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CASE REPORT

CLINICAL AND RADIOGRAPHIC FEATURES OF PARRY-ROMBERG SYNDROME

Parry-Romberg Sendromunun Klinik ve Radyografik Özellikleri

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ABSTRACT

Parry-Romberg syndrome or progressive hemifacial atrophy is a craniofacial disorder characterized by slow and progressive atrophy, generally unilateral, of facial tissues including muscles, bones and skin. The coup de sabre is a clear line of demarcation seen between the normal and abnormal structures. The severity of the facial deformity is dependent on the age of onset of the disease. Cosmetic management is the only available treatment and has to be delayed until facial growth is completed. The present case report deals with a 43-year-old woman with progressive hemifacial atrophy which started from the age of 10 months. Despite almost complete involvement of the right paramedian area and the early age of onset, she had neither eye changes nor any dental malformations.

Keywords: *Parry-Romberg syndrome; progressive atrophy; hemifacial; craniofacial disorder; treatment*

ÖZ

Parry-Romberg sendromu ya da progresif hemifasiyal atrofi; yüz bölgesinde kas, kemik ve cilt dokusunda genellikle tek taraflı olarak görülen, yavaş ve progresif atrofi ile karakterize bir klinik tablodur. Normal ve anormal dokular arasındaki belirgin çizgiye kılıç darbesine benzetilerek "coup de sabre" adı verilmiştir. Yüz deformitesinin ciddiyeti hastalığın ortaya çıktığı yaşa göre değişir. Bu hastalarda kozmetik tedavi başlıca seçenek olmasına karşılık yüz gelişimi tamamlanıncaya kadar ertelenmelidir. Bu olgu bildirisinde 10 aylıkken progresif hemifasiyal atrofi teşhisi koyulan 43 yaşındaki bir kadın hasta sunulmuştur. Sendromun erken yaşta başlamasına ve neredeyse yüzün sağ yarısının tamamını etkilemesine rağmen hastada gözlerle ilgili bir bulguya ya da dişlerle ilgili bir gelişim bozukluğuna rastlanmamıştır.

Anahtar kelimeler: *Parry-Romberg sendromu; progresif atrofi; hemifasiyal; kraniofasiyal bozukluk; tedavi*

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Introduction

Parry-Romberg Syndrome (PRS), also called as Progressive hemifacial atrophy, was first described by Caleb Parry in 1825 and later by Moritz Romberg in 1846 (1). Parry-Romberg Syndrome is a degenerative condition characterized by a slow progressive unilateral atrophy of facial tissues including subcutaneous fat, muscles, bone and skin (1, 2). Onset is usually in the first or second decade of life with the extent of atrophy dependent on the age of onset. PRS may often be accompanied by neurological complications like trigeminal neuralgia, migraine, seizures and changes in the eyes and hair (2). Some of the proposed etiologies include heredity, autoimmune disorders, trauma, hypo- or hyperactivity of the sympathetic nervous system, disorders of the trigeminal nerve and infectious diseases (3). PRS is usually a self-limiting condition with maximum progression between 2-5 years after onset and stabilising thereafter (2). Management includes reconstruction using fat or silicone implants (2).

Case Report

A 43-years- old female patient reported to the department of Oral Medicine and Radiology with complaint of progressive deformity of the right side of her face since 10 months of age. At the age of 10 months her mother first noticed a swelling on the right side of her face which increased for a few years and then became hard. As she grew older, she developed progressive asymmetry of the right side of the face. On extraoral examination, the right side showed gross facial asymmetry with marked hypoplasia. Incompetency of lips and obliteration of nasolabial fold were seen on the right side (Figure 1A). Vision and hearing were normal. A clear line of demarcation (coup de sabre) was seen extending from the right lower eyelid extending downwards till the lower border of mandible of the same side 3 cm away from the midline causing an obvious depression along its length. Notching of the lower border of the mandible was seen. Retrusion of mandible was also present (Figure 1B). Mild atrophy of right side of tongue was seen (Figure 1C).

