Acta Dermatovenerol APA

Acta Dermatovenerologica
Alpina, Pannonica et Adriatica

2020;29:193-199
doi: 10.15570/actaapa.2020.39

Parry–Romberg syndrome: a mini review

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Abstract

Parry–Romberg syndrome (PRS) is a rare disorder of uncertain etiology that is characterized by progressive atrophy of the soft and hard tissues of face, typically occurring in the first 2 decades of life. It is more commonly seen in females. The disease progresses slowly with gradual atrophy, frequently associated with neurological, ophthalmological, and other system involvement, resulting in secondary complications. The severity of deformity varies depending on the age of onset of disease. Those in whom the disease starts at an earlier age will have more severe deformity. Due to the visible facial deformity, such patients usually suffer from social and psychological trauma. Management is mainly cosmetic, which is carried out after disease progression has stopped and stabilized. This brief review describes PRS in detail and compares it with linear morphea en coup de sabre (ECDS), its close differential, which is likely to be a milder variant sharing the same spectrum of disease.

Keywords: hemifacial atrophy, linear morphea en coup de sabre, Parry–Romberg syndrome, progressive hemifacial atrophy, Rasmussen's encephalitis

Received: 21 April 2020 | **Returned for modification:** 27 July 2020 | **Accepted:** 27 September 2020

Introduction

Parry–Romberg syndrome (PRS), also called progressive hemifacial atrophy (PHA), is an acquired disorder of uncertain cause. Caleb Parry was the first to report it in 1825, and Moritz Romberg described it in 1846 (1). Albert Eulenburg (1871) introduced the term *progressive hemifacial atrophy* (2). There are reports of evidence of PRS existing over 2,000 years ago because two of 200 colored mummy portraits were found to have PRS (3). It is seen more commonly in females beginning in the first 2 decades of life. There is progressive and variable hemifacial loss of soft tissue, which can extend deeper to osseocartilaginous tissues. This is progressive for 2 to 20 years, and after that a stabilization stage is achieved (1, 4). Due to atrophy, these patients usually develop physiological and aesthetic deficits with associated complications such as neuralgia, migraine, epilepsy, and ocular and dental problems depending on the area of involvement.

Etiopathogenesis

The etiology of PRS remains unclear. Many theories regarding the possible etiology of PRS have been postulated, including the following.

Trauma: About 24 to 34% of PRS patients report a history of trauma. This may be trauma related to surgeries such as surgical removal of the thyroid, tooth extraction, or obstetric procedures such as the use of vacuum or forceps during childbirth. Accidental trauma has also been reported (5, 6).

Neurovascular: Neuro-vasculitis of the lymphocytic type has been incriminated in the etiology of PRS. Chronic lymphocyte-mediated injury to vessels is followed by partial regeneration of the lining endothelium along the branches of the neurovascular bundle containing the trigeminal nerve (7). A good percentage of individuals have a latent form of varicella-zoster virus (VZV) in the ganglia associated with the trigeminal nerve. Once reactivation of VZV occurs, the infection can reach the trigeminal nerve, leading to infiltration of the short and long ciliary nerves along with their surrounding vasculature with lymphocytes, causing disease (8).

Autoimmune: Several studies have mentioned PRS as a manifestation of scleroderma based on similar histopathology as well as frequent overlap of the two entities in the same patient (4). PRS has been reported to be associated with a number of autoimmune diseases (Table 1) and various autoantibodies have been reported in PRS patients. To date, the importance of such antibodies in PRS is questionable. It has been observed that most of these patients had an overlap with morphea en coup de sabre (ECDS). In a case

Table 1 | **Diseases associated with Parry–Romberg syndrome (PRS) (20, 56–59).**

Dermatological	Non-dermatological		
	Congenital	Autoimmune	Infections
Segmental vitiligo	Unilateral trunk and limb atrophy	Autoimmune hemolytic anemia	Borreliosis
Bandlike alopecia	Poland syndrome	Lupus erythematosus	Herpes
Hyperpigmentation	Hypertrophic cardiomyopathy	Scleroderma	Otitis
Port-wine stain	Congenital lower limb hypoplasia	Hashimoto thyroiditis	Dental infections
Klippel–Trénaunay syndrome	Congenital ipsilateral cerebral atrophy	Graves' disease	Diphtheria
Raynaud syndrome	Supernumerary nipple	Primary biliary cirrhosis	Syphilis
Lupus profundus	Microphthalmia	Inflammatory bowel disease	Rubella
Morphea	Congenital torticollis	Rheumatoid arthritis	Tuberculosis
Linear morphea en coup de sabre	Renal malformations	Ankylosing spondylitis	
Poliosis		Multiple sclerosis	
Hemangiomas			

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study of PRS by Garcia-de la Torre et al., 57% of patients had antinuclear antibodies (ANAs). Rheumatoid factor (RF), anti-histone antibodies, and anti-centromere antibodies were present in 36%, 21%, and 14% of patients, respectively. However, the anti-ds-DNA antibody was not present (9). A case of PRS in a 21-year-old male was described by Gonul et al. as exhibiting right-sided facial atrophy. Screening for various antibodies revealed positivity only for the anti-ds-DNA antibody, whereas all others were non-reactive (10).

