

FREE DIFFERENTIAL THICKNESS DE-EPITHELIALISED ANTEROLATERAL THIGH FLAP FOR SEVERE HEMIFACIAL ATROPHY IN A CHILD – A CASE REPORT

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ABSTRACT

Parry Romberg syndrome (PRS), also known as progressive hemifacial atrophy is a rare self-limiting degenerative neurocutaneous and craniofacial disorder, typically affecting one half of the face occurring in children and young adults which causes functional, aesthetic and psychological disabilities with social implications in the affected individual. It is characterized by atrophy of the skin, fat, muscles and underlying bone and cartilage affecting the face and neck unilaterally. It is associated with neurological symptoms like epilepsy and involvement of other organs like the eye. It has a slow and progressive course and its cause remains largely unknown. It was been noted that this condition is more common in females. Neuroimaging with computed tomography (CT) scan aids in demonstrating radiological features, assessing disease severity, and detecting neurological and ocular complications. The diagnosis was based on clinical features. A multidisciplinary team approach involving rheumatologists, dermatologists, plastic & maxillofacial surgeons, ophthalmologist, paediatrician, neurologist and psychologists are required for the management of this problem. We present a case of PRS in a 9-year-old female child with gross facial asymmetry, with marked wasting and atrophy involving the entire right side of the face for which a differential thickness de-epithelialised free anterolateral thigh flap was done to correct the deformity.

KEYWORDS: Parry-Romberg syndrome, Progressive facial hemiatrophy, Facial asymmetry, De-epithelialised Anterolateral Thigh Flap

INTRODUCTION

Parry-Romberg syndrome (PRS), also known as progressive hemifacial atrophy, is a developmental disorder characterized by unilateral progressive atrophy of the skin and its underlying tissues of the face and neck. It is rare with a prevalence of at least 1 in 700,000 births. (1) It was initially described by Dr. Caleb Hillier Parry in 1825 and Dr. Moritz Heinrich Romberg in 1846, but the term "progressive hemifacial atrophy," was however coined in 1871 by Eulenberg. (2-4) PRS primarily affects the subcutaneous fat but also has a variable involvement of skin, muscle, cartilage, and bone. This condition usually manifests in childhood, usually within the initial 2 decades of life, with a higher prevalence in females. (1,5,6) After disease onset, the condition follows a gradual progression that eventually reaches a stable or "burnt out" phase. (7,8) Disturbance in fat metabolism, viral infection, trauma, heredity, endocrinal or vascular disturbances, and autoimmunity are few possible factors in its pathogenesis. (5,9,10) The resultant craniofacial asymmetry causes cosmetic problems, in addition to neurological and ophthalmological complications, including seizures, trigeminal neuralgia, migraines and enophthalmos. (11,12) PRS is a clinical diagnosis, but radiological investigations are required to assess the extent of soft tissue or bony atrophy, identify intracranial involvement and monitor disease progression. (13) The treatment for the cosmetic deformity is done after the syndrome is stable and includes procedures like silicone implants, fat, skin and bone/cartilage grafting.

CASE REPORT

A 9-year-old girl presented with deformity of the right side of face for the past 6 years. She was apparently normal till 3 years of age when her mother noticed a blackish discoloration on her right cheek, which gradually progressed over time. At 6 years of age, the deformity had involved the entire right side of her face and reached

