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Dental Implications of Parry-Romberg Syndrome: A Case of Progressive Hemifacial Atrophy

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ABSTRACT

Parry-Romberg syndrome (PRS) is a rare degenerative disorder of unknown origin that causes slow, progressive atrophy usually on left side of the face. The cause may be associated with a malfunction of the sympathetic nervous system, with or without neurological symptoms. Atrophy usually begins in first and second decades of life and progresses gradually over several years. There is no definitive management for this condition, but once the condition is stabilized, reconstructive surgery of the damaged skin and soft tissue can correct the disproportion. Additionally, with the extensive systemic involvement in such a condition, a multidisciplinary approach is needed because of its devastating effects on the entire body, treatment requires a multidisciplinary approach. Further research is needed to clearly understand the etiology and provide patients with accurate treatment plans. The cause of PRS is unknown however certain resources mention its relation to autoimmune conditions. A multidisciplinary approach is essential in such patients to enhance their quality of life. The objective of this article is to present an insight into the etiology of PRS with a case report of a 9-year-old male patient, who was diagnosed with PRS and developed progressive hemifacial atrophy without neurological manifestations.

Keywords: Parry Romberg Syndrome (PRS), Hemifacial atrophy, Autoimmune, Scleroderma, Oral manifestations.

INTRODUCTION

Parry-Romberg syndrome, also referred to as Romberg's disease or progressive hemifacial atrophy, is an uncommon condition that causes the gradual wasting away of the skin and underlying tissues on one side of the face. It was first described independently by Caleb Hillier Parry in 1825 and Moritz Heinrich Romberg in 1846[1]. PRS predominantly affects children and young adults, with an estimated occurrence rate of 1 in 700,000 individuals. It most often affects the left side and tends to be more prevalent in women than men[2].

This syndrome is usually associated with depressed vertical scar along the centre of forehead and is known as linear scleroderma or 'En coup de sabre'[3]. Recent studies concluded that these two conditions are a part of the spectrum of localized scleroderma and often co-exist. This syndrome exhibits numerous intraoral and extraoral features. Although dental abnormalities like malocclusion, tooth eruption disturbances and periodontal disease were observed in PRS, they are less frequently reported.

Based on clinical, laboratory and radiographical investigations a diagnosis of early stage of PRS with En coup de sabre has been made. Certain dental findings like dental malocclusion retained deciduous teeth, reduced salivary flow with periodontal disease has been noted. In this article we report a case of PRS along with its dental manifestation.

CASE PRESENTATION

A 9-year-old male child with a chief complaint of mobile tooth in upper front tooth region presented to Department of Pedodontics and Preventive Dentistry, UCMS and GTB Hospital, Delhi. The history of the patient revealed that the patient had trauma while playing two years back, at the age of 7 years, and since then, the patient had slow "shrinking" of left side of the face, which was noted by his family members. He had no relevant medical, dental, or family history.

During the clinical examination, the patient showed normal vital signs and overall health. He was alert and in good condition, with no indications of mental or psychological issues. Mild slurring speech was noted.

Routine blood tests, antibody testing, radiographs and CT scans were performed. Patient was negative for antinuclear antibodies. CT of brain showed no significant abnormality. X ray of face and skull showed posterior axial line and spino laminar line was disrupted. Histopathology section shows orthokeratosis, mild thinning of epidermis with flattening of rete ridges. The dermis shows fibrosis with perivascular inflammation.

On extraoral examination, there was significant atrophy and a scar-like defect (coup de saber) at the centre of forehead and left cheek region (Figure1).

On intraoral examination, grade 3 mobility observed irt 61 (Figure 2a), prominent palatal rugae, midpalatine raphae and incisive papilla was observed (Figure 2b). Ankyloglossia was noted [Figure 2c,d].

Moreover, an orthopantomogram (OPG) showed delayed tooth eruption, malocclusion due to dental crowding and impacted incisors [Figure 3].

At the first scheduled appointment, preventive therapy was advocated. Fluoride gel was applied and the patient was referred for physician's consent for the extraction of 61. At the second appointment, extraction of 61 was done, following the physician's consent [Figure 4]. The treatment aimed to improve the oral condition of the patient compared to what it was before. After a week, the patient was called for follow-up. Healing was checked and found to be satisfactory. Oral prophylaxis was performed and pit and fissure sealants placed.

