

Case Report

Parry Romberg Syndrome: A Rare Entity from a Primary Care Center in Bangladesh

*Podder CS¹, Chowdhury N²**Abstract**

Parry Romberg syndrome (PRS) is an acquired, rare, neurocutaneous disorder presenting most commonly as hemiatrophy of face with characteristic involvement of skin, subcutis, bone and denture with or without neurological manifestations¹. Here we present a case of 16 year old girl, who presented with characteristic insidious, progressive, selflimited hemifacial atrophy accompanied with headache and facial pain.

Key words: Parry Romberg syndrome, Hemifacial atrophy

INTRODUCTION:

Parry Romberg syndrome or, Progressive hemifacial atrophy is a rare disorder of unknown origin. First delineated by Parry in 1825 and Romberg in 1846² the term Progressive hemifacial atrophy (PHA) was Introduced by Ellen berg in 1871. The syndrome is characterized by slowly progressive hemifacial atrophy and subcutaneous structures such as fat, fascia, cartilage, bones and others of the affected side. After a progressive phase, which may continue upto 20 years, the process settles down. Usually the disfigurements are permanent Seen most commonly among females, it is also associated with ocular, neurological and oral manifestations and associated with some common autoimmune diseases. Chronic vascular disturbance, autoimmunity, prior trauma and genetic causes have been postulated for development of PHA. Diagnosis is based on history and clinical features. Histopathology and imaging aids in diagnosis. Immunosuppressants, steroids, plastic & reconstructive surgery are some of the treatment options. Here we present a case of Parry Romberg syndrome in a 16 year old girl with trigeminal neuralgia.

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CASE REPORT:

A, 16 years old girl presented to Debidwar UHC OPD with complaints of progressive atrophy of left side of the face for last 10 years. Her mother mentioned this shrinking started at the age of 6 and following recovery from a febrile illness which persisted for about 2 weeks. It was progressive and gradually increasing over the years and for last 3 years it has settled down and there is no noticeable progress. She also complained of recurrent episodic pain in the left side of the face.

She had no diminished sensations on affected side hearing or speech difficulties, no convulsions or, noticeable denture abnormalities.

On examination, there was asymmetry of left side of the face, with atrophy of cheek and mandibular region. leftside of tongue was also atrophied. There was no sensory deficit of left side, paraesthesia, auditory or speech disturbance. Other cranial nerves & motor system examination also revealed no abnormality.

Table 1: Patient's baseline reports

Investigations	Results
Hb%	11.9 gm/dl
WBC count	7,800/cumm
Neutrophil	67%
Lymphocyte	28%
Monocyte	3%
Eosinophil	2%
Platelet count	1,75,000/cumm
TSH	2.25 mIU/L
ANA	Negative



