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Imaging in Rare Case Parry Romberg Syndrome: Case Report

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ABSTRACT

Parry Romberg syndrome (PRS) defined by the gradual and acquired atrophy of one side of the face. There is still a lack of clarity regarding the pathogenesis of PRS. In the etiology of PRS, immune-mediated mechanisms are assumed to be a fundamental role; nevertheless, there is also the possibility that the autonomic nervous system is not functioning correctly. Performing radiological evaluation and subsequent identification of this condition can help mitigate the impact on one's lifestyle and slow down the disease's advancement. In this report, we present an example of this syndrome in 12-year-old boy and intend to provide a detailed description of the many radiological manifestations of this condition.

Keywords: *Parry Romberg Syndrome, Imaging, Rare Case*

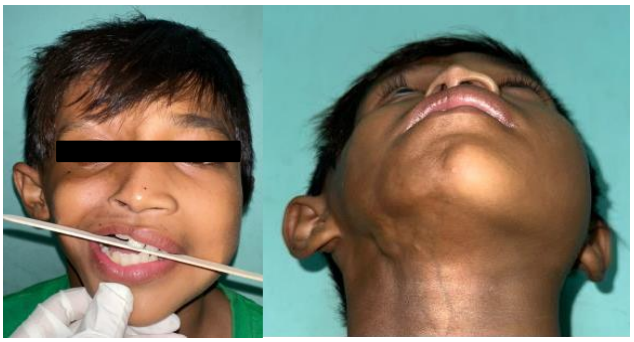
1. INTRODUCTION

Parry Romberg syndrome was found in early 1800 by Parry. Later in 1846, Romberg was the second individual to characterize this condition as an uncommon acquired hemifacial atrophy syndrome affecting skin, subcutaneous tissues, adipose tissue, muscle, and osseous structures. The diverse neurological, ocular, maxillofacial, and dermatological manifestations are not considered diagnostic hallmarks but rather associated clinical findings due to their variability [1], [2]. An indolent progressive phase that develops for twenty years and culminates in a burned-out phase is one of the defining characteristics of the condition. There have been hypotheses that have been proposed to promote autoimmune insults, disturbance of the cervical sympathetic system, vasculitis, traumatic injury, and infections as possible inciting agents; however, a precise aetiology has not been determined as of yet. Positive neuroimaging results are more commonly seen in patients who are experiencing neurological complaints. These results are typically ipsilateral to the area of the face that is afflicted [3].

2. CASE PRESENTATION

A 12-years-old-boy presented to pediatric neurology clinic with a chief complaint his face looked asymmetrical. Gradual facial disfigurement happened for six years. At age 6, he experienced parotitis and it was self-treated by his grandmother. Due to drug allergy, he got dark patches or hyperpigmentation around his cheek and jaw. Upon conducting a thorough history, it was revealed that his face slowly retracted and creased up. The patient had no headache, paraesthesia, visual disturbance,

hearing loss. Physical examination showed facial asymmetry with midline shifting to right-side. *Coup de sabre* found in frontoparietal region along with right frontal atrophy, cheek atrophy, muscle and soft tissue loss. There were also malar depressed, elevated commissure labialis, and hypoplastic alae nasi in right-side. Occlusal canting was discovered with Maximal Intercuspal Occlusion (MIO) of 38 mm as shown in Figure 1. There was no seventh cranial nerve (CN VII) abnormality. He could smile, frown, and lift his eyebrow in both sides. Visual acquity examination in both eyes >2/60.



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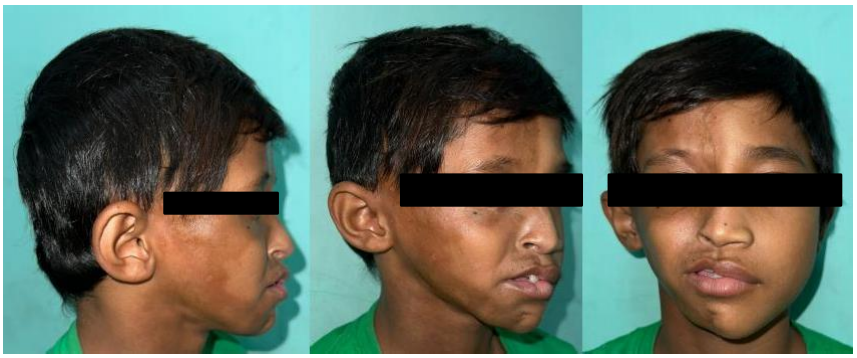

