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A RARE CASE REPORT OF PARRY ROMBERG SYNDROME

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ABSTRACT **Objectives:** To evaluate developmental disorder, parenchymal changes in brain with associated anomalies in a case of PARRY ROMBERG SYNDROME. **Methods:** The aim of this work is, through the various presentation of a clinical case, to execute a literature review concerning general features, etiology, pathophysiology, and treatment of progressive hemifacial atrophy. **Findings:** A 22-year-old female started with hyperpigmentation of left upper half of face followed by progressive hemifacial atrophy on same side with significant enophthalmos and tinnitus since the age of 14 years. **Novelty:** Although PRS may be linked to autoimmune aetiology, the current instance does not support that idea.

KEYWORDS : Atrophy, hemifacial atrophy, dermatology, connective tissue disease Parry Romberg syndrome (PRS)

INTRODUCTION

Parry Romberg syndrome is a degenerative disorder involving skin, sub cutaneous tissue, muscle, bone causing degeneration of tissues and management is by facial reconstruction surgery. Despite the prevalence of neurologic and ophthalmologic symptoms, the fundamental cause of the condition is still unclear. Radiologic tests can help rule out other probable diagnoses, assist in disease progression tracking, and evaluate post-treatment responses. The cause of the extremely uncommon condition known as Parry-Romberg syndrome is still unclear. In most cases, the malformations that are sustained are long-lasting. It affects the eyes most frequently and is occasionally accompanied (15 percent of the time) with neurological problems such as trigeminal neuralgia, facial paresthesia, headache, and focal epilepsy. It is more prevalent in females than in males. [1]

It is believed that the disease is an inflammatory condition that affects just one side of the body and is related with a persistent vascular abnormality or neurogenic origin. Only a few instances that appeared to have a genetic or hereditary component have been recorded. Other hypothesised reasons include autoimmune, trauma, and abnormalities of the endocrine system and the metabolic system. Lyme disease has been found in sporadic cases, and positive serology for Borrelia burgdorferi has been found in some of these cases. [2]

The history of the patient and their current clinical symptoms are used to make a diagnosis. Scleroderma is a condition that has similarities to histopathological findings such as epidermal atrophy and dermal fibrosis. Several neurological and vascular abnormalities can be seen on imaging examinations such as cranial computed tomography (CT) and magnetic resonance images (MRI).

In most cases, progressive facial hemiatrophy (PFH) resolves on its own. Immunosuppressants and corticosteroids are the treatments of choice in cases when the illness is actively progressing or when it occurs in conjunction with other autoimmune conditions. Reconstructive treatments like grafting and other forms of plastic surgery may be used to repair the facial abnormalities.

METHODOLOGY

The study was done in Department of General Medicine, Vinayaka Mission's Medical College, and Hospital, Karaikal, Puducherry, India and informed consent were obtained. We report a one-of-a-kind case of Parry-Romberg syndrome in a 22-year-old girl who presented with hyperpigmentation on face on one side followed by wasting of facial group of muscles on left side as observed in this patient.

CASE REPORT

A 22-year-old female patient came to OPD with chief complaints of hyperpigmentation on face on one side followed by wasting of facial group of muscles on left side since the age of 14 years, enophthalmos on left side tinnitus. Symptoms were gradual in onset and progressive in nature. There were no symptoms of neurological disturbances, and her vision was normal. There was no history of family members with similar complaints, and no incidences of trauma or infection were observed to coincide with or precede the onset of the deformity.

The patient was found to have normal vital signs and a normal systemic appearance during the clinical examination. She seemed awake and pleasant, and there were no indications that she was mentally or psychologically unstable. Hearing and speech were not affected in any way. A malformation on the right side of the face caused the face to be asymmetrical. An interpupillary is not developed as a result of the difference in level between the patient's left and right eyes. In comparison to the opposite side, the left maxillary and zygomatic area seemed to have hypoplastic development. Contracture was present on the left side of the upper and lower lip, which was accompanied by increased incisor show and a modest commissural lift. Both the nose and the chin were shifted to the left side of the face. Atrophy was seen at all levels of the face, including the eyes, malar area, lips, and mandible. A scar-like defect was identified in the facial region. The ears were symmetrical and in typical proportions. The skin and hair looked to be normal, with the exception of a "scar" defect and a patch of hyperpigmentation. (Fig.1).