a plateau, where further progression was static for the past 3 years (burnt-out phase). There was no history of seizures in the past. She was discriminated by peers with refusal to go to school and social stigma among the neighbourhood. On examination, there was gross asymmetry of the face, with marked wasting and atrophy involving the entire right side of the face. **(Fig. 1)** There was skin hyperpigmentation, skeletal growth restriction, enophthalmos, dental crowding, occlusal cant and ear deformity on the affected side. There was a 'En Coup de Sabre' deformity of the forehead. The facial nerve and facial muscle function was normal. A clinical diagnosis of Progressive Hemifacial Atrophy or Parry-Romberg Syndrome was made. Computed Tomography (CT) of the face was done with 3D reconstruction. It showed facial asymmetry with underdeveloped right hemifacial bony and soft tissue structures. **(Fig. 2)** There was hypoplasia of the right zygoma, zygomatic arch, maxilla, medial and lateral pterygoid plates and the right hemi-mandible. The right globe is displaced posteriorly with atrophy of the intraconal fat atrophy, causing enophthalmos. The right hemi-tongue exhibited mild atrophic changes, with severe hypoplasia of the right parotid and submandibular salivary glands. The right external auditory canal anterior bony wall was also hypoplastic. There was atrophy of the right temporalis, medial and lateral pterygoid, masseter, risorius, buccinator, zygomaticus major and minor, levator labii superioris, levator anguli oris and orbicularis oris muscles. There was volume loss of the right facial subcutaneous fat and skin thinning, especially of the right frontotemporal scalp. Magnetic resonance imaging (MRI) of the face was also done which showed ipsilateral right hemi-facial atrophy of the skin, subcutaneous tissues, cartilaginous, and bony structures causing facial asymmetry. The brain parenchyma appeared normal with no signs of altered signal intensity and no evidence of brain atrophy. The complex nature of the syndrome, affecting both the skeletal and soft tissue structures, required a multidisciplinary approach involving plastic surgeons, paediatricians, paediatric intensivists, anaesthetists, and other technical staff to provide a comprehensive management. We divided the face into multiple subunits for a subunit-based planning. **(Fig. 3)** We planned for a differential thickness de-epithelialised free anterolateral thigh flap with maximum bulk in malar and cheek area and lesser bulk at the temporal, infraorbital region, upper and lower lip regions with a monitoring skin paddle and fascia for lining if needed. Fat grafting was planned to fill the deformity of the en Coup de Sabre and restore volume. **(Fig. 4)** The anterolateral thigh flap was harvested as a single perforator pedicle. The skin was de-epithelialised and the flap was thinned differentially according to the plan. **(Fig. 5)** The intra-oral approach was used with a horse-shoe shaped incision and subperiosteal dissection with pockets developed for the soft portions of the flap. **(Fig. 6)** The recipient artery was the facial artery with internal jugular veins as recipient veins for anastomosis. Post-procedure, the patient was shifted to the paediatric intensive care unit and placed under observation. The post operative period was uneventful. **(Fig. 7)** Oral feeds were started for the patient on post-operative day 8. There was transient neuropraxia of the facial nerve which recovered over a few weeks. She was on regular follow-up with us for 2 years with active physiotherapy for facial musculature and mouth opening exercises. **(Fig. 8)** The patient went back to school with improved self-confidence and was readily accepted by her peers.



Fig. 1 – Clinical photograph of the patient showing hemifacial atrophy of the right side with En Coup de Sabre of the right side of the forehead



Fig. 2 – 3 Dimensional Computed Tomographic Pictures showing hypoplasia of the facial bones of the right hemi-face causing facial asymmetry



Fig. 3 – Pre-operative planning showing the region where the de-epithelialised differential thickness free anterolateral thigh flap would be placed

