

Rare case of Parry-Romberg Syndrome (Hemi facial atrophy)

B. M. Karmacharya, R. M. Karmacharya

Department of Medicine, Dhulikhel Hospital, Dhulikhel, Nepal, Aarogya Aspataal, Naya Naikap, Kathmandu, Nepal

Correspondence to: Dr. Biraj Man Karmacharya, Dhulikhel Hospital

e-mail: reachrobin773@iom.edu.np

Abstract: Parry-Romberg syndrome is a rare syndrome that is more common in females. It is characterized by gradual onset and progressive atrophy of one half of face causing considerable disfigurement, blindness and orthodontic problems. The etiology and treatment has not been clarified yet ¹. We report a case of this rare syndrome.

Key Words: Hemi facial atrophy, Parry Romberg Syndrome

Introduction

54 year old female from Naikap presented to the out-patient department of 'Aarogya Aspataal' with features of acid peptic disease. But on examination, remarkable asymmetry in her face was noticed. There was atrophy of all the musculature, cartilage as well as bone on the right half of her face. She had developed difficulty in opening her mouth and had some disfigurement over right half of face when she was 25 years old. The disfigurement increased gradually with progressive decrease in the size of right half of her face. Over few years, she even had diminution of vision with persistent opening of the right eye. Slowly, her right half of jaw too decreased in size causing loosening and finally falling of the teeth. All these progressed over the next ten years and after that it came to a halt but by that time, there already was considerable disfigurement. Right eye had already gone blind with ptosis as well as huge corneal opacity and she lacked all the teeth of the right side. On further examination, right half of her face was smaller due to atrophy of muscles, cartilage as well as bone. There was no perception of light in the right side. Voluntary eye opening was absent, hearing was absent on right side and there was prosthesis for right half of jaw. Mouth opening and speech was however normal.

Discussion

Parry-Romberg syndrome is a rare disorder in which there is progressive atrophy of the skin and soft tissues of half of the face (hemi facial atrophy). It is more common in females than in males ². The onset of the disease usually begins between the ages of 5 and 15 years. This disease usually affects the tissues above the maxilla or between the nose and the upper corner of the lip and it may progress to the angle of the mouth, areas around the eye, the brow, the ear, and the neck. Sometimes it also affects the tongue, the roof of the mouth, and the gums. The affected side may be darkly pigmented. The affected eye may be sunken along with loss of vision. There may be graying of hair with hair loss. Problems with the retina and optic nerve may occur when the disease surrounds the eye causing blindness. There may be muscle atrophy on affected half of the face along with loss of bone.

Parry-Romberg syndrome is also accompanied by seizures and episodes of severe facial pain (trigeminal neuralgia). The progression often lasts for about 10 years, and then the process seems to enter a static phase.

Until now no treatment modalities have been known so as to stop the progression of atrophy. Usually only supportive

Parry-Romberg Syndrome

and symptomatic treatment are advised. Reconstruction is to be done in severe cases. This requires microvascular surgery to repair wasted tissue³. Usually surgery is done only after there is complete halt in the progression of atrophy. One to two years of waiting is recommended by most surgeons. Muscle or bone grafts may also be helpful.

Prognosis varies amongst patients with this disease. In some, there is only cosmetic defect without any affect on function whereas in others, the atrophy ends before the entire face is affected. There is lack of adequate researches due to rarity of this disease.

References

1. Wartenberg R: Progressive facial hemiatrophy. Arch. Neurol. Psychiat. 1945; **54**: 75-96.
2. Rogers B O: Progressive facial hemiatrophy; Romberg's disease: a review of 772 cases. Proc 3d Int Cong Plast Surg. Excerpta Medica ICS 1964; **66**: 681-689.
3. Muchnick RS, Aston SJ, Rees TD: Ocular manifestations and treatment of hemifacial atrophy. Am J Ophthalmol 1979; **88**: 889-897.

