Parry–Romberg Syndrome: A Rare Entity

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Abstract Parry–Romberg syndrome or progressive hemifacial atrophy is very rare, uncommon, degenerative, poorly understood condition characterized by a slow and progressive atrophy affecting one side of the face. The incidence and the causes of this alteration are unknown. Possible factors that are involved in the pathogenesis include disturbance of fat metabolism, trauma, viral infections, heredity, endocrine disturbances and auto-immunity. The most common complications are: trigeminal neuritis, facial paresthesia, severe headache and epilepsy. Characteristically, the atrophy progresses slowly for several years and become stable after certain time period. After stabilization of the disease multi specialty approach including physician, orthodontic treatment and reconstructive surgery with autogenous fat graft can be performed to correct the deformity. The objective of this article is to accomplish a literature review concerning general characteristics, etiology, physiopathology, differential diagnosis and treatment of progressive hemifacial atrophy.

Keywords Progressive hemifacial atrophy · Parry–Romberg syndrome · Romberg’s Sign · En coup de sabre · Localized linear scleroderma

Introduction

Parry–Romberg syndrome was first reported by Caleb Parry in 1815 and then described by Moritz Romberg in 1846. Parry–Romberg syndrome is a condition in which there is slow and progressive shrinkage of the tissues and sometimes bones of one or occasionally both sides of the face. It is generally unilateral atrophy of facial tissues including muscles, bones and skin [9, 11]. More than an aesthetic trouble, this illness bring several functional and psychological problems, like ocular problem, migraine, epilepsy. The incidence and cause of this alteration is unknown. A cerebral disturbance on fat metabolism has been proposed as a primary cause [4], trauma, viral infections, endocrine disturbances, auto-immunity and heredity are believed to be associated to the pathogenesis of the disease [13, 18, 22].

Frequently, the onset of this syndrome occurs along first and second decades of life. The atrophy progress slowly during years and then it stabilizes at its own [13, 15, 20]. Patients who manifest atrophy in early ages, have bigger repercussions and it is directly related to growing age of patient [16].

This syndrome seems to have higher incidence in women [4, 9, 17]. The extension of the atrophy is frequently limited to one side of the face, and the ipsilateral involvement of body is rare. 5–10% of cases were described as being bilateral [14].

The most important features of this pathology are the enophthalmos, the deviation of mouth and nose towards the affected side, and unilateral exposition of teeth (when lips are involved).

Parry–Romberg Syndrome is an auto-limitable condition with no cure. Affected patients should have multidisciplinary attendance of plastic surgeon, physicians, dental

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surgeon, phonoaudiologists and psychologists to give a better aesthetic to patient. Nowadays, cosmetic surgeries with autogenous fat graft, injections of silicone or bovine collagen, and inorganic implants are some alternatives to correct deformities [3, 18]. Besides esthetic improvement, symptomatic treatment for neurological disorders is indicated.

Aetiology

There have been a number of theories on the etiology of Parry–Romberg Syndrome include trigeminal neuritis, a chronic autoimmune neurovasculitis, a chronic infestation with a neurotropic virus (e.g. Herpes) and an increased sympathetic nerve activity triggering facial atrophy [19]. A similar condition in rats has been initiated by performing a cervical sympathectomy during the new born.

For *Myelopathy*—Multiple sclerosis, Vitamin B₁₂ and Vitamin E deficiency, structural spinal cord disease, infectious myelopathy.

For *Peripheral neuropathy*—Vitamin B₁₂ deficiency, Inherited demyelinating neuropathies, cerebellar dysfunction, vestibular dysfunction (peripheral), drug intoxication, alcohol, cisplatin, pyridoxine (Vitamin B₁₂) overdose, anticonvulsant toxicity (especially phenytoin).

Signs and Symptoms (Clinical Features)

In hemi facial atrophy unilateral sunken cheek, eye and tongue is seen. Migraine or migraine like headaches, trigeminal neuralgia, seizures—starting in the opposite side of the body, alopecia, blanched hair, atrophy of fat and subcutaneous tissue on the affected side are common. Degenerative brain lesions, intracranial calcifications, Sensory impairment, excessive sweating, tear duct dysfunction may be seen [7, 9, 16, 21].