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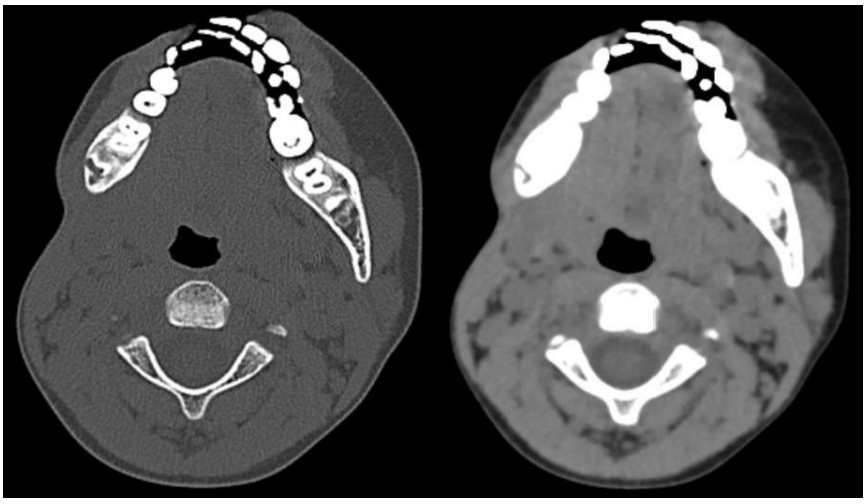
Figure 1. Clinical presentation of patient showing Progressive Facial Hemiatrophy (PFH) on the right side (a) occlusal canting (b) inferior view (c) lateral, oblique, and frontal view (from left to right)



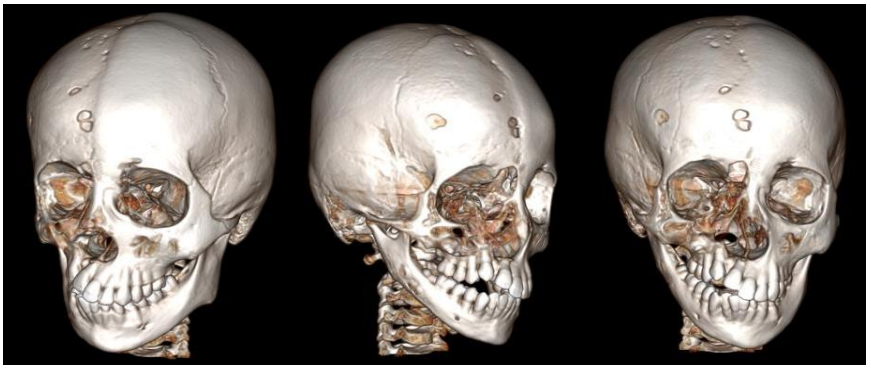
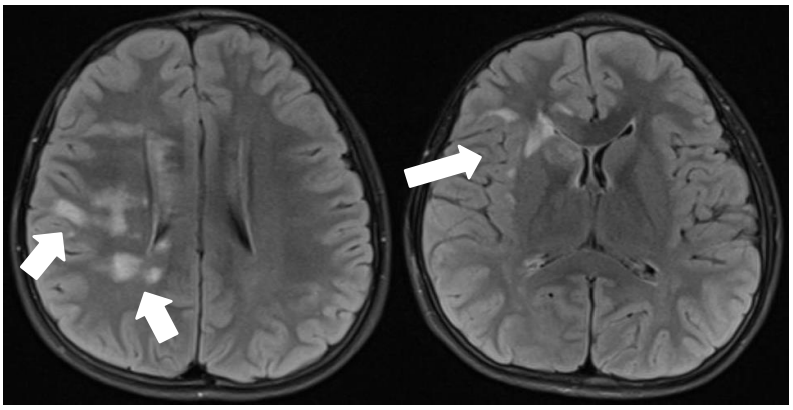


Figure 2. CT-scan and 3D reconstruction showing shrinkage and thinning, deformity in right processus alveolaris, and fat atrophy