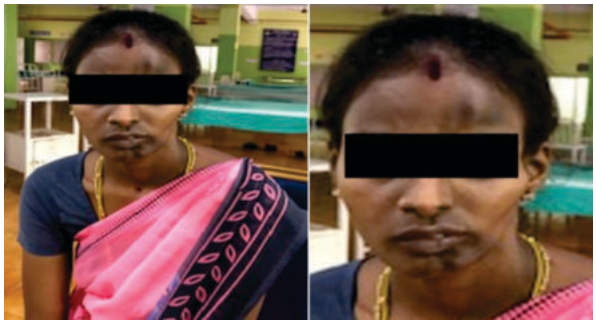

Fig.1 Clinical photograph of the patient showing facial defects

Eye examinations were found to be normal with healthy fundus visual acuity. Pure tone audiometry showed tympanic membrane perforation present on both ears. Audiometry shows bilateral conducting hear loss, more on the right side. Tonsillar hypertrophy present, hypertrophied lingual tonsils. X ray facial bones, CT brain with facial bones and MRI brain show posterior displacement of left globe (12.6 mm) with adjacent fat atrophy and extraocular muscle atrophy noted on

comparison to right side -enophthalmos. (Fig.2)

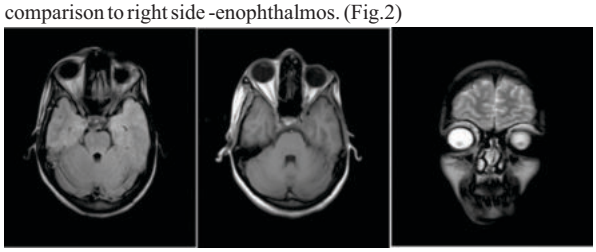

Fig.2 Photograph and CT scan showing skeletal defects

RESULTS AND DISCUSSION

It has been demonstrated that females are more likely to be affected by the sporadic sickness known as Parry-Romberg syndrome but there is no clear indication of a geographical or ethnic predisposition for the condition. The condition often shows its symptoms in the first and second decades of a person's life. It presents itself as an initially faint but developing hemi-atrophy of the face over a period of two decades to twenty years, with a little predilection for the left side of the face [2]. Madasamy et al. (2012) described a similar case with an 8-year-old girl who presented with mostly hard tissue abnormalities and no neurological or intraoral soft tissue changes. In addition to symptomatic therapy of neurological impairments, early multidisciplinary care of such cases must take development, aesthetics, speech, and masticatory function into account [4]. The evolution comes to a sudden halt, stabilises, and then proceeds to a phase that can be described as "burned out" without any obvious explanation. The diagnosis of PRS is made after considering the patient's medical history, performing an examination, and ruling out any potential alternatives. This diagnosis is then backed up by histopathologic and imaging studies [4]. This syndrome produces atrophy not only in the muscular, cartilaginous, osseous, and glandular components, but also in the epidermal and subcutaneous tissues. The illness has the potential to expand from its initial location in the maxillary or periorbital region to the forehead, the perioral region, the teeth, the jaw, and the neck. Kuah et al., 2018 reported a 48-year-old lady with a 3-year history of steadily increasing right facial hemiatrophy and right zygoma became more prominent, and she developed hyperpigmentation on her forearms and left neck. Additionally, her morning headaches and neck stiffness worsened. She was prescribed a course of methotrexate after a clinical diagnosis of PRS was established. As PRS has been associated to smaller teeth with shorter roots, involvement of teeth may assist estimate age of onset in situations where it is unclear when the condition first appeared [5]. Srinivasan et al., 2020 reported progressive hemifacial atrophy in adolescent involving the right side of the face extending from the right cheek below the eye to the angle of the mouth. Computerized Tomography of the brain revealed atrophy of the soft tissues including muscles on the right side of the face [6]. Long et al., 2020 reported Parry–Romberg Syndrome misdiagnosed as multiple sclerosis with recurrent paroxysmal weakness of the right hand, a 3-years history of unilateral tinnitus, and headache for 6 months. It was initially diagnosed but the patient was subsequently diagnosed as having PRS on the basis of clinical manifestations and radiological findings [7].

There is a correlation between the onset of PRS at an early age and its prolonged persistence throughout time. Over the course of time, several explanations have developed to explain this peculiar situation. Our knowledge of the pathophysiology that lies under the surface is still insufficient, and there is no one hypothesis that fully characterizes or predicts PRS. The illness is thought to have either an environmental stressor or a genetic predisposition to develop it, according to one idea. One theory says that the root of the problem is a dysfunction in the sympathetic cervical ganglion. The most powerful one present data suggests an inflammatory autoimmune sickness that may or may not be accompanied by vasculopathy. Immunosuppressive Taking medicine while an illness is still active can enhance clinical results. Enophthalmos caused by a decrease of retrobulbar fat is a frequent condition. the results of radiologic testing. Here we report recent finding and reports on PRS.

CONCLUSION

In conclusion a case of progressive hemifacial atrophy with hyperpigmentation, enophthalmos with atypical features was discussed above. The etiology, pathogenesis of this degenerative condition was still not elucidated. Proper diagnosis and evaluation, multistep facial reconstruction surgery may benefit patient for management of parry romberg syndrome.

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