



## CASE REPORT

### PARRY ROMBERG SYNDROME: A CASE REPORT

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#### ABSTRACT

Parry Romberg syndrome (PRS) is a rare disorder, characterized by unilateral facial atrophy of the skin, subcutaneous fat, muscles, cartilages, and even bone. Various systemic manifestations reported with this syndrome are neurologic, maxillofacial, and ophthalmologic. The etiology is still unknown. Herein we present a case report of 23 years young engineering student who had PRS and got a fracture of the right angle of his mandible after a road traffic accident. The treatment was challenging as he had nonunion at the fracture site.

#### INTRODUCTION

Parry Romberg Syndrome is a rare disorder of unknown etiology characterized by progressive hemifacial atrophy of subcutaneous fat followed by wasting of skin, muscles, cartilage, and bone. The associated neurological abnormalities are found in 20% of such cases.<sup>1</sup> Parry described it for first time in 1825 and Romberg in 1846.<sup>2</sup> The disease has an active phase (2 to 10 years) followed by burning of the atrophic process and subsequent stability.<sup>3</sup> The distribution of parry Romberg syndrome generally follows the pattern of sensory innervations of one or all the branches of the trigeminal nerve dermatomes. Bilateral manifestation has been reported in 5% to 10% of the cases. The incidence of female to male is 3:2.<sup>3</sup> Modiolus is the most common site where atrophy starts. There may be other associated findings related to neurologic, endocrinologic, maxillofacial, infectious, ophthalmologic, orthodontics, rheumatologic, cardiac and autoimmune.<sup>2</sup> Facial nerve palsy, trigeminal neuralgia, headaches, simple partial and generalized seizures and learning difficulties can be noted in such cases. A well-demarcating line between abnormal and normal skin is called en coup de sabre in these cases. Spontaneous fractures of the mandible have been reported because of ipsilateral reduction of mandibular

width and height. Parry Romberg Syndrome is rare entity so we are sharing our experience of treating the fracture of mandible in such patients. The treatment of facial bone fractures in such patients is very challenging because of atrophy of the facial bones. The purpose of presenting this case is to help other researchers and surgeons about this disease.

#### CASE REPORT

A 23 years old young boy presented to the Department of Oral and Maxillofacial Surgery, King Edward Medical University, Lahore, Pakistan with the chief complaint of non-union of the right angle of the mandible after road traffic accident 3 months back. He was operated upon for the fracture of the right angle of the mandible in some other hospital almost 3 months back. Open reduction and internal fixation with miniplate at Champy's line under general anesthesia was done. But there was nonunion at the fracture site. In his stay in our ward, he was diagnosed as a case of Parry Romberg syndrome.

According to the patient when he was 15 years old he started noticing the gradual atrophy of the right side of the face that kept on going still yet. He visited general practitioners who could not diagnose the case. By profession, he was the final

year student of engineering. On general physical examination, he was a young, thin and energetic boy who had no systemic comorbidity. On extra oral examination, he had hemifacial atrophy of right side with flat right cheek, altered ocular level and tip of nose deviated to the right side (Figure 1).



**Figure 1: Hemifacial atrophy on the right side**



**Figure 2: Fracture of the right angle of mandible**

There was a good demarcation line present on the chin and lips region between normal and abnormal skin. There was alopecia on the right side of the cheek. The skin of the right side of the face was normal in texture but there were foci of hyperpigmentation of facial skin. There was twitching of the muscle of the right cheek. According to patient twitching had increased after the road traffic accident and now it's obvious. On intraoral ex-

amination, there was mobility at the right angle of mandible region with deranged occlusion. There was no other significant finding. Informed consent was taken for surgery. Plain radiographs including a posteroanterior view of mandible and orthopantomogram were obtained (Figure 2).

Under general anesthesia using the right submandibular approach, fracture of the angle of mandible was exposed. Mini-plate was removed and fracture margins were made freshened and a reconstruction plate was applied at the fracture site. Maxillomandibular fixation (MMF) was done for four week (Figure 3).



**Figure 3: Post-operative result of the mandible**



**Figure 4: Follow-up after 5 years of treatment**

Patient was given the oral hygiene instructions and he was put on follow up. The problem was the clonic seizures of the right

cheek so the patient was given carbamazepine 400mg twice a day to control those seizures and a prolong 4 weeks of MMF was offered for better healing. He was advised to use the crepe bandage over the face to control those seizures to aid in healing. After 4 weeks MMF was removed. There was no mobility at the fracture site, mouth opening was adequate and occlusion was acceptable. After 6 months of this operation, the recon plate was removed to avoid compression atrophy of mandible. Five years follow up was kept to watch and control the disease process (Figure 4). Future recommended treatment to correct the asymmetry is fat grafting or free tissue transfer to the right side of the face.

## DISCUSSION

Parry Romberg Syndrome is a rare disorder with incidence of 1:70000 and it has a female predilection.<sup>4</sup> Mostly it affects the left side of the face and starts in second decade of life. Progression of the disease ranges from two to ten years and after that disease may cease itself.<sup>5</sup> In some cases atrophy accelerates in adolescence period or may worsen after the surgeries.<sup>6</sup> Primary cause of cerebral disturbance of fat metabolism has been proposed.<sup>7</sup> Other etiological factors include peripheral trigeminal neuritis, endocrine disturbance, lymphatic neurovasculitis, localized scleroderma, viral infection, heredity and autoimmunity.<sup>8</sup> Anatomic changes of Parry Romberg syndrome depends upon the time of the active disease period.

The diagnosis of Parry Romberg syndrome is mainly clinical and radiological investigations are only done to rule out other diseases. Radiological investigations can be done in PRS to see the extent of atrophy or for the comparison in follow-up visits. The radiological features may include the maxillary and mandibular

hypoplasia, atrophy of associated facial muscles, fat and skin.<sup>9</sup> The treatment of PRS is very challenging and demands multidisciplinary approach. The initial aim of the treatment is to stop the progression of the disease. For this methotrexate (15mg/m<sup>2</sup>) is given orally or subcutaneously once a week with combination of steroids (0.3-1mg/kg/week) orally/systemically and its first line of choice. If patients do not respond to the methotrexate the other drugs like hydroxychloroquine, mofetil, cyclosporine can be given.<sup>10</sup> Surgical management is mainly done after the cessation of disease and for correction of disfigurement. Among the surgical options there are dermal fat grafting, muscle flaps, free tissue transfer or bone augmentation.

Mazzeo et al<sup>3</sup> estimated the active disease period of his case from 3 to 7 years as the patient had atrophy of the roots of left maxillary canine and mandibular 1<sup>st</sup> and 2<sup>nd</sup> molars. In our case, no dental abnormality was found it could be because of the reason it started when all dentition has erupted. El-Kehday<sup>2</sup> reported that seizures are the most common neurologic manifestation in such patients and we also observed the local facial muscles seizures in our case. A long follow up is needed in such patients to see the extent of disfigurement or to observe any acceleration in later age.

## CONCLUSION

Parry Romberg is an uncommon syndrome with varied dental or other systemic manifestations. Dental involvement helps to determine the period of onset and duration of this disease process. General dentists may play their role in early diagnosis of Parry Romberg syndrome due to their knowledge of the growth and development of the oro-facial region.

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