Infection: Some studies support an infectious etiology of PRS, proposing cutaneous infections such as *Borrelia burgdorferi* (Lyme disease), herpes, and systemic infections such as dental infections, otitis, diphtheria, syphilis, rubella, and tuberculosis as possible infectious triggers (4, 11, 12). However, others have suggested that there is no association between PRS and anti-Borrelia antibodies, thus making this debatable (13).

Genetics: A genetic basis of PRS is controversial because there is only one case report of familial PRS to date (14). Another report of PRS occurring in only one of two monozygotic twins implies that there is probably no role of genetics in the etiology of PRS (15).

Sympathetic nervous system dysfunction: Irritation and dysfunction of the cervical sympathetic trunk has been proposed to play an etiological role in PRS. This is supported by ipsilateral development of trophic changes in animals following superior cervical ganglion ablation, mimicking clinical manifestations of PRS in humans such as hemifacial atrophy, enophthalmos, and bone atrophy on the ipsilateral side. Some patients also complain of ipsilateral Horner's syndrome, which further supports this hypothesis, whereas most patients have normal responses to standard autonomic function testing. Thus, it is suggested by multiple authors that sympathectomy can halt the progression of facial atrophy in PRS (5).

Other: Disturbances in fat metabolism, endocrine disorders, radiation exposure, and other causes have also been proposed (16).

Currently, laboratory and histopathological findings and the treatment response of PRS with immunosuppressive and immunomodulators strongly support an autoimmune and inflammatory etiology, similar to morphea ECDS.

Clinical features

Cutaneous

PRS presents as slowly progressive atrophy of the face. This atrophy usually follows the branches of the fifth cranial nerve. There is hemifacial atrophy involving skin and soft tissues that can extend deeper to muscles and underlying osseocartilaginous tissues. Cutaneous induration is rarely present (4). The atrophy and deformation progress very slowly, leading to hemifacial atrophy. This is usually associated with enophthalmos on the affected side, lingual atrophy, and deviation of the nose and mouth toward the diseased side (Figs. 1–4). The ultimate magnitude of deformity depends on the duration of the disease and the extent of involvement of the face. Figures 1–4 show the chronological events of a severe case of PRS, ultimately with the loss of an eye due to the disease (6). In the long term, the deformity becomes more visible, causing cosmetic and physiological deficits (17). Cessation of facial atrophy may occur in these patients; however, it becomes difficult to distinguish between normal disease progression and slow progression and stabilization of the disease. PRS is characterized by absent or minimal skin induration and atrophy in a paramedian location that usually extends down the face along one or more branches of the trigeminal nerve (18).

In contrast, linear morphea ECDS is a type of localized scleroderma. It presents as sclerotic, depressed indurated plaques in a linear distribution usually involving one side of the forehead and frontoparietal scalp resembling a wound from a sword (hence the name *en coup de sabre*) (19) (Fig. 5). It rarely extends down the entire face, including the nose, upper lip, chin, and neck (20, 21). There are many studies distinguishing PRS from morphea ECDS based on clinical and histopathological features (22–27). The most important clinical features differentiating PRS from ECDS include paramedian atrophy in PRS without significant skin induration and associated atrophy having a tendency to extend down the face with mandibular and orodental involvement.

In a study involving retrospective evaluation of 13 ECDS and nine PRS cases using clinical photos and histopathological slides, the authors found clinically cutaneous sclerosis (eight of the 13 ECDS cases and none of the PRS cases), scarring alopecia and hyperpigmentation in ECDS, and total hemifacial involvement with ophthalmological changes in PRS. In a histopathological examination, significant overlapping of features was seen between the two, such as collagen sclerosis (all cases of ECDS versus 2/9 cases of PRS), atrophic adnexa (11/13 ECDS versus 3/9 PRS cases), and infiltrates of mononuclear cells (all cases of ECDS versus 6/9 cases of PRS) (23).