Previous CT-scan and 3D reconstruction examination as in Figure 2 showed shrinkage and thinning in right frontal bone, right occipital, right parietal, right zygoma, right maxilla and right mandibula resulted in deformity in right processus alveolaris superior inferior from caninus 2 and fat atrophy in right temporal, right cavum orbita, masticator space, masseter space, parotid and carotid space without muscle atrophy. Prior MRI scan axial view, Figure 3 showed multiple lesions, unclear and regular margin in right frontotemporoparietal white matter. These lesions appeared isointense in T1WI, hyperintense in T2WI/FLAIR, and unrestricted diffusion area in DWI that showed non-contrast enhancement (3a). White and red arrow in coronal view showed atrophy in right facialis, right hyoglossus and right genioglossus (3b).

There was also fluid in left maxilla, ethmoid bilateral, sphenoid bilateral and frontal (3c). It represented white matter disease with muscle atrophy and alopecia in right-side of face. These imaging characteristics are typical for Parry Romberg syndrome. In addition, there are sinusitis in left maxilla and bilateral ethmoid, sphenoid, and frontal.



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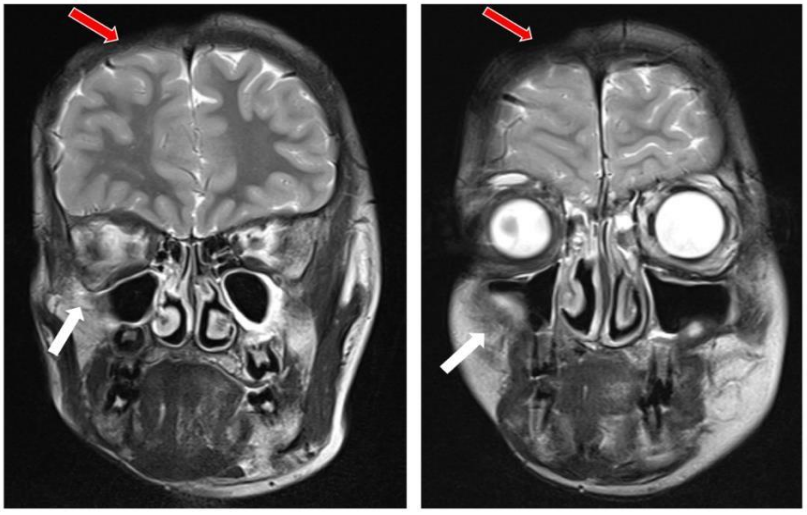

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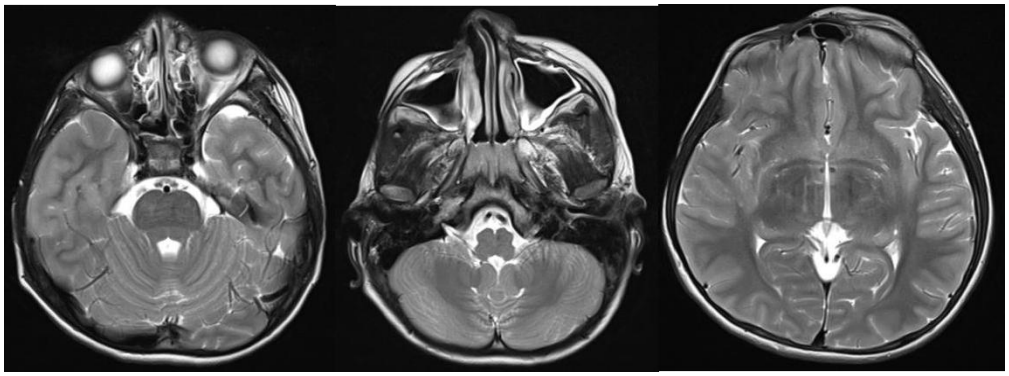

(c)

Figure 3. MRI scan showing (a) multiple brain lesions (b) fat atrophy, (c) sinusitis

