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ABSTRACT:

Progressive Hemifacial Atrophy, also known as Parry-Romberg Syndrome, is an uncommon degenerative and poorly understood condition. It is characterized by a slow and progressive atrophy affecting mostly one side of the face along with ophthalmic and neurological complications. The disease is said to overlap with linear scleroderma. No dearth of speculation has been unturned to find the causative agent since it was first described. A case report of 35 years old, female is presented.

Key words: Progressive, Hemi-facial, Atrophy, Parry-Romberg, Romberg,

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INTRODUCTION:

Among the diseases typically presenting unilaterally in orofacial region, one of the rare diseases is progressive hemifacial atrophy, also known as Parry-Romberg Syndrome (PRS). It is characterized by a progressive atrophy of the skin and subcutaneous tissue on one side of the face [1]. This rare condition was first described by Calef Parry in 1825 [2], later by Moritz Romberg in 1846 [3]. Romberg named the process "Trophoneurosis of the face," emphasizing the implication of the nervous system in this syndrome [4]. The

worldwide prevalence is unknown [5]; though Stone suggested one per half a million births [6]. The syndrome is more common in females, with onset in the first or second decade followed by a 'burning out' of the atrophic process and subsequent stability [7]. There is a predilection for the left side of the face [7]. Many causes have been proposed such as facial or body trauma, compression of the cervical sympathetic nervous system, infection including Borrelia burgdorferi, autoimmune neurovascular disorders [7- 9].

PRS is mostly diagnosed based on clinical signs and symptoms, characterized by hemifacial atrophy of the face, coup de sabre on forehead and with many oral dental manifestations such as atrophy of tongue, root resorption, tooth dilacerations [9]. Skin changes of PRS often resemble "en coup de sabre" of linear scleroderma [10].

This case study describes the clinical features of Parry Romberg Syndrome in a middle aged female. Written informed consent was obtained from the patient and ethical clearance and permission was obtained from the institutional ethical committee for the study.

CASE REPORT:

A 35 year old female patient reported to the Department of Oral Medicine and Radiology, with the complaint of gradual stiffness and hollowing of the right side of face since 6-7 years. There was no relevant medical and family history. On examination facial asymmetry was detected due to the hollowing of cheek and chin on the right side. The right eye was mildly depressed in the socket, and atrophy of facial skin and lower lip on the right side was observed. Depression of nasal bone on the right side was noticed. Because of the loss of subcutaneous fat there was prominent bony ridge on the ipsilateral side, when compared to the normal side.

Hyperpigmentation was noticed on forehead and the chin region. Localised vitiligo patches were seen in the right ear. There was atrophy of temporalis, buccinator, masseter muscles and prominent zygomatic arch on the right side (Fig 1A). Demarcation between the affected and unaffected side was evident in the lower one third of the face (Fig 1B).

On palpation muscles were very firm, nontender. The intraoral examination revealed the atrophy of tongue on the right side. All teeth appeared normal. Based on the clinical features, a diagnosis of PRS was made. Routine blood investigations were carried out which revealed all values within normal limits. Orthopantomogram and posterior-anterior cephalogram was done (Figs 2 & 3).

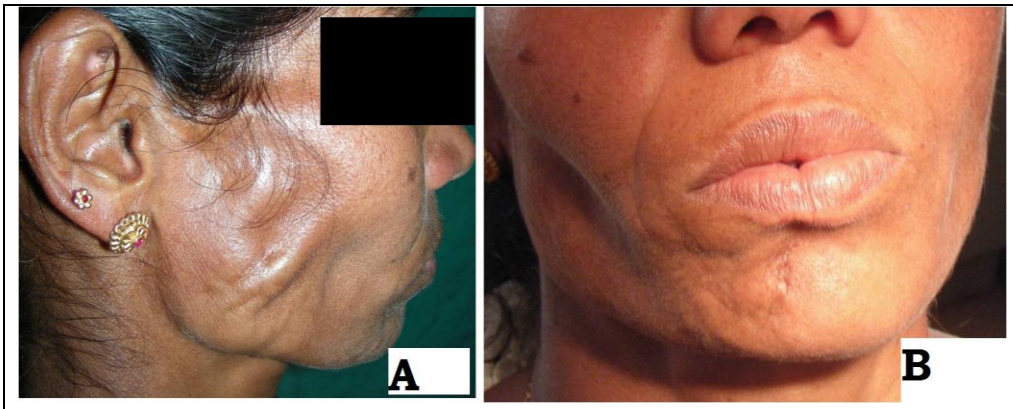

Fig. 1A: Extra oral view showing atrophy of muscles, vitiligo patches on the right ear.
Fig. 1B: Wrinkled appearance of the overlying skin due to lack of subcutaneaous fat on the right side.