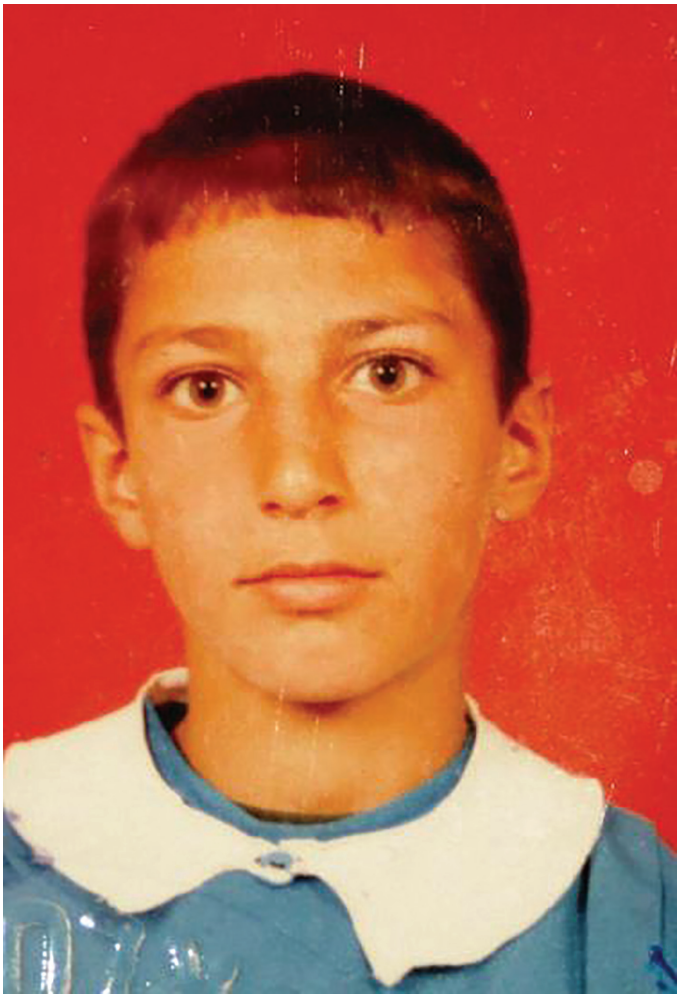

Figure 1 | At 8 years of age, very slight atrophy of the left side of the patient's face was apparent (6).

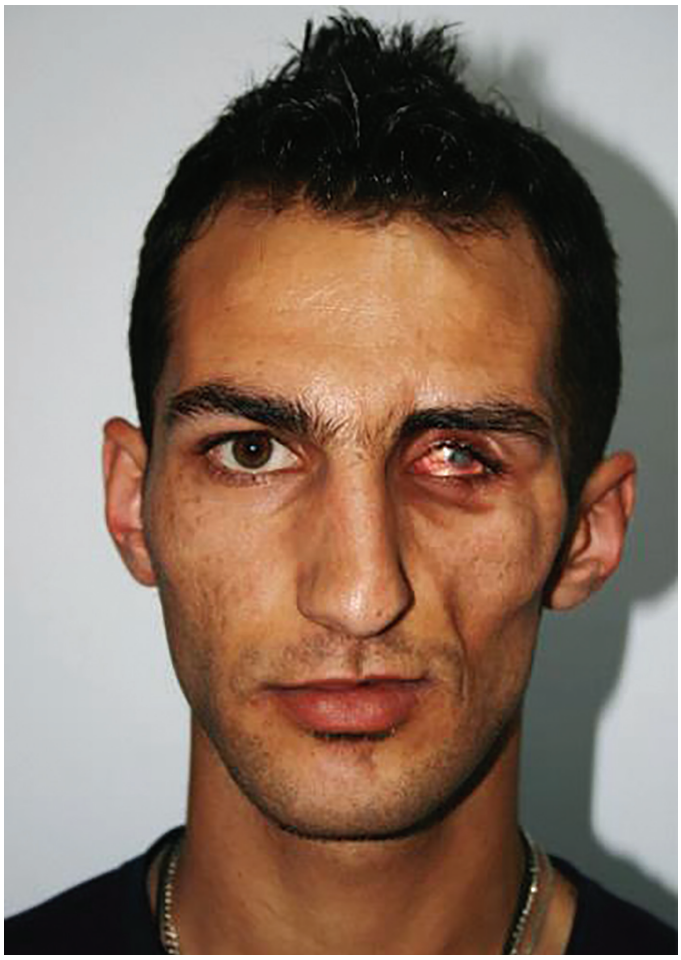

Figure 2 | Frontal view of the patient at 23 years of age, showing severe atrophy of fat and muscle tissue, and of the zygomatic arch on the left side, as well as shrinkage of the left eyeball and corneal atrophy (6).