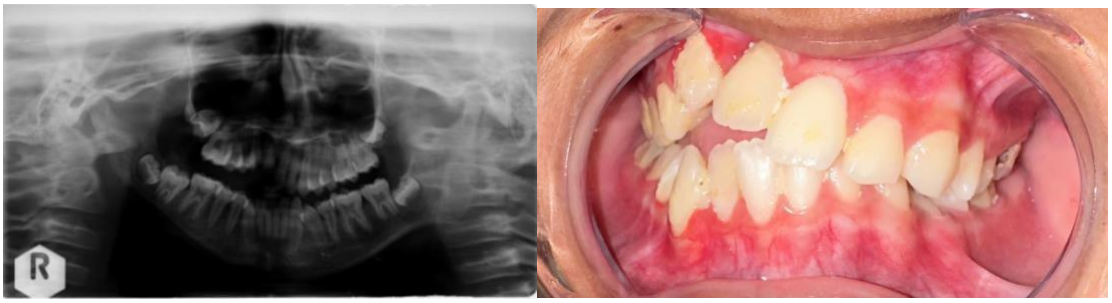

Figure 4. Dental panoramic showing deformity, impaction, caries, and periapical cyst

Dental panoramic radiography showed deformity in the right maxilla and mandibula (see Figure 4). There was impaction in tooth 17 type bucoangular class IIB, impaction in tooth 34 type mesioangular class IIA, dental caries in tooth 12 and 13, missing tooth 11, tooth bud in 28,38,48, amalgam in tooth 45, and periapical cyst tooth 26. The patient was referred to an orthodontist for dental braces. Surgery suggested if there is no more severe manifestation within one year period of follow-up.

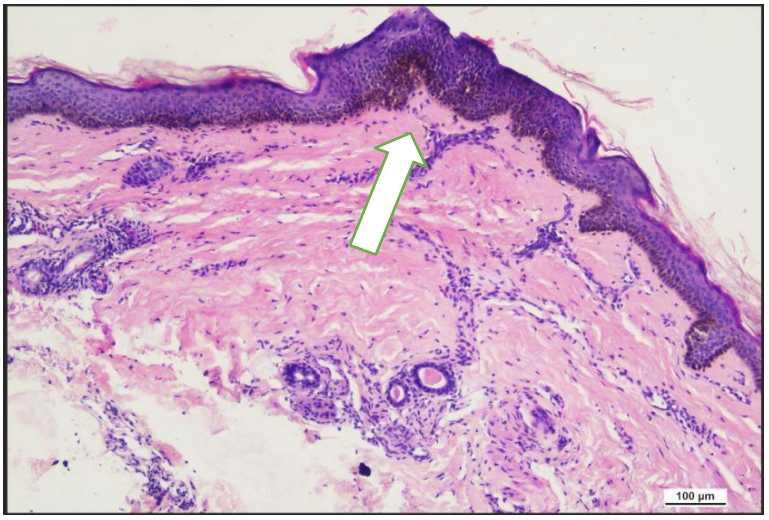

Figure 5. Skin biopsy

Skin biopsy was performed. Microscopic inspection showed sliced tissue, weight <1 gram, size 0.3x0.2x0.2 cm coated with skin 0.3x0.2cm, white gray to brown, and dense-supple consistency (see Figure 5). Macroscopic inspection revealed atrophy epidermis and shortened or flattened rete ridge. In dermal layer, it showed widened capillary vessel with perivascular lymphocyte infiltration and atrophy in skin adnexa.

3. DISCUSSION

Typically, it has been observed that females are more likely to be affected by the sporadic and uncommon disorder known as Parry Romberg syndrome (PRS). This condition does not seem to have any evident regional or ethnic bias of any kind. During the initial years of a person's life, the condition typically begins to show, often progressing slowly. Gradual hemiatrophy of the face over the course of somewhere between two and twenty years, with a little tendency toward the left side. The clinical history and examination, as well as the elimination of other potential causes, are the primary factors that contribute to the diagnosis of PRS. Histopathologic and imaging investigations assist in this process [4], [5], [6]. PRS often affects specific areas of the face that are innervated by one or more trigeminal nerve branches. The degeneration of skin, soft tissues, muscles, and bones on one side of the face is what characterizes this condition. It is sometimes preceded by the hardening of the skin. Tongue hemiatrophy might manifest as intraoral involvement. Gradual shrinking and distortion of one side of the face can result in unilateral facial atrophy, enophthalmos on the same side, and the lips and nose being pulled towards the affected area. Discoloration of the skin, such as increased pigmentation or loss of pigmentation, as well as scarring-related hair loss, may also be seen in the afflicted regions. The ultimate level of deformity may be influenced by the length of time the condition persists. The issue impacts both the visual appearance and the operational capabilities of the face [7].

