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Progressive Hemifacial Atrophy: A Case Report

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Abstract

Progressive facial atrophy also known as Parry-Romberg syndrome is a rare disorder was first described by Romberg in 1846 characterized by slowly progressive atrophy of the soft tissues of essentially half face, which is characterized by progressive wasting of subcutaneous fat, sometimes accompanied by atrophy of skin, cartilage, bone and muscle, is usually confined to one side of face and cranium. It is usually accompanied by contra lateral Jacksonian epilepsy, trigeminal neuralgia, and changes in the eyes and hair. The exact etiology is unknown although sympathetic system dysfunction, autoimmune disorders, focal scleroderma, trauma and genetic factors have been suggested. Here, a case is presented of a patient having marked left-sided facial asymmetry with atrophy without other systemic manifestation.

Keywords: Progressive hemifacial atrophy, Facial atrophy, Hemifacial atrophy, Parry-Romberg syndrome (PRS), Romberg's syndrome, Morphea

Introduction

Progressive hemifacial atrophy, also known as Parry–Romberg syndrome (PRS), was initially described by Parry in 1825 and later by Romberg in 1846.[1,2] Other names used to describe this disorder include Parry-Romberg syndrome, idiopathic hemifacial atrophy, progressive facial hemiatrophy (PFH), and Romberg's syndrome.[2] This syndrome is a rare degenerative condition characterized by slow and progressive but self-limited unilateral atrophy of facial tissues, including muscles, bones, skin, and cartilage. It is caused by the

gradual loss of subcutaneous fat, often accompanied by atrophy of the skin, cartilage, bone, and muscle. This condition leads to aesthetic, functional, and psychological problems due to facial asymmetry.[1,3,4]

The etiology of PRS remains unknown, although various factors have been suggested to play a role in its pathogenesis, including trauma, viral infections, genetic factors, autoimmunity, endocrine disturbances, peripheral trigeminal neuritis, increased cervical sympathetic nerve activity, and cerebral disturbance of fat metabolism.[1,3]

It is a developmental craniofacial disorder associated with different systemic manifestations. In particular, PRS is associated with maxillofacial manifestations (such as wasting of masticatory muscles, delayed ipsilateral tooth eruption, unilateral tongue atrophy, and jaw hypoplasia), neurological abnormalities (including trigeminal neuralgia, migraine, and seizures), and ophthalmologic abnormalities (such as enophthalmos, glaucoma, endothelial precipitates, Horner's syndrome, and ophthalmoplegia). Moreover, it is frequently associated with linear scleroderma and referred to as en coup de sabre.[1,5] The term "en coup de sabre morphea" refers to a lesion of linear morphea generally located in the frontoparietal scalp and/or the paramedian forehead, often resembling a stroke from a sword. It may extend down the face. The relationship between these two entities is not entirely clear, although studies have suggested a close relationship exists.[6]

This case report presents a 20-year-old female who manifested extensive left-sided facial asymmetry with atrophy, along with en coup de sabre morphea and oral submucous fibrosis.

Case Description

A 20-year-old female reported to the oral medicine and radiology department with a chief complaint of facial asymmetry and intermittent occasional headaches on the left side. The patient had been relatively asymptomatic before 6-7 years, after which she noticed mild depression and gradual progressive left-sided facial atrophy, accompanied by a decrease in mouth opening without pain, so she visited multiple private hospitals where oral medication was given along with physiotherapy for mouth opening. She didn't get relief so; she visited to OMR department for needful.

There was a history of trauma approximately 8-9 years ago while playing in sports activities, as the patient was a national-level player of dodgeball and handball. There was no history of tooth extraction, seizures, or lancinating pain suggestive of trigeminal neuralgia. The patient denied experiencing joint pains, rash, dyspnoea, double vision, or neurological deficits in the extremities. Family history was non-contributory. Additionally, the patient had a habit of chewing pan masala containing tobacco, betel nut, and lime for approximately 5 years, 2-3 times per day, lasting 2-3 minutes each time, which ceased about 6-7 years ago.

Extraoral examination revealed marked facial asymmetry with a decrease in fullness on the left side of the face. There was evident deformity due to depression and deviation of the nose and lips on the left side. A coup de sabre was observed in the left side of the mid-face region near the left ala of the nose. No hyperpigmentation was observed on the affected skin, and the texture of the skin appeared normal. The patient's mouth opening was measured at 13 mm (**Figure 1**). **Intraorally**, blanching was observed on the bilateral buccal mucosa. Restricted movement of the tongue and soft palate was noted. Lower anterior crowding was observed. There was no evidence of atrophy on the tongue or any other oral structure. All teeth appeared normal and clinically vital. Palpation revealed fibrous

bands on the bilateral buccal mucosa over the molar and premolar region. The patient was in good overall health condition, without any systemic illness that could explain the cause of the facial atrophy (**Figure 2**).