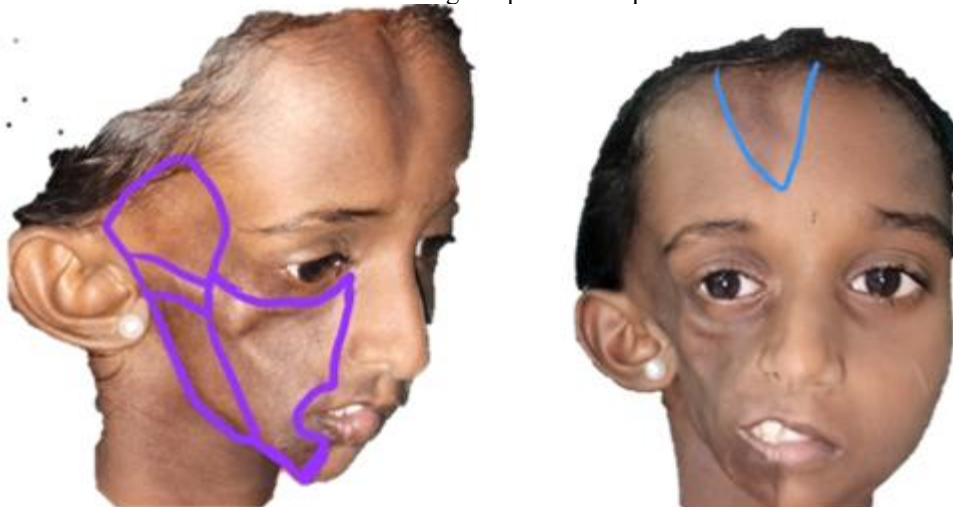




Fig. 4 – Areas marked where the de-epithelialised differential thickness free anterolateral thigh flap would be placed and the site of injection of the fat graft



Fig. 5 – Pre-operative planning and marking of the de-epithelialised free anterolateral thigh flap with areas of thinning of the flap (shaded areas)



Fig. 6 – Zones where the de-epithelialised differential thickness free anterolateral thigh flap would be inset



Fig. 7 – Early post-operative photographs (1 week)



Fig. 8 – 2 year follow-up photographs

DISCUSSION

PRS is a rare, sporadic, neurocutaneous degenerative disorder characterized by a slow progressive tissue atrophy on one side of the face, involving the facial tissue, including muscles bones and skin. (14,15) It exhibits a female predilection seen in 1:70000 of the population, with a male-female ratio of 1:2.2, and a tendency for the left side of the face to be more commonly involved. (6,13,14,16) The onset of the disease is usually in the first two decades of life, with a progressive phase lasting for 2 to 20 years. This is followed by a stable or a burnt-out phase where the disease process arrests in most patients. (5,17) In some cases, disease flare or worsening may be associated with stress including surgery. (1) There is atrophy of the skin and subcutaneous tissues, and as the disease progresses, they could also develop atrophy of the underlying muscular, cartilaginous, bony, and glandular structures but typically of adipose tissue. (7) Clinically, patients present with varying degrees of atrophy primarily affecting subcutaneous fat and skin and progressing to muscular, osseous and glandular structures. The damage is mostly confined to the distribution of the trigeminal nerve and is classified as mild, moderate and severe, depending on the extent of hemifacial atrophy. Guerrerosantos et al. classified the syndrome into four types, namely types 1, 2, 3, and 4 tissue depressions. Types 1 and 2 show mild involvement of the soft tissues of the face and the skin, while types 3 and 4 are severe diseases with involvement of bone and cartilage. Type 1 is a very mild depression or soft tissue loss, often seen during the acute phase of the disease, with atrophy of soft tissues limited to a single sensory branch of the trigeminal nerve, without rotation of the occlusal plane. In Type 2, there is reduced thickness of facial soft tissues (skin and subcutaneous fat), with no bone or cartilage involvement. There is involvement of nostrils and lip commissures in addition to the trigeminal dermatome, extending to two branches of the trigeminal nerve with no atrophy of skeleton and relatively horizontal occlusal plane. In Type 3, the soft tissue is thinner than in Type 2, with initial bone and cartilage involvement. Type 4 is the most severe type of facial depression, where the skin is often close to the bone and significant bone and cartilage involvement is present, encompasses atrophy affecting all three branches of the trigeminal nerve. There are also rotation and transverse cant of occlusal plane, atrophy of maxilla, mandible and zygoma, deviation of chin and nose to the affected side.