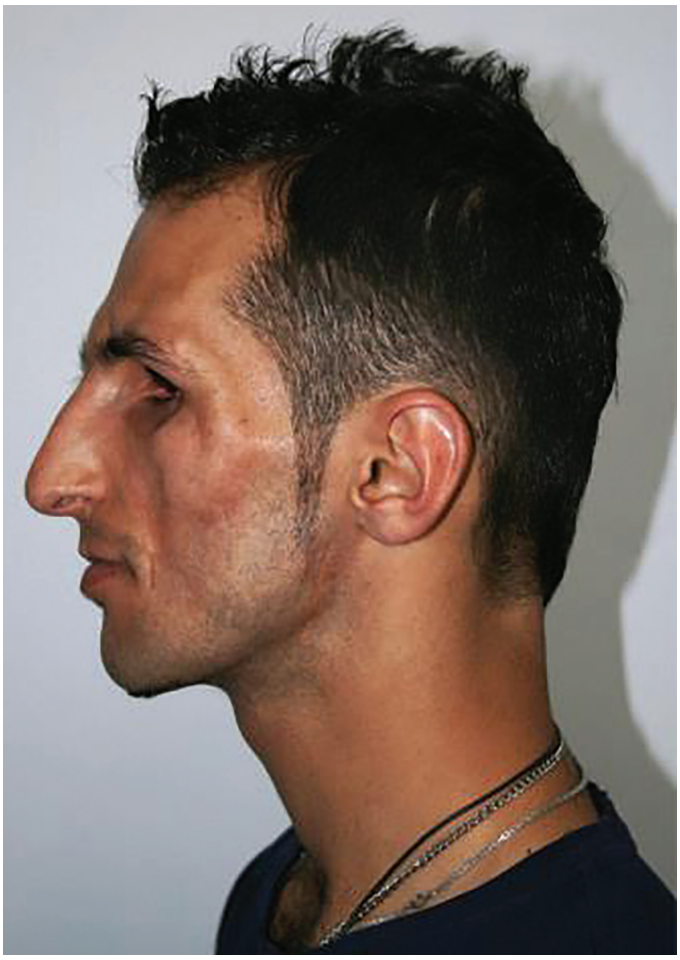

Figure 3 | Lateral view of the left side of the face (6).