Radiographically, on panoramic radiograph (OPG) and cone beam computed tomography (CBCT), normal crown and root length ratio, normal periodontal ligament (PDL) space, and intact lamina dura were observed. A normal trabecular pattern of bone was also noted. However, there was an alteration in the size and shape of the condyle on the left side (**Figure 3**). The MRI of the left side of the face revealed atrophy of subcutaneous fat, retromolar fat, parotid gland, and masseter muscle (**Figure 4**).

Based on these clinical and radiographic findings, moderate-stage hemifacial atrophy on the left side with oral submucous fibrosis (OSMF) was diagnosed. Treatment initiation involved intralesional injection of corticosteroids (2 mL Dexamethasone + 1500 IU hyaluronidase) once a week for 6 weeks along with supportive supplements (Multivitamins and Antioxidants). After 6 weeks, the patient's mouth opening had increased up to 28.12 mm along with improvement in facial aesthetics (**Figure 5**). The patient was satisfied with the treatment, and a referral was made to the dermatology department for further evaluation and management as needed.

Discussion

Progressive hemifacial atrophy, also known as Parry-Romberg syndrome (PRS), is a rare neurocutaneous disorder of unknown origin. It is characterized by slowly progressive facial hemiatrophy, affecting all tissue layers from the skin to the bone. PRS has a prevalence rate of 1 in 70,000 with no significant difference in rates observed between different ethnic groups. The condition predominantly affects females and typically involves the left side of the face. In most cases (71%), the onset of the disease occurs before the age of 15 years, with only 8% experiencing onset after the age of 25 years.[4,5] A linear lesion of morphea that is classically located on the paramedian aspect of the forehead, en coup de sabre morphea, has been associated with Parry-Romberg syndrome as well, although these two entities are not clearly related.[6,7] The presented case demonstrates a similar pattern, being a 20-year-old female with onset of atrophy occurring approximately 6-7 years prior. The patient exhibits feature consistent with PRS, including en coup de sabre and deviation of the nose and lips on the left side, along with oral submucous fibrosis.

According to the literature, Parry-Romberg syndrome (PRS) is associated with neurological manifestations such as migraine, trigeminal neuralgia (TN), partial seizures, and occasionally neurological deficits in the extremities and abnormal movements.[2,3,8] However, our presented case demonstrates facial asymmetry due to atrophy on the left side, along with occasional headaches on the left side, without other systemic manifestations such as trigeminal neuralgia or epilepsy.

The etiopathogenetic mechanism for progressive facial atrophy is controversial. Literature analysis has revealed several potential causative factors including sympathetic dysfunction, local trauma, localized scleroderma, autoimmunity, and infection.[3] The patient in this case report has a history of trauma while playing sports, which aligns with one of the aforementioned factors that could contribute to the development of progressive facial atrophy. The differential diagnosis of progressive hemifacial atrophy (PHA) includes congenital hemiatrophy and primary hemifacial hypertrophy, such as Barraquer-Simons syndrome. Congenital hemiatrophy is present since birth, whereas hemifacial hypertrophy is characterized by asymmetric enlargement of half of the

head without enlargement of other body parts. Though unilateral face enlargement is observed instead of atrophy as seen in PHA, this disorder may still be considered in the clinical differential diagnosis of an asymmetric unilateral facial deformity. Baraquer-Simons syndrome, on the other hand, is an acquired partial progressive cephalothoracic lipodystrophy. It presents with a gradual onset of symmetrical bilateral subcutaneous fat loss from the face, neck, upper extremities, thorax, and abdomen but spares the lower extremities.[2]

According to the literature, the disease is self-limiting and has no definite cure. Patients affected should have multidisciplinary care, involving experts such as dermatologists, dentists, and psychologists. Treatment typically focuses on repositioning adipose tissue lost due to atrophy and stoppage of the active disease process.[1,2,4] It encompasses both medical and surgical management. Medical management often includes the use of corticosteroids (topical and intralesional), antioxidants, and immunosuppressants such as methotrexate and retinoids. Surgical options include facial grafts, muscle grafts, pedicle flaps, microvascular flaps, and free grafts, including injection of aspirated fat and alloplastic graft materials.[5]

In the present case, the chosen treatment modality was intralesional injection of corticosteroids, which is a commonly utilized approach for managing both progressive hemifacial atrophy (PHA) and oral submucous fibrosis (OSMF). Following this, there was improvement in both mouth opening and facial esthetics, and patent was satisfied with the treatment outcome.

