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Parry Romberg Syndrome-A Review of Treatment Options

BANU SARGUNAR[1], VIJAY EBENEZER[2], R. BALAKRISHNAN[3] and SWARNA PRIYA[4]

Department of Oral and Maxillofacial Surgery, Sree Balaji Dental College and Hospital,
Bharath University, Chennai, 600100, India.
*Corresponding author E-mail: drbanuabe@gmail.com

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ABSTRACT

Parry-Romberg Syndrome, also known as Progressive Hemifacial Atrophy, is an uncommon degenerative condition, characterized by a slow and progressive atrophy affecting one side of the face. A cerebral disturbance of fat metabolism, atrophic malformation of Cervical Sympathetic Nervous System has been proposed as the primary cause. Other possible factors that are involved in the pathogenesis include trauma,viral infections, heredity, endocrine disturbances and auto-immunity. The objective of this work is, to accomplish a literature review concerning general characteristics, etiology, physiopathology, differential diagnosis and treatment of progressive hemifacial atrophy. A review of literature with variety of treatment options have been discussed in an attempt to treat the atrophic deformties from mild to severe cases. These have included free fat grafts, dermis fat grafts, fascia,muscle grafts, cartilage and bone augmentation ,Orthognathic surgery,microvascular reconstruction and stem cell regeneration of bone and soft tissues.

Key words: Hemifacial atrophy, Parry-Romberg syndrome, physiopathology, treatment options.

INTRODUCTION

Hemifacial atropy is a uncommon degenerative slowly progressing unilateral atrophy of the facial tissues including the skin, subcutaneous fat, muscle, cartilage and bone. This was first described by Parry and Romberg in the year 1846. Onset of Disease occurs at first decade of life and progresses slowly over years frequenly for 2 to 10 years and reaches a stable burn out phase. [10] It is uncommon and unilateral with a higher incidence rate in females[18]. The extension of the atrophy is frequently limited to one side of the face[16], and cranium, it may occasionally spread to the neck and one side of the body and it is accompanied usually by ocular involvement, the most frequent manifestation is enophthalmy, deviation of mouth and nose to the affected side, and unilateral exposure of teeth (when the lips are involved). Occasionally, there may be some neurological complications, such as trigeminal neuralgia, facial paresthesia, severe headache and contra lateral epilepsy. The presence of antinuclear antibodies in the serum suggested that the Parry-Romberg syndrome may be a form of localized scleroderma.

Clinical features

The shape and symmetry of head is abnormal with a cleft on the midline. The head is flat in the anterior region along the midline. Flattening is seen in the cranial vault at midline extending till hair line. Depression or furrow is present at anterior fontanelle region and at the mid line of fore head close to hair line. Shape of the forehead is asymmetrical with a depression on midline of forehead close to glabella. Enopthalmus of the right eye due to atrophy of orbital fat occurs. Supra orbital margin appears normal with a prominent infraorbital margin. Nasal bones appear normal with depression seen on the affected side of nose close to the tip.

The nose is deviated to the right side. Ears appear normal. Zygoma, maxilla and mandible appear asymmetrical on the affected side. Atrophy of soft tissue is seen in the infraorbital, zygomatic and mandibular region. On palpation, the supraorbital, infraorbital, zygoma, maxilla and mandible appear almost symmetrical on both the sides. There is loss of soft tissue bulk on upper and lower eye lid. The eye looks sunken on the affected side due to loss of orbital fat[14]. There is also loss of hair in the lower eye lid. Due to complete loss of soft tissue bulk in the cheek and the mandibular region on the affected side, there is stretching of skin on the entire aspect of the right side, causing depression of ala of nose, retraction of upper and lower lip, prominent exposure of infraorbital rim and zygoma, angle of mandible. Skin is pigmented more on the right zygomatic region, ramus region and the corner of the mouth. There is scarring in the infraorbital region extending till hair line in front of the ear. On clenching, the masseter and temporalis muscles appear prominent. Muscle bulk is comparatively less on the affected side. There is excessive exposure of teeth on the affected side during smiling. Upper and lower lip is thin. There is a deep cleft seen on the affected side of the chin. Maxilla and mandible appears to be prognathic. The most important features of this pathology are enophthalmy, the deviation of mouth and nose to the affected side, and unilateral exposition of teeth when lips are affected[13,16].