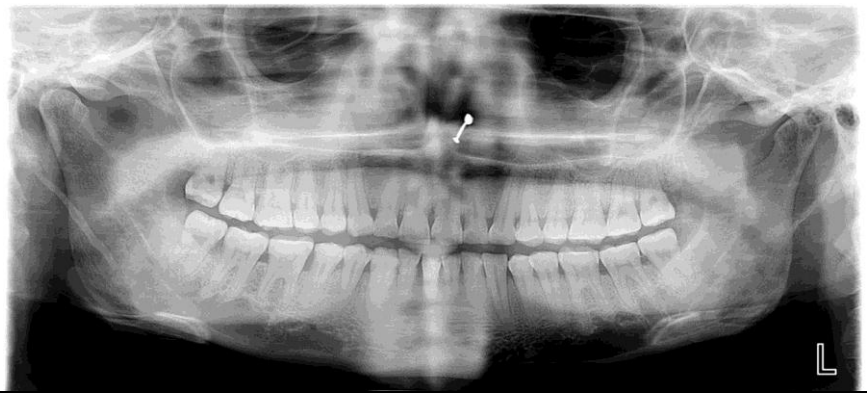

Fig 2: Orthopantomogram shows no abnormality.

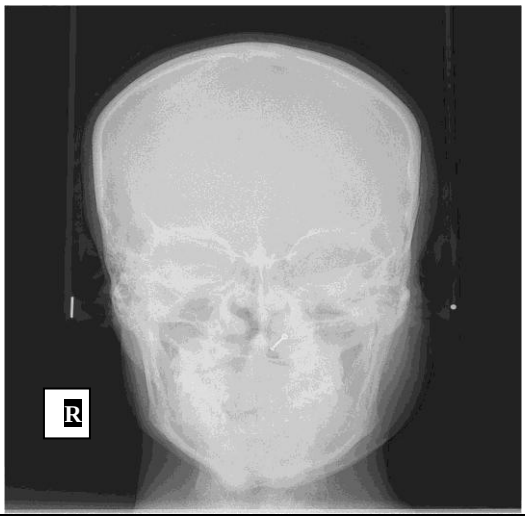

Fig. 3: PA cephalogram shows asymmetry of the jaws with severe atrophy of soft tissues on right side

Except for small maxillary sinus on right side no abnormality was observed in orthopantomogram. The posterior-anterior cephalograms revealed asymmetry of the jaws with severe atrophy of soft tissues on right side. Alloplastic implants or omentum graft suggested to the patient for correction of aesthetic deformity, but the patient could not afford it for a financial reasons. Psychological counseling was done for the patient as well as her relatives. They were assured of the benignness of the condition and explained about the course of the disease. The patient was followed up for a period of six months and no obvious changes were noticed.

DISCUSSION:

Hemi-facial atrophy is a rare disorder characterized by slowly progressive self-limited degeneration of the soft tissues of one half of the face (hemifacial atrophy) including dermis, subcutaneous tissue, fat, cartilage, and sometimes bone [11]. This condition is rarely manifested bilaterally. Along with the aesthetic abnormality there is functional disturbance often involving the eyes, neurological abnormalities. The disease may be associated to neurological abnormalities such as epilepsy, facial pain and migraine. Half of patients have eye or vision problems. Some develop autoimmune disorders like vitiligo and thyroid disease [5]. Symptoms and physical findings associated with PRS usually become apparent during the first decade or during the early second decade of life [12]. In some cases, the disorder is apparent at birth. Though in this case the patient noticed the abnormality in her third decade of life and in contradiction to most of the cases, the right side was affected. The degree of facial deformity is usually more severe if atrophy begins in the first decade, as growth is rapid during this time [9]. In the present case facial disfigurement has forced the patient to seek dental consultation rather than any functional disturbance. Clinically, the skin can be dry and hyperpigmented which is also seen in presented case. Some patients present with a demarcation line between normal and abnormal skin, known as “coup de sabre” [13], which was not seen in our case.