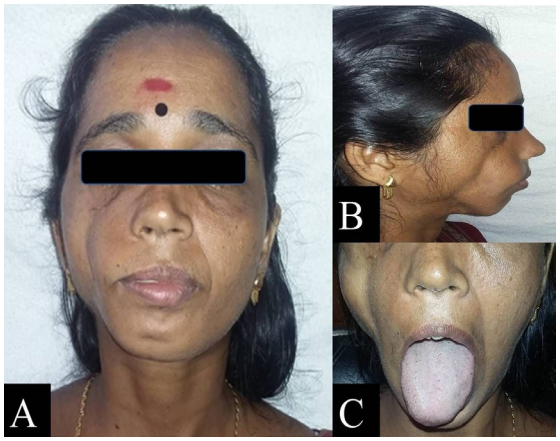

Figure 1. (A) *Extraoral photograph showing facial asymmetry with deviation of the nose to the right side* ***(B)*** *'Coup de sabre' appearance extending from the medial canthus of the eye to the lower border of the mandible. (****C****) Deviation of the tongue to the right on protrusion.*

Based on the observed clinical features, a provisional diagnosis of Parry-Romberg syndrome was given with a differential diagnosis of localized scleroderma and hemifacial microsomia. Orthopantomograph revealed prominent antegonial notch on the right side with decreased ramus and body height (Figure 2). Postero-anterior cephalometric view showed right mandibular asymmetry (Figure 3). Lateral cephalogram showed retruded mandible and decreased height of right body of mandible (Figure 4). Based on the clinical and radiographic features, a final diagnosis of Parry-Romberg syndrome or progressive hemifacial atrophy was given. Treatment plan included orthodontic alignment of teeth followed by surgical correction.

Discussion

The term 'progressive hemifacial atrophy' was coined by Eulenberg in 1871 to describe a condition characterized by unilateral atrophy of facial structures (4). It is more commonly seen in women with a ratio of 3:2 (5, 6). It commonly affects the left side of the face though the present case showed involvement of the right side.

The exact etiology of hemifacial atrophy is unknown. Due to its similarity to localised scleroderma, it has been considered as an autoimmune disorder. Cerebral disturbance of fat metabolism has also been suggested as one of the etiological factors (5). Other proposed etiologies include trauma, viral infection and sympathetic malfunctions.

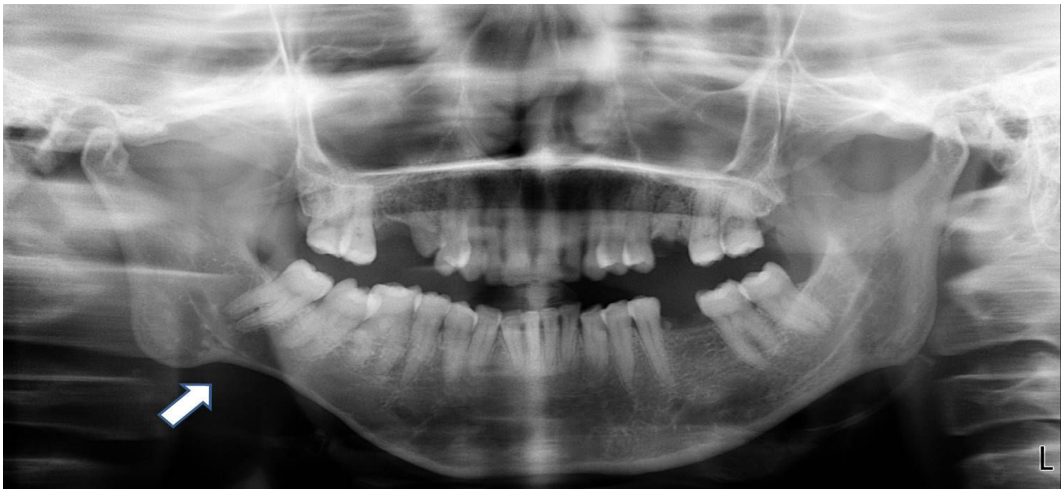

Figure 2. *Orthopantomograph showing prominent antegonial notch on the right side (white arrow) with decreased ramus and body height.*