The condyle is slightly larger on the affected side compared to the other. The roots of the posterior teeth on the affected side appear slightly shorter.

There is mild deviation of the nasal septum to the affected side. The mandible and condyle appears larger on the affected side. There is pain and tenderness over messetric region on wide opening.

128 slice CT of facial bones shows reduction in the size of right side of skull bones and thickness of soft tissues. There is thinning of pericranial and facial soft tissues. The orbit and the maxillary sinus on the affected side appear smaller in size. Brain shows no focal lesion. CT and MR findings included unilateral focal infarctions in the corpus callosum, diffuse deep and subcortical white matter signal changes, mild cortical thickening.[20]

Transaxial FLAIR MR Images demonstrate atrophy of skin and subcutaneous tissues overlying the left frontal calvarium as well as ipsilateral cerebral atrophy and diffuse white matter hyperintensities involving the left frontal, parietal, and occipital lobes, external capsule, and corpus callosum splenium. Axial T2*-weighted, gradient-echo MR images demonstrate ipsilateral microhemorrhages involving the left isthmus of the cingulate gyrus, parietal and occipital white matter, thalamus, and corpus callosum splenium. Additionally, a cystic lesion lined by hemosiderin is demonstrated in the left superior frontal lobe, consistent with old, encapsulated hematoma[21].The finding of unilateral cerebral microhemorrhages ipsilateral to facial hemiatrophy suggests that some cases of Parry-Romberg syndrome may be secondary to a small-vessel neurovasculopathy.

According to Wells and luce classification of defect, Parry Romberg syndrome is considered as type 2 defect

Type 1	Cutaneous defect, subcutaneous and underlying bony frame work intact eg. nevi, scar
Type 2	Deep soft tissue defect, involving muscles, require greater bulk to restore facial contour Eg. Romberg syndrome, lipodystrophy, hemifacial microsomia
Type 3	Full thickness defect of cheek due to resection of malignant neoplasm
Type 4	Deformities of bony structures or frame work of maxilla and zygoma leading to esthetic and functional morbidity of eye and dentition
Type 4a	Partial loss of maxilla with loss of palate and alveolar ridge
Type 4 b	Extensive loss of maxillary bone including nasomaxillary, zygomaticomaxillary region and floor of orbit

Aetiology

Theories proposed include neurogenic, vascular, exogenous insult and autoimmune mediated processes[19]. A popular hypothesis is the vasomotor trophoneuritis theory, involving the sympathetic nervous system which results in the atrophy of facial tissues.[2] Immunological evidence shows involvement of noradrenergic system[15] in the brain stem causing hyperactivity of the brain

stem sympathetic centers, possibly caused by an autoimmune process [16,17]may be the primary cause for cutaneous and subcutaneous atrophy in Parry-Romberg Syndrome .Wartenburg considered the primary factor to be a cerebral disturbance leading to increased and unregulated activity of sympathetic nervous system, which in turn produced the localized atrophy through its trophic functions conducted by way of sensory trunks of trigemminal nerve. Other workers suggested extraction of teeth, local trauma, infection and genetic factors could also be a cause. In a paper published in 1973, Poswillo attributed the development of facial deformities to the disruption of stapedial artery. The stapedial artery functions as a stopgap vascular channel during days 33-45 of embryologic development (ref)

Differential diagnosis

Localized Scleroderma, 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[16, 21]. Anti-nuclear anti-body titres are often raised with active linear scleroderma, but rarely so with Romberg's disease. The presence of antinuclear antibodies in the serum suggestive of Parry-Romberg syndrome which may be a form of localized scleroderma. The other differential diagnosis include, lupus erythematosis, trigeminal neuritis and chronic neurovasculitis.

Treatment options

Medical management

- Corticosteroids (topical and intralesional)
- Retinoids
- Antioxidants
- Immunosuppressants (methotrexate).