Atrophy of underlying muscles, bones, and cartilage is responsible for the typically aged appearance of the patient. Though absent in the present case, blanching of the hair or bald patches on the scalp and loss of eyelashes and eyebrows may occur [14].

The skin overlying affected areas may become hyperpigmented with patches of vitiligo. Localised vitiligo patches were seen in the right ear. The ear can be misshapen, smaller than

normal, or bat-eared as a result of tissue atrophy, none of these features were seen in the present case [10]. Anterior to posterior growth can be altered by deviation of the entire middle and lower third of the face to the affected side, carrying the nose and chin with it. When the lips are involved, they can be showing a unilateral dentition. There may be atrophy of the upper lip and tongue, which was observed in our case [10]. Intraoral soft tissues and muscles of mastication can be affected, but they usually function normally. A decreased depth and width of the retromolar region of the pharynx may occur. Delayed eruption, missing teeth, deficient root development, or resorption of the roots of teeth, dilacerations of the tooth of affected side, have been reported [9]. The mandible and alveolar ridge may be smaller on the affected side. The mandibular body may be shorter than normal, the ramus can be deficient vertically, and there can be a delay of mandibular angle development. The jaw disturbances can result in a unilateral malocclusion on the affected side and deviation of the facial and dental midlines. Spontaneous fracture of the mandible has also been reported [9]. Rarely there may be associated involuntary jaw closure [15]. In the current case no dental abnormality was observed in the patient.

Some individuals may experience other associated symptoms like neurological abnormalities such as epilepsy, migraine and facial pain, trigeminal neuralgias, facial paresthesias, abnormalities of hair like alopecia, ophthalmic disorders such as heterochromia, uveitis, enophthalmos due to atrophy of fat around the eye [6]. Enophthalmosis was evident in the present case.

There is an age-old argument about the relationship between linear scleroderma and PRS [16]. Controversy remains regarding the relationship of PRS to linear scleroderma, mainly about considering them as distinct entity or including PRS in the spectrum of linear scleroderma [5]. In linear scleroderma, the lesions usually are limited to the skin and to the subcutaneous tissue beneath the cutaneous lesions; rarely, however, the underlying muscles and bones are also affected. In PRS, the atrophy is deeper than that seen in linear scleroderma. The skin is less often bound down. More extensive involvement of the lower face is another feature of PRS [17].

Skin biopsy of PRS is indistinguishable from that of linear scleroderma [16]. The histopathological characteristics of linear scleroderma consist of two stages including, early inflammatory and late sclerosis stages. There are no inflammatory changes in PRS, even in its early stages. The epidermis is normal; the collagen bundles in the reticular dermis often appear thickened and closely packed and stain more deeply eosinophilic than

in normal skin; the eccrine glands appears markedly atrophied [18, 19]. There is no clear cut difference in autoantibodies between localized scleroderma and Parry Romberg syndrome. The coexistence of auto-antibodies such as anti-dsDNA in both the disorders may confirm that Parry Romberg syndrome and linear scleoderma en coup de sabre represent overlapping conditions [16].

Management of hemifacial atrophy is multidisciplinary, including reconstructive surgeries, orthodontic treatment, and psychosocial support, management of ophthalmic and neurologic complications [20]. Reconstructive facial surgery employing techniques including lipo-injection, dermis fat grafting, silicone implants, tissue transfer are needed to repair wasted tissue and diminish facial asymmetry [6, 13] however the outcome is unpredictable. Orthodontic treatment (corrective osteotomy) can help in the correction of any associated mandibular malformation [21].

None of these surgical treatments could be carried out in our patient because of financial reasons. Along with the surgical correction it is of paramount importance that these patients are provided with psychological support as it is very hard to cope with the gross facial deformity [9].