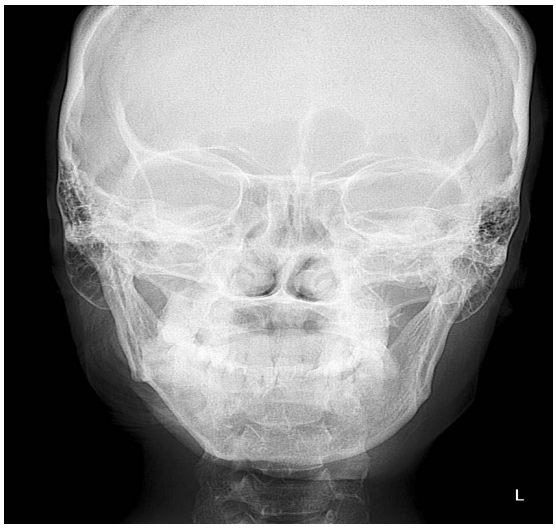

Figure 3. *Posteroanterior cephalometric view showing right mandibular asymmetry.*

Neurotrophic pathogenesis was described by Cassirer in 1912 since the process of atrophy follows the distribution of trigeminal nerve. The anatomic changes seen in Parry Romberg syndrome disturb the growth potential of hard tissue during active growth periods. Due to this, the accompanying soft tissue shrinks with loss of adipose tissue. Since facial growth is almost complete in the second decade of life, hemifacial atrophy shows fewer changes as compared to atrophy initiated in first decade (5).

There are three types of progressive hemifacial atrophy. Type I, called as morphea, is a localized area of depression of the superficial epidermal layer. It follows the distribution of the fifth cranial nerve (trigeminal nerve) with additional lymphocytic neurovasculitis. In Type II, the features of inflammation are more prominent compared to Type I.

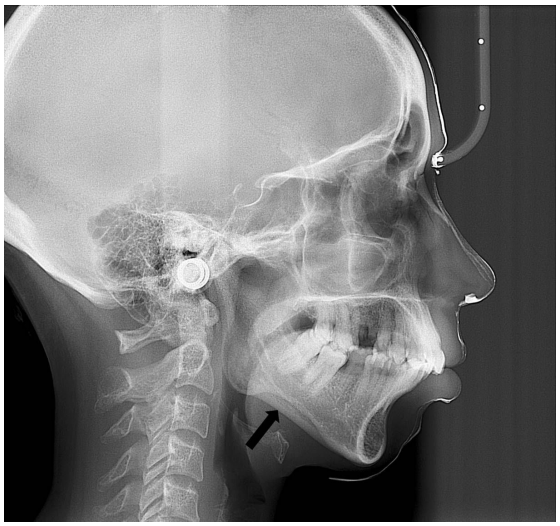

Figure 4. *Lateral cephalogram showing decreased height of right body of mandible (black arrow).*

In 1976 Rees further divided this class into two subgroups: Parry-Romberg syndrome and linear scleroderma. Type III is characterized by bilateral scleroderma where deterioration of soft and underlying skeletal tissues is present (7). Type III accounts for only 5%-10% cases of hemifacial atrophy (8).

The prime clinical feature noted is atrophy of the facial tissues, usually adipose tissue (9). Rarely, skin and other connective tissue including bone may

be affected. Severity of the atrophy can range from barely noticeable asymmetry to severe disfigurement (4). The present case showed a clear line of demarcation from the right lower eyelid extending few centimetres anterior to the angle of the mandible. Around 10-35% of cases show ocular involvement, the most encountered manifestation is enophthalmos characterized by sunken eye due to loss of adipose tissue around the orbit. No orbital changes were seen in our case. Neurological complications occur in 45% of cases, which usually manifests as severe headache, trigeminal neuralgia, facial paresthesia, and contralateral epilepsy. However these findings were not found in the present case (6). Around 50% of affected individuals show dental changes. This includes deviation of nose and mouth to the affected side. Unilateral exposure of teeth may be seen when lips are involved (6). Unilateral atrophy of tongue and deviation of dental midline has also been reported. The present case showed mild atrophy of right side of tongue. The osseous lesions are related to the age of onset and can vary. In children, when the onset is before 5 years of age, the fronto-orbito-zygomatic area is usually involved. Late onset usually causes skeletal changes in the lower third of the face. The most commonly reported bony lesions consist of maxillary and mandibular hypoplasia with deviated mid-line of the face towards the affected side. Rare reports describe spontaneous fractures of the affected mandible (4). In the present case, there was mandibular hypoplasia of the right side with prominent antegonial notch and decreased ramus and body height. Teeth on the affected side may show deficiency of root development, crowding and late eruption leading to unilateral cross bite (4). Present case showed no changes in the teeth despite early onset. Differential diagnosis includes hemifacial microsomia, Goldenhar syndrome and localised scleroderma. Localised scleroderma does not involve atrophy of underlying bone. Anti-nuclear antibody titres may be raised in scleroderma but not in PRS.