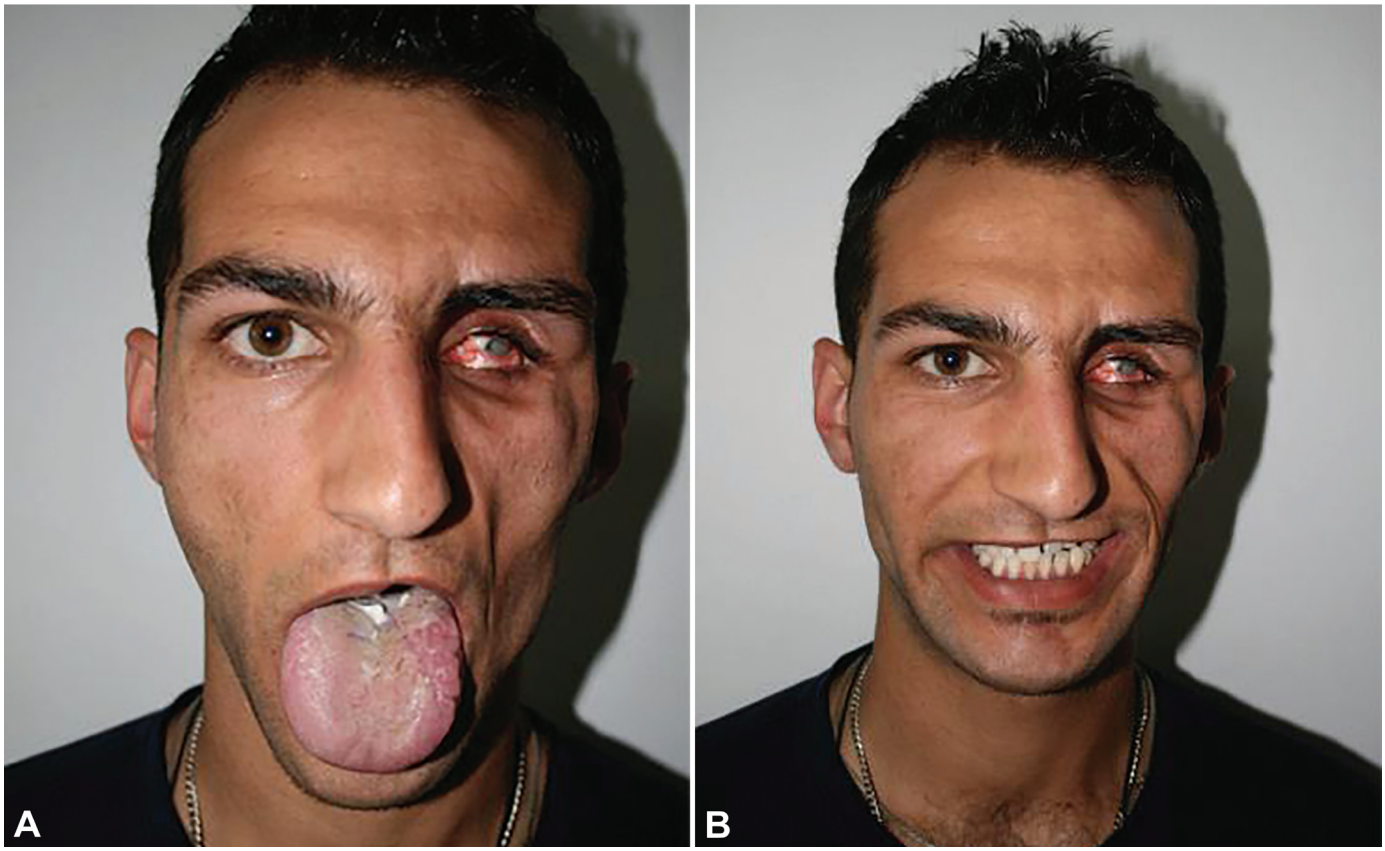

Figure 4ab | a) Atrophy and fissuring of the left hemiglossus (6). b) Maxillary atrophy has displaced the left upper teeth in an upward and backward direction (6).

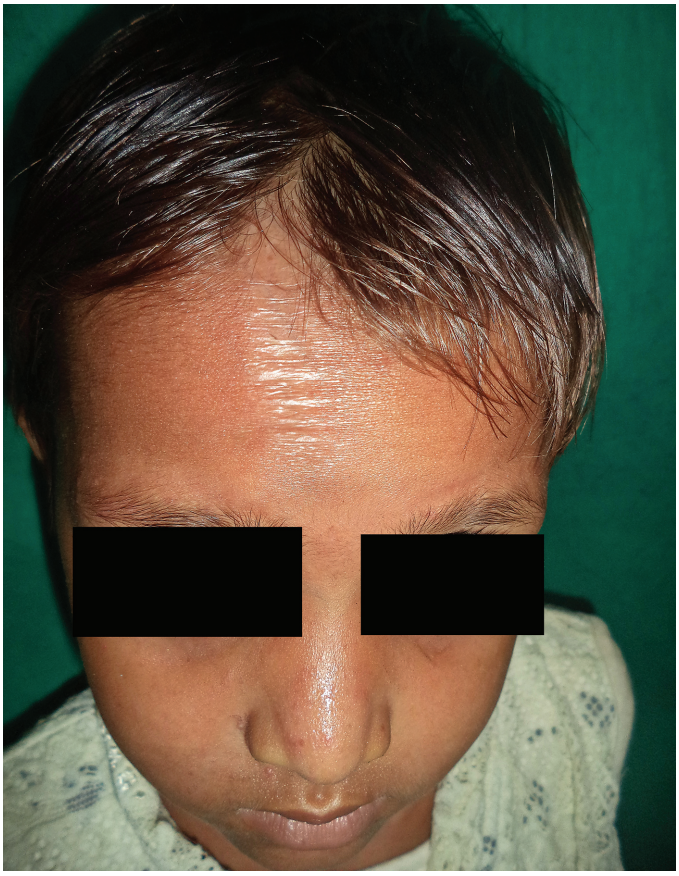

Figure 5 | En coup de sabre (ECDS) in a child. There is a linear atrophic plaque on the right paramedian side of the forehead extending upward to the frontal scalp with associated scarring alopecia.

Extracutaneous/systemic findings

PRS can have extracutaneous involvement, the most common system involved being the central nervous system and ocular involvement, followed by dental and rarely otorhinolaryngological involvement.

