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Case Report- A case of Parry Romberg Syndrome (Progressive Hemifacial Atrophy)

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Abstract
Parry Romberg Syndrome or progressive hemifacial atrophy (PHA) is an uncommon degenerative condition characterized by slowly progressive atrophy of muscles, bone and skin of the face before spontaneously entering remission. It usually involves one half of the face. Rarely both halves of the face may be involved. The various proposed causes include cerebral disturbance of fat metabolism or a trophic malformation of the cervical sympathetic nervous system. Possible factors involved in the pathogenesis include heredity, trauma, viral infections, endocrine disturbance and autoimmunity. Differential Diagnosis include Juvenile Localized Scleroderma, Rasmussen Encephalitis, Barrarquer-Simon Syndrome, Congenital Hemiatrophy, Primary Hemifacial Hypertrophy. The main aim of treatment is aesthetic restoration. There is no cure or treatment to stop the progression of the disease. We report case of Parry Romberg Syndrome who reported to the paediatric outpatient department of a peripheral hospital.

Keywords: parry romberg syndrome, progressive hemifacial atrophy, idiopathic hemifacial atrophy

1. Introduction
Parry Romberg Syndrome or progressive hemifacial atrophy (PHA) is an uncommon degenerative condition characterised by slowly progressive atrophy of muscles, bone and skin of one half of the face before spontaneously entering remission [1]. True incidence and epidemiological characteristics of PHA have been elusive due to the rarity of the disease and lack of standardised criteria for the diagnosis. It is more common in females than males. It was first described by Caleb Hillier Parry in 1825 and then in more detail by Moritz Heinrich Romberg in 1846.

2. Case report
Seven year old female child presented with facial deformity involving right half of the face, gradually progressing since two years. On examination, the patient had facial asymmetry with marked hypoplasia of the right half of the face, and deviation of the lips and nose to the right side (Fig 1 & 2). A line of demarcation (*coup de sabre*) between normal and abnormal skin was seen below the lower lip. In addition, the right ear was slightly smaller than the left ear with a prominent tragus. Intra orally there was mild hypotrophy of the right half of the tongue with atrophic tongue papillae.
3D CT Scan of Face and Brain revealed significant hypoplasia of the right mandible, including right ramus and condyloid process. The glenoid fossa of the right temporal bone was relatively hypo plastic with inadequate approximation of the temporo-mandibular joint. The posterior part of the right zygomatic arch appeared deficient. The right half of the maxilla was also hypo plastic (Fig 3 & 4).

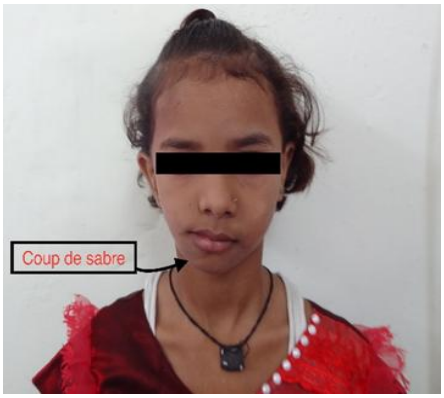

Fig 1: Coup de Sabre

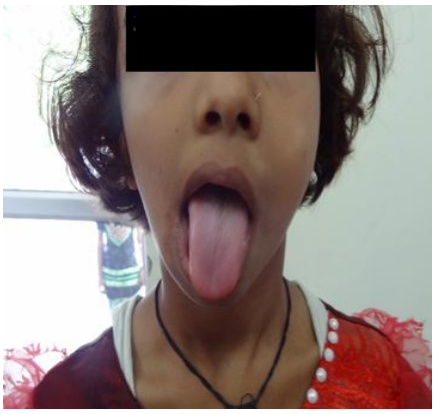

Fig 2: Facial Asymmetry

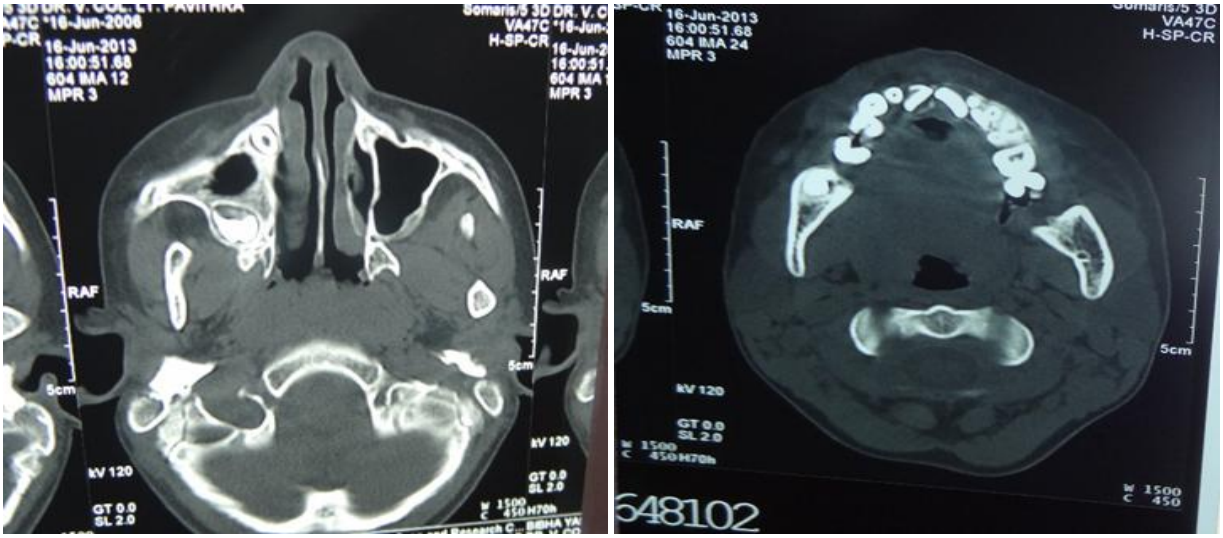

Fig 3 & 4: Hypoplasia of the right mandible, including right ramus and condyloid process. The glenoid fossa of the right temporal bone is relatively hypo plastic. The posterior part of the right zygomatic arch appears deficient.