Diagnosis of Parry Romberg syndrome is based on patient history and clinical examination and is corroborated by imaging and histopathological studies. (18,19) The etiology and incidence are unknown. The primary cause is proposed to be a cerebral disturbance of fat metabolism. Viral infection, trauma, endocrine imbalance, neuro-vasculitis, autoimmunity, heredity, vascular abnormalities and dysfunction of the sympathetic cervical ganglia are considered to be pathogenetic. (5,7,8,20,21) Current evidence strongly supports PRS as an autoimmune

disorder, indicated by the presence of coexisting autoimmune conditions like systemic lupus erythematosus and rheumatoid arthritis, as well as clinical improvement with immunosuppressive therapy during active disease. (5,7)

Clinically, the initial signs are thinning of the skin and subcutaneous tissues, usually preceded by alopecia areata, or vitiligo. (15,22) Atrophy typically begins at the maxillary or periorbital region but may extend to other areas of the face, with accompanying changes in pigmentation of the overlying skin. (21,23) Around 10% to 35% of patients show ocular involvement; the most encountered manifestation is enophthalmos characterized by sunken eyes due to loss of adipose tissue around the orbit and atrophy of the retrobulbar fat. (5) Around 50% of affected individuals show dental changes. (24) The most frequent neurological manifestation of Parry-Romberg syndrome are seizures (60.5%) and focal seizures ipsilateral to brain calcifications are also common (50%). Headache (44%), behavioural disturbances, and trigeminal neuralgia (8.5%) may also be present. (9,25,26) Neurological complications occur in 15%-20% of cases, presenting as headaches, seizures, trigeminal neuritis, facial paraesthesia, cranial nerve dysfunction and fixed focal neurological deficits. Headaches and seizures, often refractory to treatment, are the most common. (5) Frank elevation of the mouth, nose, facial midline, and dental midline is characteristic. Upper lip atrophy leads to exposure of maxillary anterior teeth. There can be atrophy of the tongue with deviation of mouth and nose to the affected side. (27) Dental involvement in the form of overcrowding and short crowns and roots of teeth may be observed in some patients. Cognitive and behavioural problems have also been reported. (5)

Neuroimaging findings in PRS are reported to be abnormal in 20%-50% of patients, with those exhibiting neurological symptoms more likely to show positive findings on brain imaging. (21) CT scan best demonstrates calcifications and bony changes of the hemicranium and facial skull while MRI is sensitive in identifying gliosis and atrophy. These abnormalities are usually non-enhancing, but iodinated contrast medium and gadolinium were useful in a few cases in showing areas of active inflammation on head CT and MRI, respectively. (28) Typical findings on MRI are hyperintense white matter lesions on T2-weighted and FLAIR sequences, which are distributed predominantly on the ipsilateral hemisphere but are sometimes bilateral. The Computed Tomography (CT) and Magnetic Resonance Imaging (MRI) facial findings of PRS include varying degrees of hemiatrophy by fat plane obliteration, ipsilateral deviation of the aerodigestive tract and enophthalmos. The most common CT and MRI intracranial findings of PRS include ipsilateral subcortical calcifications, white matter changes, and ipsilateral focal or diffuse brain atrophy. Other findings include cortical gyration loss, encephalomalacia, ventricular dilatation, corpus callosum infarct, leptomeningeal thickening and enhancement and hamartomas. (29) Vascular abnormalities of PRS such as micro or macro-hemorrhages, malformations, stenoses and aneurysms have been observed. Ultrasonography can identify sclerosis, and monitor the disease activity and progress of treatment by taking measurements of dermal thickness and noting the echo pattern of the affected sites. Colour Doppler is also useful in detecting dermal blood flow, and increased flow suggests active disease. Increased blood flow and hypoechogenicity of the ipsilateral parotid and salivary glands also suggest inflammation and an active disease process. (5)