Central nervous system: PRS can affect the neural and vascular tissues of central nervous system with or without peripheral nervous system involvement. Among neurological manifestations, headaches and seizure disorder are the most common. Seizures (simple or complex partial type) that arise from cerebral cortex of the same side and are resistant to treatment have frequently been reported. Neuropathies involving several cranial nerves (the third, fifth, sixth, and seventh) have been described (24, 28–31). Impingement of the trigeminal nerve due to vascular inflammation and destruction of surrounding bone causes secondary trigeminal neuralgia in these patients. This manifests as chronic facial pain that is usually resistant to treatment (32, 33). Other findings include speech disorders, cognitive impairment, behavioral disorders, paresthesias, asymptomatic white-matter changes, infarction, hemorrhage, vascular malformations, and cysts (18).

Ocular: Ocular findings in PRS are commonly seen due to involvement of the eye and periocular soft and hard tissue in the disease process, ranging from subtle impairment of vision to complete blindness (34). As the tissue atrophy extends deeper, such patients usually suffer from enophthalmos and dysfunction of the orbitalis muscle. Apart from this, the disease process can affect other structures such as the eyeball, other extraocular muscles, and eyelids. In some cases when the disease is severe, restrictive strabismus may ensue (35–38). PRS patients may occasionally develop retinal vasculitis, which supports a vascular etiology. Less common findings reported in PRS include uveitis, neuro retinitis, glaucoma, papillitis, cataracts, changes of the retinal pigment epithelium, and heterochromia iridis (39–45).

Dental and oral: Bone resorption in PRS can lead to hypoplasia of the mandible and maxilla, resulting in crowded, shorter tooth crowns and roots. Involvement of soft tissues causing atrophy of the lips, tongue, salivary glands, and gingiva leads to difficulty eating and smiling. Abnormally short and crowded teeth are prone to early caries. Involvement of masticatory muscles can cause trouble smiling, speech problems, painful chewing, spasms, jaw locking, and pain in the temporomandibular joint (36, 46–53).

Otorhinolaryngological: The nose can be deviated secondary to skeletal atrophy with secondary changes. An association of PRS with dysphonia has also been described (54, 55).

Associations with Parry–Romberg syndrome

Various dermatological and non-dermatological disorders have been described as associated with PRS (Table 1) (20, 56–59).

Differential diagnosis

ECDS forms a close differential for PRS, and occasionally the two become indistinguishable. The two disease entities share several features, such as similar age of onset, slow progression, comparable neurological and ocular complications, and a similar response to immunosuppressive agents. They may overlap in the same patient at the same site and may rarely coexist in same patient at different sites (Fig. 6). On histopathology both show dermal sclerosis with thickened collagen bundles, chronic lymphocytic infiltrate, and atrophy of adnexa (Fig. 7). The characteristics of the two diseases are summarized in Table 2 (10, 18, 23, 31).

Rasmussen's encephalitis (RE), congenital disorders with facial asymmetry, acquired posttraumatic atrophy, partial lipodystrophies, and rarely unilateral facial hypertrophy may mimic PRS. Rasmussen's encephalitis is considered an immune disorder that usually affects half of the cerebrum. It manifests with seizures that are refractory to treatment and hemiplegia, which is progressive in nature. Onset of disease is usually in children under age 10 (60–62). Radiologically, hyperintense T2 signal areas are present in one side of cerebral hemisphere, which may later develop into cerebral atrophy. Such imaging features are usually difficult to differentiate from those of PRS patients (61, 62). RE and PRS have been reported to coexist in the same patient. This suggests a probable pathophysiological relationship between the two (60, 63). Disorders with facial asymmetry such as Goldenhar syndrome and hemifacial microsomia (a syndrome involving the first and second brachial arches) may clinically look similar with unilateral atrophy. However, these disorders are nonprogressive in nature and considered congenital disorders (64, 65). In hemifacial hypertrophy, there is facial asymmetry attributed to hyperplasia and overgrowth of one side rather than atrophy, which is seen in PRS (65, 66). Barraquer–Simons syndrome is a type of partial lipodystrophy that may mimic PRS. However, manifestations in the former are typically bilateral. Silent sinus syndrome may sometimes simulate PRS. However, it can be differentiated from PRS by its late onset of presentation, atelectasis and opacification of the maxillary sinus on imaging, and obstruction of the ostiomeatal complex on the affected side (67). Very rarely, unilateral post traumatic atrophy may mimic PRS.