3. Discussion

Parry-Romberg syndrome is a devastating disease resulting in severe disfigurement and potential functional impairment after years of progressing hemifacial atrophy. Eventually, the atrophy will halt as the disease enters remission; however, there can be significant tissue destruction in the interim. It usually involves one half. Rarely both halves of the face may be involved. First described by parry in 1825 and Romberg in 1846, this condition was given the nomenclature of progressive hemifacial hemiatrophy by Eulenberg in 1871 [2]. The other names given to this disorder are idiopathic hemifacial atrophy, progressive facial hemiatrophy and Romberg's syndrome. The illness appears to occur randomly. The true incidence and epidemiology of PHA is not clear owing to rarity of the disease. Prevalence is higher in females than males, with a ratio of roughly 3:2. The condition is observed on the left side of the face about as often as on the right side [3]. The various proposed causes include cerebral disturbance of fat metabolism or a trophic malformation of the cervical sympathetic nervous system. Possible factors involved in the pathogenesis include heredity, trauma, viral infections, endocrine disturbance and autoimmunity [1].

The onset of the disease is usually between 5-15 years. The progression of atrophy often lasts for 2-10 years and the disease enters a stable phase. There is progressive atrophy of unilateral facial tissues including muscles, bone and skin. Alopecia and pigmentation is often seen on the affected side. Other features include enophthalmy and deviation of the mouth and nose on the affected side. Ocular manifestations include corneal and retinal changes. Neuro-ophthalmological manifestations include involvement of optic nerve and pupillary dysfunction [4]. There may be delayed eruption of the teeth on the affected side due to deficiency in the root development. Complications that may be associated with this condition include trigeminal neuralgia, facial paresthesia, severe headache and epilepsy. Intra oral soft tissue is usually normal without any speech or deglutition problems [5]. Histology shows atrophy of the epidermis, dermis and subcutaneous tissues, along with lymphocytic and monocytic infiltration in the dermis. Degenerative changes are seen in the vascular endothelia on electron microscopy.

Differential Diagnosis include juvenile localised scleroderma (responds to treatment and ANA positive), Rasmussen encephalitis (chronic focal encephalitis), Barrarquer-Simon Syndrome (progressive cephalothoracic lipodystrophy- usually bilateral), congenital hemiatrophy and primary hemifacial hypertrophy [6]. There is no cure or treatment to stop the progression of the disease. Medical management may involve immunosuppressive drugs such as methotrexate, corticosteroids, cyclophosphamide, and azathioprine. No randomized controlled trials have yet been conducted to evaluate such treatments, so the benefits have not been clearly established. The main aim of treatment is aesthetic restoration. Reconstructive and microvascular surgery is the treatment of choice. Treatment is usually based on repositioning of the adipose tissue- for which autogenous fat grafts, cartilage grafts, silicon injections, and prosthesis, bovine collagen, and in organic implants can be used [7]. Surgical intervention is usually recommended when the disease has reached a stable phase and facial growth is complete.

4. References

1. Thiago Pastor da Silva Pinheiro, Camila Camarinha da Silva, Carolina Souza Limeira da Silveira, Patrícia Cristina Ereno Botelho, Maria das Graças Rodrigues Pinheiro, João de Jesus Viana Pinheiro. Progressive Hemifacial Atrophy- Case Report. Med Oral Patol Oral Cir Bucal, 2006; 11(2):E112-4.
2. Hiren Patel, Chintan Thakkar, Kajal Patel Parry Romberg Syndrome. A Rare entity. J Maxillofac oral Surg. 2010; 9(3):247-250.
3. Xu M, Yang L, Jin X, Xu J, Lu J, Zhang C *et al*. Female Predominance and Effect of Sex on Parry Romberg Syndrome. J Craniofac Surg. 2013; 24(4):1195-1200.
4. Bucher F, Friche J, Neugebauer A, Cursiefen C, Heindl LM. Ophthalmological manifestations in Parry Romberg Syndrome. Sur ophthalmol. 2016; 61(6):693-70. J Survophthal 2016.03.009
5. Sande A, Risbud M, Kshar A, Paranjpe AO. Progressive Hemifacial Atrophy. Dent Res J (Isfahan), 2013; 10(1):108-11.

6. Stanilav Tolkachjov N, Nirav Patel G, Megha Tollefson M. Progressive Hemifacial Atrophy- A Review. Orphanet Journal of Rare Diseases. 2015; 10:39. DOI-10.1186/s13023-015-01250-9.
7. Agostini Tommaso MD, Spinelli Giuseppe MD, Marino Gaetano MD, Perello Raffaella MD. Esthetic Restoration in Progressive Hemifacial Atrophy (Romberg Disease): Structural Fat Grafting Versus Local/Free Flaps. J Craniofac Surg. 2014; 25(3):783-7.