DISCUSSION

Surgical treatment options

- Fascia grafts
- Muscle grafts
- Pedicle flaps
- Microvascular free flaps allow for the permanent correction of large deformities
- Free fat grafts-Injection of aspirated fat
- Alloplastic graft materials

Hemifacial atrophy treated with autologous fat transplantation: [6,7]**.** Extraction of adipose tissue through a cannula or a needle and the fat was harvested from the buttocks using a 10cc syringe to which a 16 G lumbar puncture needle was attached. To and fro' movement of the LP needle in the subcutaneus plane (tunneling approach), creates a culture of adipocytes which is believed to lead to more rapid revascularization and less reabsorption. The prepared fat was injected into the target area in the subcutaneous plane with a 16 G needle in a retrograde manner depositing ribbons of fat in the desired area. Over correction was made to compensate for subsequent resorption. The injected fat, which was pliable, was sculpted into the designed contour by manual pressure.

In spite of the satisfactory results achieved with lipofilling it may be considered an interesting solution for soft tissue augmentation of the face especially for moderate adipose defects, due to its repeatability, no donor site morbidity, no complications at the recipient site such as lesions resulting from dissection, bleeding, necrosis, etc. This technique can be performed in a day-hospital with short surgery time, at low cost and without a highly skilled team. For severe grades of adipose atrophy, because of the low blood supply to these tissues which interferes with take of any type of autograft, we think that free flaps actually represent one of the best solutions for soft tissue augmentation[11].

Advantages

- Cost effective
- Simple technique
- No rejection and allergy
- Good results at three years appear encouraging

Disadvantage

Over-correction of the region with gradual atrophy may require further treatment[8].

Free vascularized tissue transfer should be considered for young patients, because the free flap is the best among the procedures for Romberg's disease for maintaining volume.

Reconstruction of hemifacial atrophy with a free flap of omentum:[9]

- Provide adequate tissue in three dimensional plane for reconstruction,
- Adequate blood supply,
- Lack of sufficient atrophy of the soft tissue mass after several years follow-up.

Disadv

Intraabdominal harvest,Difficulty in fixation, and excessive bulk

Free Jejunal flaps

Segment of jejunum can be removed without impairing absorption and digestion

Advantages

Pliable and elastic allows the surgeon to mold and adapt to reconstruct the defect

Galea temporoparietal fascia flap,temporalis muscle flap[13]

Reconstruction of orbital ,forehead and cheek region

Aesthetic treatment of progressive Hemi facial atrophy

use of a pedicled platysma muscle flap.

The platysma muscle flap was used for reconstruction in four cases of severe to moderate disease.[7]The platysma flap was transected at the clavicular level, turned at the mandibular margin, and spread subcutaneously on the affected side from the nasolabial fold up to the orbital margin and laterally to the anterior part of the ear.The flap masked the atrophy relatively well, and no complications were seen during or after the procedure.

Candidate for free flap:

- Patients with severe abnormalities
- Young patients
- Timing for reconstruction: Wait for 18-24 months after atrophy stops

Allografts

Implantation of autogenous tissue for major contouring is associated with morbidity from the donor site, Prolongation of the surgery, Possibility of absorption if the tissue is not vascularised,Insufficient amounts of autogenous material, Difficulties in contouring of grafts, necessitates the use of alloplastic material for facial augmentation.

The ideal host bed for allografts is one that is covered by well-vascularized thick skin and subcutaneous tissue that is not subjected to the stresses of trauma or motion.

- High density Porous polymer (Medpor)
- Porous polymers
- Polytetrafluroethylene-proplast
- Expanded fibrillated PTFE-goretex
- Solid polymers
- Silicone,
- Polymethylmethacrylate
- Liquid silicone
- Meshed polymers –mersilene ,Marlex
- Poly –L-lactic acid:
- Poly-L-lactic acid (PLLA) which is biocompatible and biodegradable
- *Sculptra* is used primarily to augment the soft tissues of the face, replacing fullness in areas of fat loss (lipoatrophy)
- . Injections of a hyaluronic acid filler[24] into the right side of the upper lip improved the cosmetic appearance and further treatment with autologous fat transfer is planned.
- *Stem cells* The controlled study, conducted by Dr Kyeung-Suk Ko and Dr Jong-Woo Choi and led by Dr Jeong-chan Ra of RNL Stem Cell Technology Institute, painlessly removed a few ounces of fat from one group Parry-Romberg Syndrome patients, harvesting stem cells from these patients' fat.s Adding stem cells to standard-of-care therapies, described a revolutionary finding, that "adult" mesenchymal stem cells saw unprecedented improvement in the effectiveness of therapies. stem cell transplantation is considered as standard of care of treatment[23].

