


Oral manifestations of Parry-Romberg syndrome

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Abstract:

Introduction: Parry-Romberg syndrome (PRS) or Idiopathic hemifacial atrophy is a rare neurocutaneous syndrome, characterized by slowly progressive atrophy located on one side of the face, primarily involving the skin, fat and connective tissue and it becomes stable after certain time period. It exhibits many intraoral and extraoral features. The incidence and the causes of this changes remains unknown. **Objective:** We present this case report to improve the understanding of this rare condition. **Case Report:** We present a case of 33 year old male patient who reported to our department with progressive hemifacial atrophy on left side of the face, diagnosed as Parry-Romberg syndrome after clinical and radiographic examination. **Conclusion:** Parry-Romberg syndrome is a rare condition of progressive deformity of the face with various oral manifestations. It may also involve certain ocular and neurological disturbances. The major treatment option is the esthetic management and other symptomatic treatments.

Keywords: Facial Hemiatrophy; Alopecia; Uveitis.

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INTRODUCTION

In 1815 Caleb Hillier Parry was first to report Parry–Romberg syndrome and then it was described by Moritz Heinrich Romberg in 1846¹. Parry–Romberg syndrome is a condition in which there is slow and progressive shrinkage of the tissues and sometimes bones of one or occasionally both sides of the face.^{5–10%} of cases were described as being bilateral². When the onset is before the second decade of life, the underlying bone and cartilage is also involved. A sharply demarcated line may develop between the normal and abnormal skin, called as *coup de saber*. The involved area varies from a discrete lesion to a widespread, extensive malformation.

The most important features of this pathology are the enophthalmia, uveitis, iris atrophy, deviation of mouth and nose towards the affected side, and unilateral exposition of teeth (when lips are involved)². Alopecia and pigmentation of skin may also be seen³. More than an aesthetic trouble, this illness brings several functional and psychological problems, ocular problem, migraine, epilepsy⁴. Parry–Romberg Syndrome is an auto-limitable condition with no cure. Affected patients should have multidisciplinary attendance of plastic surgeon, physicians, dental surgeon, phonoaudiologists and psychologists to give a better aesthetic to patient. Besides esthetic improvement, symptomatic treatment for neurological disorders is indicated.

CASE PRESENTATION

A 33 year old male patient reported to our department of oral medicine and radiology with a progressive deformity of left side of face since 10-12 years. The patient has a good general health and there was no history of any past illness that may cause a facial asymmetry. On extraoral examination, there was facial asymmetry on the left side of the face with marked hypoplasia and also deviation of lips and nose to the left side. A big linear dark scar (*coup de saber*) of about 6cm in length was noted on the left pre-auricular region figure 2. Enophthalmia and uveitis were noted on the left eye. An atrophy of the right and left iris was also noted. Mild alopecia was found on the left eyebrow towards the distal side and also patchy loss of eyelashes of the lower eyelid was observed figure 1. The left ear was slightly smaller than the right ear.

On intraoral examination, one of the relevant alteration was the atrophy of the tongue papillae on the left side figure 3. The maxillary 1st premolar on the affected side was found to be missing and all other tooth was erupted, also crowding of the lower anteriors was noted. There was shift of midline towards the affected



Figure 1. Showing hemifacial atrophy, mild alopecia of the left eyebrow.



Figure 2. Shows the demarcating line 'coup de saber'.



Figure 3. Showing atrophy of papillae on the lateral border of the tongue.

side and also the occlusion was disturbed. There was generalized tooth mobility and gingival recession for the tooth. A traumatic ulcer was found on the left lateral posterior 1/3rd of the tongue.

Radiographically, oral panoramic radiograph shows an impacted maxillary 1st premolar and 3rd molar on the affected side. The mandible have a decreased ramus height on the affected side and it also confirms the midline shift and lower anterior crowding. The overall eruption pattern of the tooth on the left side was retarded as compared to the right side figure 4.