Facial imaging features that are commonly observed on radiologic examination include variable degrees of hemiatrophy, which is associated by the obliteration of fat planes, ipsilateral deviation of the aerodigestive tract, and enophthalmos as a result of the loss of retrobulbar fat. Neither an abnormal CT attenuation nor an abnormal MR signal has been detected inside the damaged face structures. Intracranially, the following are the findings that are most frequently observed: 1) ipsilateral linear or discontinuous subcortical calcifications in the frontal lobe, 2) white matter hypoattentuation on CT correlating to hyperintense T2 signal on MR imaging, and 3) ipsilateral localized or hemispheric brain atrophy. These are the three specific symptoms that are associated with this condition [8], [9], [10]. In this patient, there was a sudden and gradual hemiatrophy in right-side of his face with intracranial multiple lesions accompanied by muscle- fat atrophy.

Between 15% and 20% of people experience neurological symptoms, with the most common being headaches localized on one side of the head, facial pain, and seizures that may be resistant to treatment. Other neurological symptoms that have been recorded include trigeminal neuritis, facial paresthesia, cranial nerve dysfunction, persistent focal neurologic abnormalities, hemiparesis, and cognitive decline. Previous single center study in London reported out of all the patients, 50% showed abnormalities in their brain scans. The most common results were abnormalities in the white matter signal (43%), dystrophic calcification (36%), leptomeningeal enhancement (29%), and sulcal crowding (21%) [11]. Studies examining neuroimaging in this condition reveal consistent patterns across modalities: hyperintense white matter signals on T2-weighted MRI corresponding with reduced white matter density on CT imaging. Additional findings include brain volume loss affecting one hemisphere, cavernous malformations with microhemorrhages, and calcifications in subcortical regions, predominantly affecting the frontal lobe. Notably, these cerebral abnormalities associated with Parry-Romberg Syndrome occurred with significantly greater frequency in epileptic versus non-epileptic patients (94% versus 47%). Furthermore, the spatial distribution of these brain findings demonstrated stronger concordance with the affected craniofacial region in patients experiencing seizures compared to those without epilepsy (81% versus 20%) [12]. Ophthalmologic symptoms manifest in 10%–35% of individuals and often affect the same side orbit. Enophthalmos is frequently observed due to the degeneration of the fat behind the eyeball. Other possible abnormalities in the eye socket may include inflammation of the uvea and changes in the retina or optic nerve [2], [4]. However, neurologic and ophthalmologic symptoms did not come out in our patient.

Progressive hemifacial atrophy is an incurable illness since it naturally resolves independently. Nevertheless, several therapies have been attempted to halt the progression of the illness. The treatments for this condition encompass the use of oral steroids, D- penicillamine, methotrexate, cyclophosphamide, antimalarials, cyclosporine, and azathioprine. Affected patients require a comprehensive strategy including many disciplines, such as doctors, dentists, psychologists, and speech and hearing therapists. The therapy often involves repositioning adipose tissue that had atrophy as a result of the illness. The surgical or cosmetic procedure should be postponed until there are no further indications of atrophy, indicating that the situation has stabilized [1].

The patient was referred to an orthodontist for fixing occlusal canting using dental braces. Surgery was also planned after one year observation and it was not done due to loss to follow-up. The surgical approach was tailored to the specific illness severity of each patient. The surgical techniques employed consisted of free-fat grafts, cutaneous fat grafts, and bone grafts in conjunction with a temporoparietal fascia flap [13]. There are aesthetic, functional, and psychological issues that are connected with PRS, which is a progressive condition that affects the skin, subcutaneous fat, and bone. Although early onset is linked to alterations in the eyes and teeth, the present case solely presented with facial and mandibular defects. Consequently, PRS can manifest with a diverse range of alterations, varying from moderate to severe As such, the clinical symptoms of this illness are influenced by a great number of circumstances [1].

4. CONCLUSION

Parry Romberg syndrome (PRS) is a rare disorder; diagnosis and identification of PRS are difficult and even highlight challenging task for management. A highly competent medical practitioner is required to evaluate the affected PRS patient. PRS has wide range of symptoms. Surgical treatment is indicated for some patients, however disfigurement may recur after surgery. Radiology plays a crucial role, providing essential imaging techniques for diagnosing and managing PRS.

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