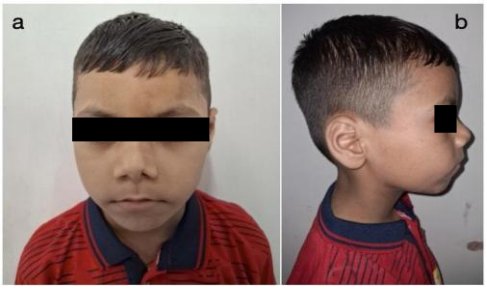

Figure 1a, b: Significant atrophy and a scar-like defect (coup de saber) at the centre of forehead and left cheek region

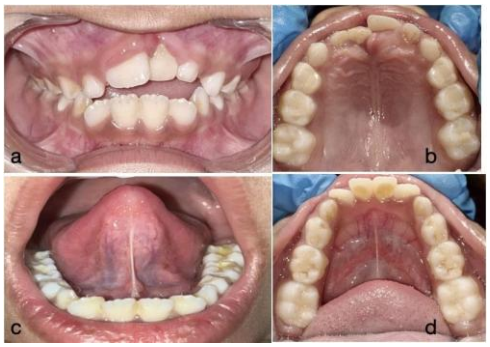

Figure 2 a: Intraoral examination reveals Grade 3 mobility in relation to 61
2b: Prominent palatal rugae, midpalatine raphae and incisive papilla
2c, d. Ankyloglossia was noted.

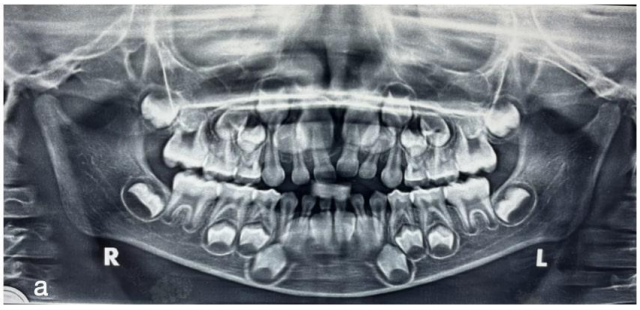

Figure 3: Orthopantomogram (OPG) showing delayed tooth eruption, malocclusion due to dental crowding and impacted incisors.

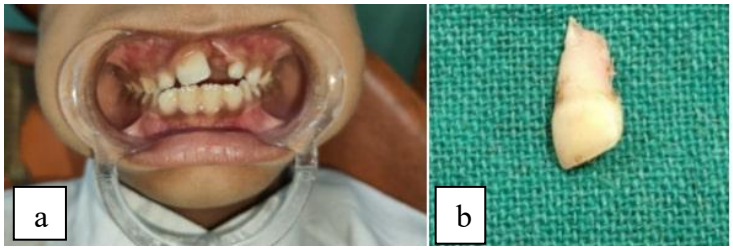

Figure 4 a,b: Extraction of 61 was done

DISCUSSION

Parry Romberg syndrome is a rare, acquired, neurocutaneous syndrome characterised by a progressive but self-limiting atrophy of the skin and subcutaneous tissue on one side of the face[4]. The condition is most commonly seen in association with linear scleroderma. The condition is more commonly seen in females and affects mostly left side of the face leading to facial asymmetry. The prevalence rate is atleast 1 per 70,000 in the general population[5]. The pathogenesis of PRS is related to brain disorders in fat metabolism, local facial trauma, endocrine disorders, autoimmunity, heredity, decreased or increased sympathetic nervous system activity, trigeminal nerve abnormalities and viral infections[6]. The striking feature is hemiatrophy of the facial tissues typically fat, but variably skin, other connective tissue, and sometimes bone. But in our case, since the patient is young, we can notice a depressed vertical scar along the centre of forehead known as linear scleroderma or 'En coup de sabre and we can note that the lesion is not progressed because of his young age. Usually, the atrophy progresses slowly during the years and then becomes stable. In the case reported here, there is involvement of left side of the face. The most common clinical features include enophthalmos, deviation of mouth and nose to the affected side leading to facial asymmetry, unilateral exposition of teeth, dry and hyperpigmentation of the

skin[7]. Ocular features like enophthalmos are due to fat loss around the orbit and is not observed in our patient. Atrophy of fat around the ear, alopecia on the involved side is also a presenting feature[8]. The teeth on the affected side have a normal clinical appearance and are vital. Jaw hypoplasia, deformity of mouth, nose on the affected side, atrophy of tongue papilla, left lateral border, lingual atrophy, oral mucosal changes, defects in tooth eruption, and delayed tooth formation and maturation, dental malocclusion are some of the oral manifestations seen in Parry Romberg syndrome[9]. Treatment of atrophy will include remodelling by adipose tissue, some therapeutic options like autogenous fat grafts, cartilage grafts, silicne infections, and inorganic implants[10]. In our case, the patient had early features of PRS, so frequent monitoring of condition is required to avoid complications. Affected patients should have multidisciplinary attendance of plastic surgeon, physicians, dental surgeon, phonoaudiologists and psychologists to give a better aesthetic to patient.

CONCLUSION

PRS is an exceptionally uncommon condition that leads to gradual hemifacial atrophy and may also be linked to linear scleroderma. It can result in severe consequences due to its multiple systemic effects, making a multidisciplinary approach necessary. Further research is essential to determine the underlying cause and develop a clear management guideline.

Declaration by Authors

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Conflict of Interest: The authors declare no conflict of interest.

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