Prognosis[16]

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. It is an auto-limitable condition and there is no cure. The treatment is usually based on reposition of adipose tissue[22]

REFERENCES

1. Plastic and Reconstructive Surgery, 113(2): (2004).
2. Clin Exp Ophthalmol **31**: 363-5 (2003).
3. Neurology india, **47**(4): 342-3 (1999).
4. Journal of dental science,Dept of restorative dentistry awolowo university
5. Indian journal of dermatology **50**(3): 150-151 (2005).
6. Howard a. Tobin,toronto, canada Internet journal of dental science
7. Plastic and Reconstructive surgery **96**(1): (1995)
8. Journal of Reconstructive Microsurgery, **21**(1): 15-19 (2005).
9. James wallace International journal of surgical reconstruction, **32**(1): 15-18 (1979).
10. Mazzeo N, Fisher JG, Mayer MH, Mathieu GP, Mcade FGG: Progressive hemifacial atrophy (Parry Romberg Syndrome).*Oral Surg Oral Med Oral Pathol Oral Radiol Endod,* **79**: 30-35 (1995).
11. Roddi .R. Riggio, Clinical evaluation of techniques used in the surgical treatment of progressive hemifacial atrophy. *J Craniomaxillofac Surg.*, **22**(1): 23-32 (1994).
12. Progressive hemifacial atrophy – Case report Thiago Pastor da Silva Pinheiro **1**, Camila Camarinha da Silva **2**,Med Oral Patol Oral Cir Bucal **11**: E112-4 (2006). Parry-Romberg Syndrome
13. Cheung LK. The vascular anatomy of the human temporalis muscle: implications for surgical splitting techniques. *Int J Oral Maxillofac Surg.*, **25**: 414-421 (1996).
14. Ong K, Billson FA, Pathirana DS, Clifton-Bligh P. A case of progressive hemifacial atrophy with uveitis and retinal vasculitis. *Aust N Z J Ophthalmol* **19**: 295-8 (1991).
15. Neurology India Parry Romberg syndrome : newer concepts in pathophysiology. Aleem MA, Meikandan D, Year : **47**(4): 342-3 (1999). Parry–Romberg Syndrome: A Rare Entity
16. Hiren Patel, Chintan Thakkar, and Kajal Patel,Maxillofac *Oral Surg.* **9**(3): 2 (2010).
17. Finesilver B, Rosow HN. Total hemiatrophy. *J Am Med Assoc.* **110**(5): 366-368, 47–250 (1938).
18. Hiren patel,Journal of oral and maxillofacial surgery, **9**(3): 247-250 (2010).
19. El-Kehdy J, Abbas O, Rubeiz N. A review of Parry-Romberg syndrome.J Am Acad Dermatol. **67**(4): 769-84 (2012).
20. R C Cory, D A Clayman, American Journal of Neuroradiology , Advanced Search Bottom of Form Clinical and radiologic findings in progressive facial hemiatrophy **262**: 721-725 (2012).
21. Marisa K. Blitstein, BS, Michael J. Vecchione, DO,Journal of radiology, **40**(1): (2011).
21. Beata Bergler-Czop*, Anna Lis-Zwi'ty and Ligia BrzeziDsk,"Scleroderma linearis: hemiatrophia faciei progressiva (Parry-Romberg syndrom) without any changes in CNS and linear scleroderma "en coup de sabre" with CNS tumor, *BMC . Neurology* **9**: 39 (2009).
22. Vykuntaraju ,*Indian Pediatr* **50**: 255 (2013).
23. Dr Kyeung-Suk Ko and Dr Jong-Woo Choi , Dr Jeong-chan Ra International plastic surgery journal Annals of Plastic Surgery
24. Tameka K Lane MD, Jessie Cheung MD , Parry-Romberg syndrome with coexistent morphea,*Dermatology online journal* **14**(1): (2008).
25. Jon Stone,Neurological Rarity-Parry-Romberg syndrome, *Pract Neurol* **6**: 185-188 (2006).