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Case Report

Progressive hemifacial atrophy

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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 but self-limited atrophy affecting one side of the face. The incidence and the cause of this alteration are unknown. A cerebral disturbance of fat metabolism has been proposed as a primary cause. Possible factors that are involved in the pathogenesis include trauma, viral infections, heredity, endocrine disturbances and auto-immunity. The most common complications that appear in association to this disorder are: trigeminal neuralgia, facial paresthesia, severe headache and epilepsy. Characteristically, the atrophy progresses slowly for several years and, it becomes stable. The objective of this work is, through the presentation of a clinical case, to accomplish a literature review concerning general characteristics, etiology, physiopathology and treatment of progressive hemifacial atrophy.

Key Words: Progressive hemifacial atrophy, Parry-Romberg Syndrome, Romberg's disease

INTRODUCTION

Progressive hemifacial atrophy, also known as Parry-Romberg syndrome, is an uncommon degenerative condition characterized by a slow and progressive but self-limited atrophy of facial tissues, generally unilateral, including muscles, bones, skin and cartilage. More than an aesthetic trouble, this illness brings several functional and psychological problems, when a "symmetric" face loses its identity.[1]

The first published reports on progressive hemifacial atrophy were attributed to Parry' in 1825 and Romberg in 1846.[2]

The incidence and cause of this alteration is unknown. A cerebral disturbance on fat metabolism has been proposed as a primary cause. Trauma, viral infections, endocrine disturbances, auto-immunity and heredity are believed to be associated with the pathogenesis of the disease.[3]

Frequently, the onset of this syndrome occurs in the first and second decades of life. Characteristically, the atrophy progress slowly for many years and then it becomes stable. This syndrome seems to have higher incidence in women. Ocular involvement is common and the most frequent manifestation is enophthalmos.[4]

It is usually accompanied by neurological complications like trigeminal neuralgia, partial seizures have found to be most common neurological complication.[5] Radiographically, the teeth on the involved side appear small and usually have short roots. The extension of the atrophy is frequently limited to one side of the face, and the ipsilateral involvement of body is rare.[4]

CASE REPORT

A 45-year-old male patient reported to our Department of Oral Medicine and Radiology, Vasantdada Patil Dental College and Hospital, Kavlapur, Tal-Miraj, Dist. Sangli, Maharashtra, India, with the chief complaint of facial asymmetry and missing teeth in the upper right posterior region.

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The patient had noticed shrinkage of the right side of the face when he was 20 years old. The patient had visited a local physician, who suggested cosmetic correction for the same, but could not carry out the treatment because of financial reasons. The patient's past dental, medical and family history were non-contributory.

On inspection facial asymmetry was detected on right side [Figure 1]. The face on the affected side appeared smaller. Eyes appeared normal and of the same size. The pinna of the ear on the right side was pulled towards the front giving it a bat ear appearance. Ala of nose on right side was small as compared to normal side [Figure 1].

The zygomatic bone appeared more prominent on right side due to depression in the cheek region.

On inspection the tongue was normal but on protrusion it deviated slightly towards the affected right side. The corner of mouth on right side was retracted. When patient was asked to clench, the prominence of the masseter muscle could not be palpated on right side.

On palpation, the ramus area and the depression below the zygomatic bone felt bony hard as there was absence of muscles. The skin on right side of the face was taut and not of the same texture when compared with the other side.

Local examination, measurements were taken from the nasion to the tragus, nasion to angle of mandible, and mid chin to tragus of both right and left side [Table 1]. The measurements showed hemifacial atrophy of the right side.

The intraoral examination revealed edentulous area in first quadrant [Figure 2]. The ridge in first quadrant was thin. Patient also gave history of absence of deciduous and permanent teeth in the first quadrant. Generalized periodontitis was seen with the remaining teeth. Patient also gives history of exfoliation of lower molars on right side 10 years back.

Based on the clinical features and facial measurements, a diagnosis of Parry-Romberg syndrome was made. Routine blood investigations were carried out which revealed all values within normal limits.

As a part of radiographic investigations an orthopantomogram, posterior-anterior cephalogram, transcranial, posterior-anterior chest X-ray and CT and MRI was made.

The orthopantomogram revealed that the changes in the mandible were more apparent than in maxilla. There was absence of maxillary teeth on right side. The affected right side of the jaw was small in overall dimension than the opposite side.

The condyle and coronoid process of mandible were normal in shape but smaller in size on right side. The body of the mandible was reduced in size on the affected side. Prominent antegonial notch on the affected side was evident [Figure 3]. The midline of the jaw was deviated towards the affected side. Transcranial radiograph and chest X-ray did not show any abnormality.