The differential diagnosis for PRS includes en coupe de sabre (ECDS), hemifacial microsomia (first and second branchial arch syndrome), Goldenhar syndrome, Rasmussen encephalitis, post-traumatic atrophy, and partial lipodystrophy (Barraquer-Simon Syndrome). (24) En coup de sabre (ECDS), a variant of linear scleroderma, shares similar epidemiological and clinical features with PRS. These conditions often coexist and are believed to be entities on the same spectrum of disease, with reported cases of ECDS converting to PRS. (5,11,21) Distinguishing between these two conditions can be challenging, however, bony, muscular, and cutaneous atrophy favour a diagnosis of PRS, while predominant involvement of the fronto-parietal region with no extension below the eyebrow and cutaneous sclerosis are more characteristic of ECDS. (5,7,21) Hemifacial microsomia and Goldenhar syndrome are congenital, nonprogressive disorders. Rasmussen encephalitis is an acquired inflammatory autoimmune disorder that manifests in the paediatric population with intractable partial epileptic seizures and progressive neurological deficits. While similar brain imaging findings may be observed in Rasmussen encephalitis and PRS, a history of refractory epileptic seizures with the absence of facial asymmetry should favour a diagnosis of Rasmussen encephalitis. (21)

Parry-Romberg syndrome has a significant biopsychosocial impact on the patient's life due to functional and aesthetic limitations. (19) With regard to management, it is always a challenge and involves a multidisciplinary team consisting of paediatricians, dermatologists, plastic surgeons, intensivists, radiologists, dentists, speech therapists and psychologists. (5,8) The initial goal of treatment is to slow down or halt the disease progression. Methotrexate at a dose of 15 mg/m² given orally or as a subcutaneous injection once a week in combination with a systemic corticosteroid at a dose of 0.3–1 mg/kg/week given orally or as an injection is recommended as first line treatment for active disease. Other drugs like hydroxychloroquine, mycophenolate mofetil, cyclosporine and cyclophosphamide may be tried in those patients who have not responded to methotrexate. (5,30)

Treatment of PRS can be challenging, as there are currently no standard treatment guidelines in place. The primary objective of treatment is to impede the progression of the disease using immunosuppressive agents and to manage seizures through the administration of anticonvulsants or lobectomy. PRS-related scleroderma is treated with immunosuppressive therapies, topical and systemic corticosteroids, immunomodulators and plasmapheresis. Antimalarials, antibiotics, vitamin D3 analogs and penicillamine are treatment options that have demonstrated variable responses. (31) The success of these therapeutic measures however varies among

individuals. (5,7) Surgical interventions to address cosmetic disfigurement are typically employed once the disease has stabilized. This approach necessitates a multidisciplinary strategy and may involve procedures such as pulse dye lasers, autologous fat grafts, muscle flap grafts, silicone injections, collagen, porous polyethylene implants, bone augmentation, free tissue transfer and orthodontic rehabilitation. (7,21) Additionally, patients benefit from psychological support, highlighting the importance of addressing the emotional and psychological aspects of living with PRS. (32) The grafting of cartilage, bones, fat, and skincare treatment are reserved for more severe cases. (33,34) The main aim of psychological treatment is the social reintegration of these patients. (34)

CONCLUSION

PRS is an uncommon self-limiting condition characterized by slow progressive hemifacial atrophy, often accompanied by changes, ranging from mild to severe, involving the skin, subcutaneous fat, muscles, cartilage and bones, neurologic, ophthalmologic and cutaneous manifestations. The exact etiology and pathophysiology are still unknown. A precise clinical diagnosis necessitates comprehensive knowledge of the condition, detailed review of the patient's medical history, thorough physical examination and neuroimaging. Imaging assessments also facilitate the exclusion of other differential considerations, help monitor disease progression and evaluate post-treatment responses. This syndrome causes aesthetic, functional and psychological problems, which require a well-planned, specific and multidisciplinary treatment. In this paper, we presented a case of Parry Romberg syndrome with classical features in a young girl. We corrected the deformity using a surgical approach of a differential thickness de-epithelialised free anterolateral thigh flap for the face and free fat graft of the forehead deformity. She will also need a comprehensive orthodontic and final restorative treatment. We achieved good results with high satisfaction of the patient after the initial surgery.

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