Diagnosis

Romberg's Sign

The Romberg test is named after the nineteenth century German Ear Specialist, Moritz Heinrich Romberg (1795–1873). It is a neurological test to detect poor balance, examine proprioception also to some extent, cerebellar function. The test is performed by having the patient with feet together and eyes closed, which eliminates the visual cues that help to maintain posture. Patients with positive Romberg's sign shows diminished proprioception begin to fall or move their feet to maintain balance. Patients with vestibular dysfunction may also exhibit

positive Romberg's sign. Romberg's sign shows cerebellar lesion and its not seen in a normal individual.

Romberg's sign, when positive perform other neurologic screening tests like proprioception, knee bend and to walk straight line placing heel to toe, evaluate his gait, patient's awareness of body part position while his eyes are closed, direct the movement, repeat the movement several times, gradually increasing the speed, vibratory sense nystagmus was not observed.

Investigation

Diagnosis can be made clinically if the patient is having facial asymmetry (physical sign). MRI and CT scan should be done if the patient is having neurological symptoms [1, 6]. Patient having epilepsy should be investigated with lumbar puncture and auto antibodies. Thermographic response shows that the affected part is warmer than the unaffected part [2, 10, 12].

Complications

Fits, poliosis, facial, abnormality, enophthalmos, headache, alopecia and hyperpigmentation.

Differential Diagnosis

Linear scleroderma “en coup de sabre” (or localized linear scleroderma) is a well recognized entity by dermatologists and rheumatologists. Clinically, linear scleroderma may present in childhood and it involves intense loss of subcutaneous fat with ensuing thinning and pigmentation of the skin. It is commonly seen in the paramedian forehead region. In “en coup de sabre” atrophy of underlying muscle or bone is not seen. There is prolonged nerve conduction in areas affected by scleroderma which do not exist in Romberg's. Anti-nuclear anti-body titres are often raised with active linear scleroderma, but rarely so with Romberg's disease.

Discussion

Progressive hemifacial atrophy is disease of unknown etiology and rare pathology. It not only involves soft tissue but sometime involve bone causing severe esthetic discrepancy. There is wasting of muscle and subcutaneous tissue. Ocular involvement is usual so enophthalmos occur due to fat loss around orbit. The eye works normally. Ear can be smaller due to atrophy. There may be localized area of alopecia. There is neurological complication including trigeminal neuralgia, epilepsy, facial paraesthesia, headache etc. [7, 9, 16]. Facial and dental midline deviation along with mouth

Fig. 1 Patient with hamifacial atrophy put in discussion at in our case (shown in photograph), atrophied right side of lip and right tongue is seen along with facial atrophy

and nose on the affected side is seen. Unilateral atrophy of muscle of the tongue and upper lip (orbicularis oris) is seen.

In our case (shown in photograph), atrophied right side of lip and right tongue is seen along with facial atrophy.

No radiological deficiency seen although in some cases when there is involvement of bone in dentition deficiency of root development and delayed eruption causing dental crowding, unilateral posterior crossbite [4, 5, 15]. Intraoral soft tissue and muscles of mastication is also involved but the normal function like speech, deglutition are not disturbed [13, 17] (Fig. 1).

Histologically dermis and epidermis of skin and subcutaneous tissue are atrophied [17]. There is atrophy of subcutaneous fat and degenerative changes are seen in vascular endothelium in electron microscope [17]. Treatment usually based upon, reposition of adipose tissue, autograph, silicon injection, prosthesis [3, 22]. These treatment modalities momentary resolved facial esthetic problems. Cosmetic treatment recommended when progress of the disease stops. This is the reason why our patient as not undergone any surgical treatment.

Treatment

The list of treatment mentioned in various sources for Parry–Romberg Syndrome includes the following list.

Always seek professional medical advice about any treatment or change in treatment plans. Reconstructive or microvascular surgery, fat graft and dermis graft, fat cell injection (lipoinjection or lipofilling), silicone implantation, muscle graft, bone graft, orthognathic surgery, allografts, along with symptomatic and supportive therapy like assistive devices (e.g. cane, walker), surgical therapy for compressive myelopathy, spondylitic myelopathy [3, 8, 20, 22]. Supplementation of Vitamin B₁₂, eliminate exposure to offending toxic substances, multiple sclerosis is treated with steroids, interferons, glatiramer acetate, mitoxantrone.

Prognosis

In some cases, the atrophy stops before the entire face is affected. In mild cases, the disorder usually causes no disability other than the cosmetic effects.

Recovery period for overall prognosis of Parry–Romberg syndrome is unpredictable.

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