With all these clinical and radiographic findings the diagnosis of Parry Romberg Syndrome was made. The patient is provided with the emergency treatment procedures like extraction of tooth with a hopeless prognosis followed by prosthetic rehabilitation of those tooth. The patient is now under regular review checkup for the periodontal problems.

DISCUSSION

Progressive hemifacial atrophy is a rare pathology of unknown cause, whose degenerative condition affect not only the esthetic but also the functionality of attained hemiface. There have been a number of theories on the etiology of Parry–Romberg Syndrome include trigeminal neuritis, a chronic autoimmune neurovasculitis, a chronic infestation with a neurotropic virus (e.g. Herpes) and an increased sympathetic nerve activity triggering facial atrophy and in association with a connective tissue disorder, particularly scleroderma⁵.



Figure 4. Shows a decreased ramus height on the affected side. It also shows a generalized bone loss and impacted maxillary left 1st premolar, and 3rd molar.

Possible factors that are involved in the pathogenesis include disturbance of fat metabolism, trauma, viral infections, heredity, endocrine disturbances and auto-immunity. A cerebral disturbance on fat metabolism has also found to be a primary cause. None of these theories support proper investigation and thus the cause for Parry Romberg Syndrome remains unresolved. Recently extracutaneous involvement has been described in literature raising awareness that this is not only a cutaneous disease^{5,6}.

The condition is more often found in female population and has predilection for the left side of the face. The prevalence rate is estimated to be at least 1 per 7,00000 in the general population⁵. The onset of this disease occur along the first and second decades of life. Alterations in the involvement, duration and deformity may stabilize in any stage of the growth and development. The condition is usually unilateral as that in our case. The skin on the affected side appears to be dry and can be with pigmentation.

In some patients there may be a linear scar demarcating the normal and abnormal skin known as '*coup de saber*'⁴ as seen in our case on the left pre-auricular area. Another important feature is enophthalmos and other ocular involvement, due to fat loss around the orbit. Uveitis, iris atrophy and also the deviation of mouth and nose towards the affected side, and unilateral exposition of teeth are seen in most cases. Thinning of the ear due to atrophy of the fat around it. Alopecia on the involved side are also seen in patients with parry romberg syndrome as like the alopecia seen on the left eyebrow in our case. Certain neurological complication like trigeminal neuralgia, facial parasthesia, severe headache and contralateral epilepsy may also be found. Parry Romberg Syndrome can be featured as a condition of progressive hemifacial atrophy without cutaneous scleroderma.

Although the intraoral soft tissue and muscles of mastication are involved, the normal function like speech, deglutition are not disturbed.^{7,8} Mouth and the nose will be deviated to the affected side and also there will be marked atrophy of the tongue papillae⁷ as seen in our case. The condition may also be associated with deficiencies of the soft and hard palates in all dimensions. Root atrophy, delayed tooth eruptions and retarded tooth formation may also be observed^{9,10} in parry romberg syndrome which is also evident in our case. However, the affected teeth are normal and vital clinically.⁹ Frequently, there is unilateral posterior crossbite, as a result of jaw hypoplasia and delayed teeth eruption.⁹

The treatment is usually based on reposition of adipose tissue that was lost due to atrophy¹¹. Autogenous fat grafts, cartilage grafts, silicon injections and other prostheses, bovine collagen and inorganic implants are some alternatives to aesthetic correction of the atrophy^{2,4}. The treatment modalities are mainly for the esthetic correction of the facial appearance and the psychological problems that may require a multidisciplinary approach.

CONCLUSION

Parry-Romberg syndrome is a rare condition, which manifest as atrophy of hemifacial structures. In most cases, Parry-Romberg syndrome appears to occur for unknown reasons. It has got many oral manifestations like deviation of mouth and nose, atrophy of tongue papillae, root atrophy, delayed tooth eruption. And there are no valid guidelines to be followed in the treatment

of parry romberg syndrome and the major treatment objective is the esthetic correction for this disfiguring disease.

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