Table 2 | **Features of Parry–Romberg syndrome (PRS) and linear morphea en coup de sabre (ECDS) (10, 18, 23, 31).**

Parameter	Parry–Romberg syndrome	Linear morphea en coup de sabre
Description	Progressive hemifacial atrophy of soft and hard tissue of one side of face, usually left, frequently associated with ocular and neurological abnormalities	Localized scleroderma with indurated and hyperpigmented skin over face, usually upper face above eyebrows with prominent deep paramedian groove separating normal and involved tissue with relative sparing of deeper soft and hard tissues
Age of onset (years)	13.6	10
Female: male	3:1	2:1–3:1
Skin involvement	Normal to hyperpigmented skin, normal-appearing hair with minimal or no skin induration	Shiny indurated thick skin with hypo- or hyperpigmentation and alopecia
Facial involvement	Mostly lower half, predominantly maxilla and mandibular regions	Mostly upper face, above eyebrows with linear depressed paramedian groove (en coup de sabre)
Scalp alopecia	No	Very common, sometimes involving eyebrows
Involvement of deeper structures	Yes, including muscles/bones	Mostly superficial, rarely deeper involvement
Extrafacial involvement	Central nervous system, ocular, otorhinolaryngological, oral, and dental	Less frequent
Preceding induration / skin inflammation	No	Yes
Histopathology	Dermal sclerosis with inflammation in early stages with fat atrophy and relative shrinking of adnexa at later stages	Severe dermal sclerosis with mononuclear cell infiltrates and loss of adnexa
Autoantibodies related to scleroderma/ANA	Not always seen	Mostly elevated
Treatment	Topical calcineurin inhibitors, systemic methotrexate, corticosteroids, other immunosuppressants, and surgical modalities once disease stabilizes	Topical steroids, calcineurin inhibitors, systemic methotrexate, corticosteroids, etc.

ANA = antinuclear antibody.

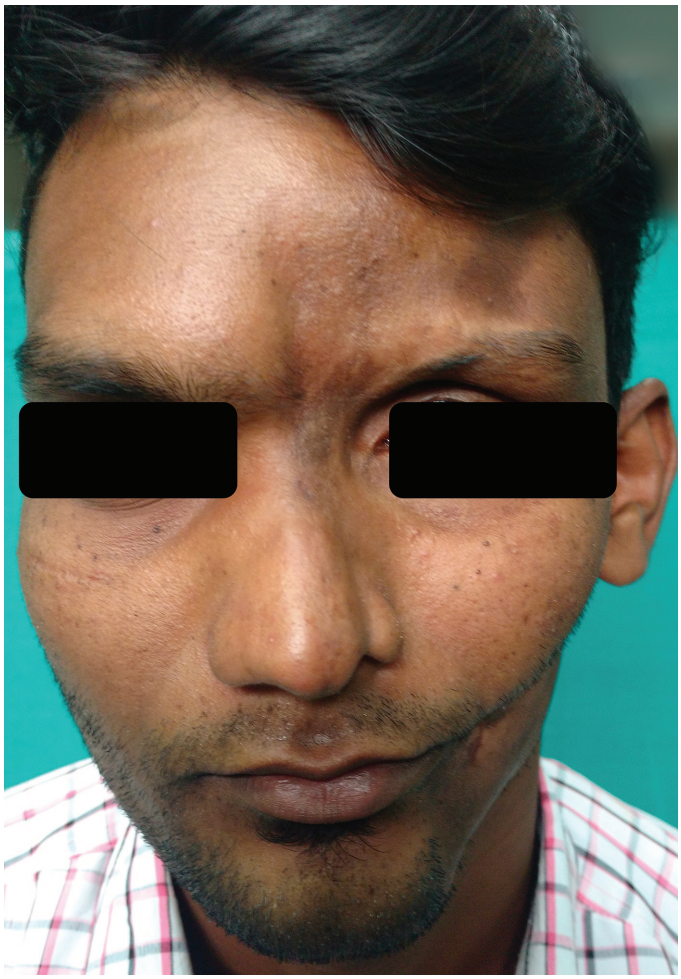

Figure 6 | En coup de sabre (ECDS) and Parry-Romberg syndrome (PRS) in the same patient on the same side. This patient also had scarring alopecia in a linear pattern on the frontal scalp as a result of ECDS. There was visible atrophy on the left side of face behind the angle of mouth that was associated with atrophy of left side of the tongue consistent with PRS. (Image credit: Swetank and Mohammad Adil; MLB Medical College, Jhansi, Uttar Pradesh, India and Jawaharlal Nehru Medical College, Aligarh Muslim University, Aligarh, India).