Fig 1: Patients photograph showing left sided hemifacial atrophy

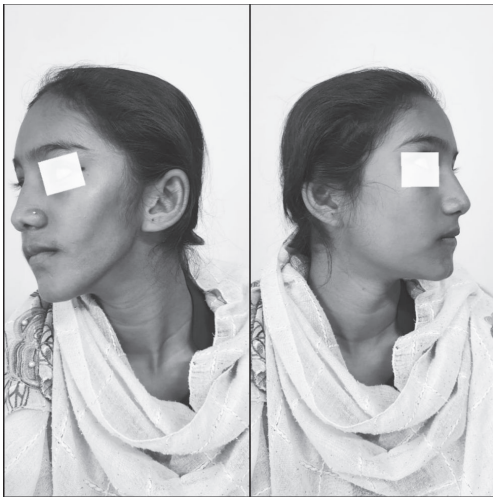


Fig 2: Showing marked facial Asymmetry



Fig 3: Showing atrophy of left half of tongue

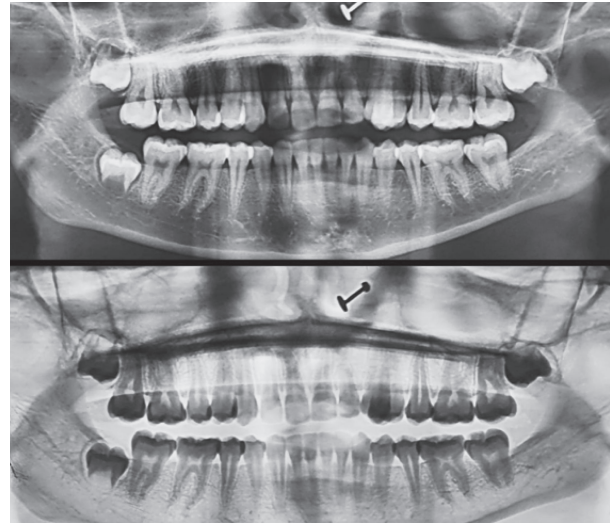


Fig 4: The orthopantomography shows decreased vertical height of ramus along with loss of gonial angle prominence on the affected side. There is also Class 3 malocclusion in right side, left side class 1



Fig 5: X ray Skull showing no obvious bony deformity

DISCUSSION:

Parry Romberg syndrome is a rare disorder of unknown aetiology and pathogenesis. It has been propositioned to be related to prior trauma, genetics, infection, vascular

malformation, cervical sympathetic overactivity and autoimmunity.^{3,4,7,8,9}

Parry Romberg syndrome has a female predilection, presenting more commonly in 1st and 2nd decades and average age of onset is 13.7 year.^{3,4} Interestingly left side of the face most common involved.⁵ After initial presentation, the course is insidious, slowly progressive and is self-limiting.¹³ Characterized by hemifacial atrophy, absence of underlying skin induration, atrophy of subcutaneous tissue, fat, skin, muscle, bone (100%), dental abnormalities (50%), migraine/facial pain (45%), hemimasticatory spasm (35%), tongue atrophy (25%), vitiligo (20%), hemiatrophy of ipsilateral/contralateral arm/trunk/leg (20%)⁶. Trigeminal neuralgia, facial paraesthesia, severe headache and epilepsy are most common neurological manifestations. Our patient presented at the age of 6, was progressive and later on halted on its own, had left hemifacial atrophy and subcutis, but no obvious involvement of bone. The mentioned OPG findings are correlatable with PRS. Notably, she was complaining of episodic sharp lancinating pain, which was very much typical of trigeminal neuralgia. She had no history of epilepsy. she had a preceding history of fever but, the it's aetiology was unknown. She had no history of trauma to affected side or positive family history for any autoimmune diseases. Closest differential of it is linear scleroderma, "en coup de sabre" (ECDS). But, distinctive clinical features and histopathological findings help to differentiate between these two entities., due to financial constraints and lack of resource, histopathology had not been done. However, 28-42% of patients have been reported to have concurrence of these two diseases^{8,14}

Overall prognosis of PRS is unpredictable. Usually there is only cosmetic effect rather than any life threatening disability. Treatment for PRS is very challenging. Methotrexate is the standard therapy for active disease. Methotrexate is often combined with oral prednisolone in tapering dosage. For prolonged remission a 12-24 month course of methotrexate is recommended^{9,10,11}. Other immunosuppressive agents have variable response. Surgery has been recommended when the disease process halts. It needs a multidisciplinary approach and autologous fat grafting, injectable fillers, lipoinjection, dermal fat grafts, adipofascial flap, bone grafts are some of the treatment options for aesthetic augmentation^{11,12}. As, our patient when presented to us, had no evidence of active disease, no treatment was offered. However, she was counselled about available possible treatment options, but she refused. She

had been counselled about the disease course, prognosis and possible complications and had been asked for periodic followup.

AUTHORS CONTRIBUTION:

CSP was involved in diagnosing the case and management of the patient. NC was involved in literature review and manuscript writing.

Conflict of interest: Nothing to declare

Consent: Informed written consent was taken from the patient for publication of this case report with accompanying images.

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