Table 1: Measurements between anatomical landmarks

Landmarks	Right side	Left side
Nasion to tragus	13.3	15
Nasion to angle of mandible	11.5	13
Mid chin to tragus	14	16

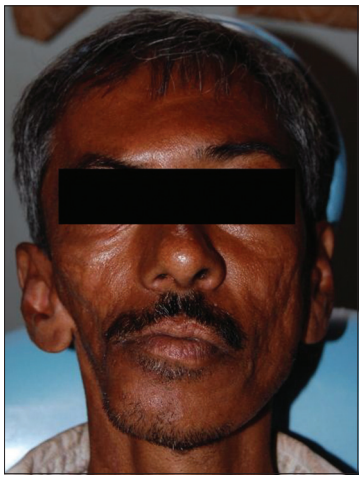

Figure 1: Extra oral photograph showing right side facial atrophy

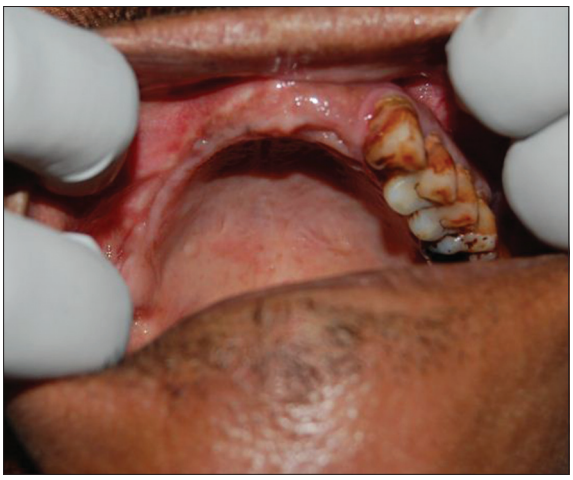

Figure 2: Intra oral photograph showing edentulous 1st quadrant

Computerized tomogram (CT) of head and neck showed, hypoplastic right mandible and its condyle, absence of right masseter muscle as well as absence of right parotid and submandibular salivary glands [Figures 4 and 5].

MRI also confirmed the absence of right masseter muscle and right parotid and submandibular salivary glands [Figure 6].

DISCUSSION

Parry-Romberg syndrome is an uncommon degenerative and poorly understood condition. It is characterized by a slow and progressive unilateral atrophy of the facial tissues, including muscles, bones and skin. The condition is more often found in female population and has predilection for the left side of the face, as a rare case we found it in a male patient and on right side. More than an aesthetic concern, this disease brings several functional and psychological problems due to asymmetry of the face. The main feature is hemiatrophy of the facial tissues, typically fat, but variably skin, other connective tissue, and sometimes bone. All these features were present in the case herein. The prevalence rate is estimated to be at least 1 per 700,000 in the general population.[6] Characteristically, the atrophy progresses slowly over many years and then becomes stable as in our case.

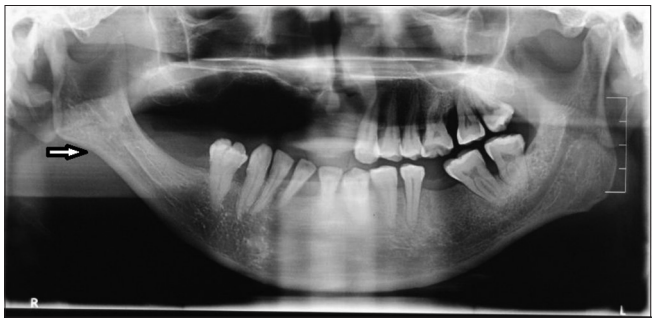

Figure 3: Orthopantomogram showing antegonial notch on right side

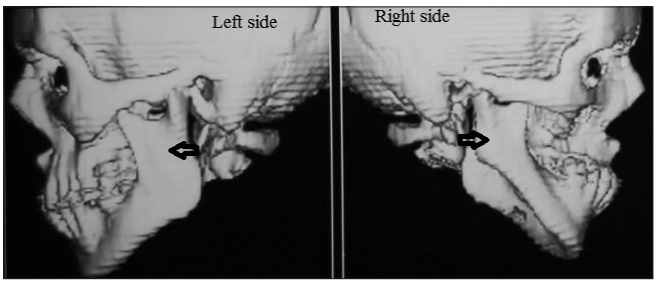

Figure 5: 3D reconstruction CT showing hypoplastic right mandible and condyle

The extension of atrophy is frequently limited to one side of the face, and the ipsilateral involvement of body is rare (10 to 23% of cases were described as being bilateral).[7] In the case presented here, there was involvement of only one side of the face.