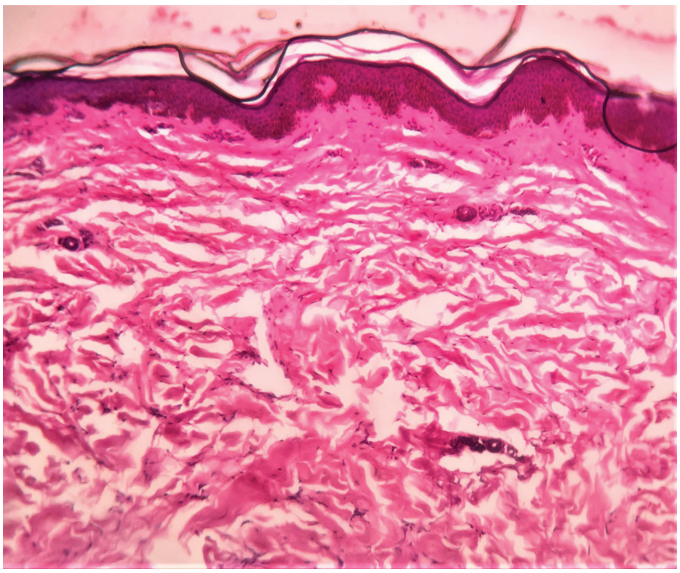

Figure 7 | Histopathology of Parry–Romberg syndrome (PRS) showing mild epidermal atrophy. Dense collagen bundles in the dermis with sparse inflammatory infiltrate. Findings similar to scleroderma (H&E 40×).

Treatment

A multimodal approach is needed in the treatment of PRS. The main objective of treatment is to halt the disease activity with medical therapy followed by surgical intervention for correction of residual deformities. Methotrexate (0.3–1 mg/kg/week) is frequently used as standard therapy in early stages of PRS. Due to delayed action of methotrexate, oral steroids such as prednisolone (1 mg/kg/day) are started with methotrexate and given for up to 3 months. Prednisolone is tapered after the first 2 months and stopped after 3 months, whereas methotrexate is continued for 1 to 2 years based on disease activity. Alternatively, monthly pulse therapy with high-dose intravenous methyl prednisone (1,000 mg for 3 days/month) is also suggested for 6 months to avoid side effects from daily prednisolone therapy (18). Other immunosuppressive agents that have

been tried with variable success include cyclosporine, mycophenolate mofetil, cyclophosphamide, and antimalarials (18).

Surgical treatment for PRS includes repeated and stepwise procedures based on the degree of defect and area of involvement. Timing of surgical intervention in PRS is controversial. One school of researchers is of the opinion that surgical intervention should be delayed until stabilization is acquired. This will help in avoiding multiple surgical interventions. The other group of experts recommends earlier procedural therapy in spite of having active disease to promote normal development of facial structures. Their argument is supported by the fact that patients that underwent earlier surgical interventions scored higher on satisfaction (68, 69). It should be remembered that the appropriate timing for procedural therapy needs to be individualized for each case with guided orthodontic appliances and rehabilitation (70, 71). PRS with mild to moderate atrophy can be corrected with fat grafting, lipoinjection and soft tissue fillers, and other techniques. Severe atrophy needs procedures to augment both skeletal and soft tissues such as bone grafting, implants made of porous and biocompatible polyethylene, injections of autologous fat, grafts of dermal fat, and adipofascial flaps (68, 69, 72, 73).

For a better cosmetic appearance, various plastic surgeries or cosmetic procedures can be performed, including eyebrow lifting, eyebrow repair, lip repair, lip augmentation, nasal reconstruction, facelift, and hair transplantation (18, 72 ,73).

Conclusions

From the current literature review, it seems that both linear morphea ECDS and PRS probably have some autoimmune etiology and belong to same disease spectrum, with one milder form involving superficial tissues and the other severe form having deeper tissue involvement. Although PRS is self-limiting, proper evaluation with early imaging of central nervous system and eye involvement with regular follow-up involving sequential periodic photographs may be helpful in assessing the progression of the disease and planning early orthodontic treatment.

Acknowledgements

We are thankful to M. Kaya, C. Sel Yilmaz, H. Kurtaran, and M. Gunduz (Department of Otolaryngology Head & Neck Surgery, Turgut Özal University Hospital, Faculty of Medicine, Turgut Özal University, Alparslan Turkes Caddesi no. 57, 06510 Ankara, Turkey) and Hindawi Limited, London, UK, for allowing us to use Figures 1–4 using Creative Commons Attribution License 4.0. Thanks to Swetank (MLB Medical College, Jhansi, Uttar Pradesh, India) and Mohammad Adil (Jawaharlal Nehru Medical College, Aligarh Muslim University, Aligarh, India) for contributing Figure 6.

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