Conclusion

This case highlights a 20-year-old female with Parry-Romberg syndrome, presenting with facial asymmetry and en coup de sabre morphea. Trauma history may contribute to the syndrome's development. Multidisciplinary management involving dermatologists, dentists, and psychologists is crucial, with treatment options including corticosteroids, antioxidants, and surgical interventions to address aesthetic and functional concerns associated with the condition.

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Legend Figures

Figure 1: Extra oral findings (facial asymmetry due to atrophy of the left side of face and Coup de sabre)

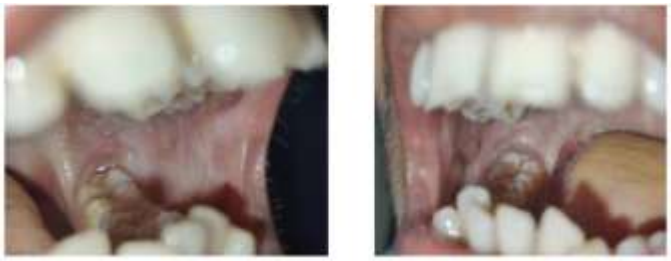

Figure 2: Intra oral findings (blanching present on bilateral buccal mucosa, no erosion or ulceration, normal size, shape and structure of teeth

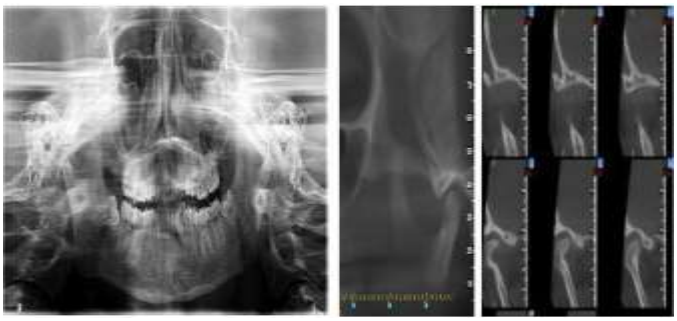

Figure 3: OPG and CBCT shows normal crown: root ratio, normal PDL space and intact laminadura.. Normal trebecular pattern. Altered size and shape of the condyle

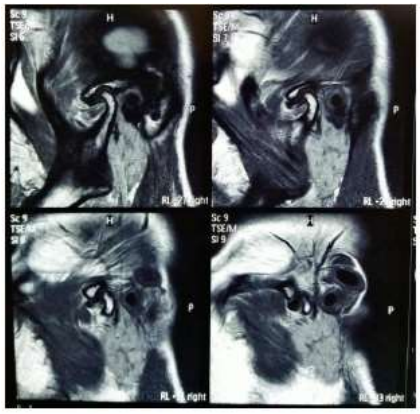

Figure 4: MRI of left side of face showed atrophy of subcutaneous fat, retromolar fat, parotid gland and messeter muscle

Figure 5 A: Before treatment: Left facial atrophy and restricted mouth opening

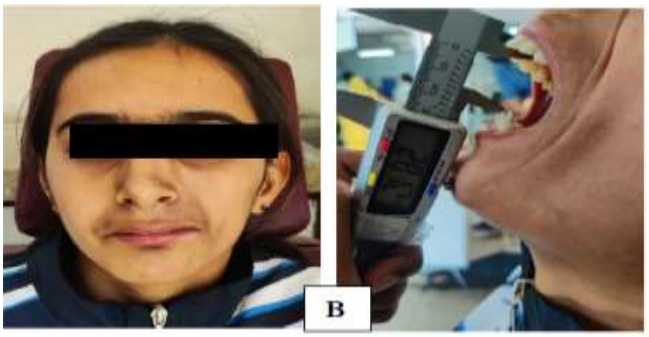

Figure 5 B: After treatment: Improved facial aesthetics and increased mouth opening

Abbreviations: Parry-Romberg syndrome (PRS), progressive facial hemiatrophy (PFH), periodontal ligament (PDL), oral submucous fibrosis (OSMF), trigeminal neuralgia (TN)