CONCLUSIONS:

PRS is an uncommon condition, which manifest as atrophy of one side of the face. The pathophysiology of the syndrome remains unknown. This case reports highlights the features of progressive hemifacial atrophy.

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The authors report no conflicts of interest related to this study.

REFERENCES:

1. Gulati S, Jain V, Garg G. Parry Romberg syndrome. Indian J Pediatr. 2006; 73(5):448-9.
2. Parry CH. Collections from unpublished papers. London: Unterwood, 1825.
3. Romberg MH. Trophoneurosen. In: Klinische Ergebnisse Berlin: Forstner, 1846:75–81.
4. DeFelipe J, Segura T, Arellano JI, Merchán A, DeFelipe-Oroquieta J, Martín P et al. neuropathological findings in a patient with epilepsy and the Parry–Romberg Syndrome. Epilepsia2001; 42(9):1198–1203.
5. Slimani S, Hounas F, Rezig A L. Multiple linear sclerodermas with a diffuse Parry–Romberg syndrome. Joint Bone Spine 2009; 76(1): 114-116.
6. Stone Jon. Parry Romberg syndrome clinical characteristics of 205

respondents. Neurology 2003; 61:674e6.
7. Moko SB, Mistry Y, Maurice T, de Chalain B. Parry–Romberg syndrome: intracranial MRI appearances. Journal of Cranio-Maxillofacial Surgery 2003; 31: 321–324.
8. Lonchampt P, Emile J, Pelier-Cady MC, et al. Central sympathetic dysregulation and immunological abnormalities in a case of progressive facial hemiatrophy (Parry-Romberg disease). Clinical Autonomic Research 1995; 5:199e204.
9. O'Flynn S and Kinirons M. Parry–Romberg syndrome: a report of the dental findings in a child followed up for 9 years. International Journal of Paediatric Dentistry 2006; 16: 297–301.
10. Rangare RA, Babu GS, Thomas PS, Shetty RS. Parry-Romberg Syndrome: a rare case report. J Oral Maxillofac Res 2011; 2(2):e5.
11. Miller MT, Spencer MA. Progressive hemifacial atrophy: a natural history study. Transactions of the american ophthalmological society 1995; 73: 203–15
12. Rogers BO. Progressive facial hemiatrophy: Romberg's disease: a review of 772 cases. Proceedings of the 3rd International conference on plastic surgery. Exerpta Medica ICS 1964; 66: 681–689.
13. da Silva-Pinheiro TP, da Silva C C, da Silveira CS, Ereno-Botelho PC, Rodrigues-Pinheiro MG, Viana-Pinheiro JJ. Progressive hemifacial atrophy – Case report. Med Oral Patol Oral Cir Bucal 2006; 11:E112-e4.
14. Zafarulla MYM. Progressive hemifacial atrophy: a case report. British Journal of Ophthalmology 1985; 69: 545–547.
15. KS Anand KS, Sabhnani ST, Prasad A. Parry-Romberg Syndrome with Involuntary Jaw Closure: An Extremely Rare Association. JIACM 2006; 7(3): 258-60.
16. M Gonul, B Dogan, Y Izci, G Varol. Parry–Romberg syndrome in association with anti-dsDNA antibodies: a case report. JEADV 2005; 19: 740–742.
17. Menni S, Marzano AV, Passoni E. Neurologic abnormalities in two patients with facial hemiatrophy and sclerosis coexisting with morphea. Pediatr Dermatol 1997; 14: 113–116.
18. Lever WF, Schaumburg-Lever G. Histopathology of the Skin 7th edn. JB Lippincott Co., Philadelphia, 1990: 511-12.
19. Fleischmajer R, Nedwich A. Generalized morphea. I. Histology of dermis and subcutaneous tissue. Arch Dermatol 1972; 106: 509–514.
20. Waters MF, Bhidayasiri R, Shields WD. Favorable longitudinal outcome in a patient with Parry–Romberg syndrome. Acta Neurol Scand 2005: 112: 192–193.
21. Kurian K, Shanmugam S, Mathew B, Elongavan. Facial hemiatrophy – a report of 5 cases. Indian J Dent Res 2003; 14 (4): 238-45.