral orbit is observed. Enophthalmos which present is mainly due to atrophy of the retrobulbar fat. Other potential orbital abnormalities include uveitis and retinal or optic nerve alterations [13].

Many theories have developed during the recent years in an attempt to explain this syndrome. There is no single theory which has been satisfactory in fully characterizing and predicting PRS. Understanding of the underlying pathophysiology of this syndrome remains limited. Proposed aetiologies includes trauma, genetic predisposition, infection, radiation exposure, embryonic developmental dysfunction, sympathetic cervical ganglion dysfunction and metabolic and endocrinologic disturbances. [14] Currently, the strongest laboratory and histologic evidence supports the etiology to be of that an inflammatory autoimmune disorder with or without associated vasculopathy, which is also supported by clinical improvement observed in patients affected by PRS with immunosuppressive therapy during active disease [3, 15].

Diseases in which facial asymmetry is a prominent clinical feature includes hemifacial microsomia (first and second brachial arch syndrome) and Goldenhar syndrome, but unlike PRS, these conditions are typically congenital and nonprogressive. Hemifacial hyperplasia causes asymmetry of the face as well, but rather than atrophy, this entity is characterized by overgrowth and hyperplasia [16]. The clinical description of Rasmussen syndrome consists of intractable seizures, progressive hemiparesis and focal atrophy of brain [17]. Partial lipodystrophy (Barraque-Simons syndrome) may have manifestations similar to those of PRS, but typically these are bilateral.

Table: Various differential diagnosis of parry Romberg syndrome

Disease	Prominent clinical feature
Parry Romberg syndrome	Atrophy of skin, fat, muscles and bone. Generally unilateral, progressive and developmental
Hemifacial hyperplasia	Asymmetry of the face, characterized by overgrowth and hyperplasia
hemifacial microsomia (first and second brachial arch syndrome)	These conditions are typically congenital and nonprogressive
Goldenhar syndrome	Congenital and non-progressive
Partial lipodystrophy (Barraque-Simons syndrome)	Similar to those of PRS, but typically these are bilateral.
Rasmussen syndrome	intractable seizures, progressive hemiparesis and focal atrophy of brain

There is no standard treatment algorithm which has been accepted for PRS especially owing to the fact that response to treatment is difficult to assess. In general; treatment goals are to obtain seizure control if seizures are associated with this syndrome. In cases where present, provide symptomatic relief and halt disease progression [3]. PRS-related seizures are usually treated with anticonvulsive therapy despite their commonly refractory nature. Additionally, similar to treatments for scleroderma, immunosuppressive therapies, ranging from topical corticosteroids to systemic corticosteroids, immunomodulators, and plasmapheresis, have been used with varying degrees of success [5].

Once the disease stabilizes, a finalized treatment plan has to be fabricated in accordance to the extent of the disease [18]. The various modalities to address the deformity includes pulse dye lasers, dermal fat grafts, autologous fat grafts, muscle flap grafts, free silicone injections, and bone augmentations. The success of all these modalities is yet to be evaluated both in terms of aesthetics and success [19].

4. Conclusion

Parry Romberg syndrome is an uncommon condition, which manifest as atrophy of one side of the face. This syndrome requires early diagnosis and careful treatment approach. A valid treatment needs to be developed and randomised clinical trial to be performed in order to establish best treatment approach for this rare but severely disfiguring disease.

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