There is no cure for progressive hemifacial atrophy as it is a self-limiting condition. However, various treatments have been tried to arrest the disease. These include administration of oral steroids, D-penicillamine, methotrexate, cyclophosphamide, antimalarials, cyclosporine, and azathioprine (4). Affected patients require a multidisciplinary approach including physicians, dentists, psychologists, speech and hearing therapists (10). The treatment is usually focused on repositioning of adipose tissue that underwent atrophy due to the condition. The surgical or cosmetic treatment should be deferred until there are no further signs of atrophy therefore suggesting that the condition is stable (11).

Conclusion

Parry-Romberg syndrome is a progressive disorder involving the skin, subcutaneous fat and bone which is associated with aesthetic, functional and psychological problems. Despite early onset being associated with ocular and dental changes, the present case presented only with facial and mandibular defect. Thus PRS can present with a wide spectrum of changes ranging from mild to severe. Many factors, therefore, influence the clinical manifestations of this syndrome.

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Conflict of interest
None declared.

References

1. Ayyildiz O, Ayyildiz S, Durukan AH, Sobaci G. Progressive hemifacial atrophy with contralateral uveitis: A case report. Iran Red Crescent Med J 2015;17(9):e16168.
2. Balan P, Gogineni SB, Shetty SR, D'Souza D. Three-dimensional imaging of progressive facial hemiatrophy (parry-romberg syndrome) with unusual conjunctival findings. Imaging Sci Dent 2011;41(4):183-187.
3. Kaya M, Sel Yilmaz C, Kurtaran H, Gunduz M. Chronologic presentation of a severe case of progressive hemifacial atrophy (parry-romberg syndrome) with the loss of an eye. Case Rep Otolaryngol 2014;2014:703017.
4. Sudarshan R. Progressive hemi facial atrophy- a review. SEAJCRR 2012;1(1):16-21.
5. Deshingkar SA, Barpande SR, Bhavthankar JD, Humbe JG. Progressive hemifacial atrophy (parry-romberg syndrome). Contemp Clin Dent 2012;3(Suppl 1):S78-81.
6. Khan M, Khan M, Negi R, Gupta N. Parry romberg syndrome with localized scleroderma: A case report. J Clin Exp Dent 2014;6(3):e313-316.
7. Grippaudo C, Deli R, Grippaudo FR, Di Cuia T, Paradisi M. Management of craniofacial

development in the parry-romberg syndrome: Report of two patients. Cleft Palate Craniofac J 2004;41(1):95-104.
8. Pinheiro TP, Silva CC, Silveira CS, Botelho PC, Pinheiro M, Pinheiro Jde J. Progressive hemifacial atrophy--case report. Med Oral Patol Oral Cir Bucal 2006;11(2):E112-114.
9. Erfan M, Khan I, Ahmed AS, Hasnain S. A tale of two diseases: Parry romberg syndrome and en coup de sabre: A case report. Int J Pathol 2015;13(2):82-86.
10. Kumar AA, Kumar RA, Shantha GP, Aloogopinathan G. Progressive hemi facial atrophy - parry romberg syndrome presenting as severe facial pain in a young man: A case report. Cases J 2009;2:6776.
11. Gomez-Diez SG, Lopez LG, Escobar ML, Gutierrez LJ, Oliva NP. Progressive facial hemiatrophy with associated osseous lesions. Med Oral Patol Oral Cir Bucal 2007;12(8):E602-604.

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