The important features of this disease are enophthalmos, which was not evident. Deviation of tongue and comparatively less development of nose on the affected side were evident. Unilateral absence of maxillary teeth (first quadrant) was found in our case. Some patients present a demarcation line between normal and abnormal skin, known as "coup de sabre" (French term which means "cut of the sword")[1,6] which is not seen in our case. Thinning of the ear due to atrophy of the fat around it and forward pulling of pinna of ear giving it a bat ear appearance was a manifestation registered

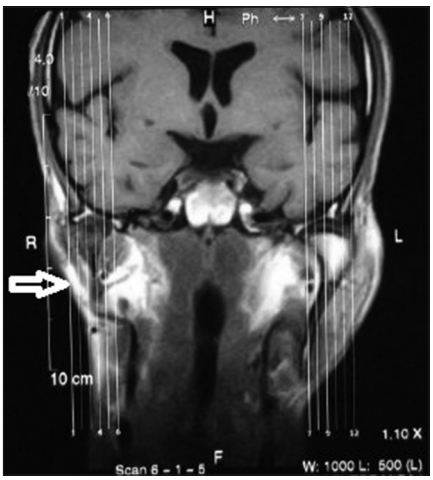

Figure 4: Coronal section of CT showing absence of masseter muscle on right side

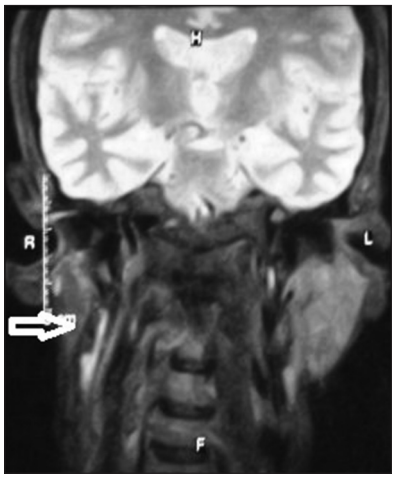

Figure 6: MRI showing absence of parotid and submandibular salivary glands

in our patient.[1] One of the striking features in our case was absence of masseter muscle, parotid and submandibular gland of the affected side due to which patient had decreased salivation. The neurological complications, such as trigeminal neuralgia, facial paresthesia, severe headache and contralateral epilepsy can also be present,[8,9] but were not diagnosed in our case. Our case showed most of the classical clinical manifestations of the disease [Table 2].

Radiographically, the teeth of patients with Parry-Romberg syndrome have short roots and appear small, when compared to the uninvolved side.[1] In our case complete absence of deciduous and permanent teeth in first quadrant were noted [Figure 2].

The treatment is usually based on reposition of adipose tissue that was lost due to atrophy. Autogenous fat grafts, cartilage grafts, silicon injections and prostheses, bovine collagen and inorganic implants are some alternatives to aesthetic correction of the atrophy.[10] The treatment modalities mentioned, resolve just momentarily the good appearance, whereas all the structure projected in the cosmetic surgery is lost with time, due to gravity action, and the patient usually requires new intervention.[1] Pan-facial volumization with autologous fat is an excellent tool for replacing volume and restoring contour to the aging face.[11] Our patient was suggested a treatment using alloplastic implants to improve the facial appearance, but could not undergo the treatment due to financial and logistical reasons.

Table 2: Comparison of the features seen in the syndrome with features seen in the case

Important features of Parry-Romberg syndrome	Features seen in our case
Facial asymmetry on affected side	Evident
Enophthalmos	Not evident
Atrophy of facial skin on affected side	Evident
Ala of the nose small on affected side	Evident
Pigmentation over the skin on affected side	Not evident
Angle of mouth retracted	Evident
Bat ear appearance on the affected side	Evident
Deficient eyebrows	Not evident
Zygomatic prominence	Evident
Prominent antegonial notch on affected side	Evident
Atrophy of masseter muscle	Absence of masseter muscle on right side
Roots of deciduous and permanent teeth are small and have short roots	Absence of parotid and submandibular salivary glands on right side Absence of deciduous teeth according to history given and permanent teeth in 1st quadrant according to clinical examination Hypoplastic right mandible and its condyle

CONCLUSION

Parry-Romberg syndrome is an uncommon condition, which manifests as atrophy of one side of the face. In most cases, Parry-Romberg syndrome appears to occur randomly for unknown reasons. The pathophysiology of the syndrome remains unknown. There is no definitive treatment for this condition but an attempt to use restorative plastic surgery which includes fat or silicone implants, flap/pedicle grafts, or bone implants can be done to improve facial disfigurement.

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