



Parry Romberg Syndrome - A Pediatric Case Report

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Abstract

Parry-Romberg syndrome, a rare disease, is characterized by atrophy of the skin, fat, muscles and underlying bone and cartilage structures that usually affects the face and neck unilaterally. It is associated with neurological symptoms like epilepsy and involvement of other organs and systems. It has a slow and progressive and its cause remains largely unknown. A female sex predilection has been noted. We report a case of Parry Romberg syndrome in a 9 years old child who presented to us for the evaluation of seizure.

Introduction

Parry-Romberg syndrome, otherwise known as progressive hemifacial atrophy, is characterized by progressive atrophy of the skin and its underlying tissues, primarily affecting the face and neck, usually unilaterally. Caleb Hillier Parry and Moritz Heinrich Romberg were the first to describe the case in 1825 and 1846 [1]. The incidence is about three cases per 100,000 people per year and occurs more commonly among females [1]. The disease manifests between the age of 2 and 20 years with a slow and progressive course [2]. The pathogenesis of this disease is unknown [3]; nevertheless, a variety of theories have been proposed, including immunological, traumatic, infectious, endocrinological, neurological, and genetic causes [1,4]. Epilepsy is the most frequent abnormality associated with Parry-Romberg syndrome.

Case Presentation

A 9-year-old male child presented to the outpatient department with his mother for evaluation of seizures. He was the firstborn child, delivered by vaginal route to non-consanguineous parents. He was apparently well and developmentally normal till the age of 6 years, when he started developing discoloration (hyperpigmentation) on the left side of the face, which became progressive over the year. Parents did not seek medical consultation for the same, and by the age of 8 years, parents also noticed a gradual change in his facial features, noticeably a slow shrinking of the left side. At eight and half years of age, he had a seizure episode of a generalized tonic-clonic type, lasting for about 30 min. At first, he was treated with Ayurvedic medications, but since the seizure episodes recurred mother brought him to our center for further evaluation and management. On examination, the child was moderately built and nourished. We observed an atrophic lesion on the left side of the face, extending from the trichion down to the chin with a depth of 2 mm (Figure 1). Madarosis was noted on the left side. A zone of alopecia was found in the left frontal region (Figure 2). Blood workup, including complete blood count, electrolytes, and calcium, was found to be normal. Magnetic Resonance Imaging (MRI) of the brain showed a well-defined FLAIR and T2W hyperintense oval area in the caudate nucleus and the lentiform nucleus on the left side (Figure 3), which also appeared hyperintense on the T1W images. Electroencephalogram (EEG) showed diffuse changes in the temporal region, suggestive of seizures. He was started on Sodium valproate and has been on regular follow-up thereafter. The dosage required optimization for control of seizure attacks.

Discussion

Parry Romberg syndrome is a rare and sporadic condition reported to be more common in females [5,6]. The onset of the disease is in the first two decades, with a progressive phase lasting for 2 to 20 years. This is followed by a stable phase which usually culminates in resolution [7]. Patients distinctly experience atrophy of the skin and subcutaneous tissues, and as the disease progresses, they could develop atrophy of the underlying muscular, cartilaginous, bony, and

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Received Date: 23 Dec 2021

Accepted Date: 20 Jan 2022

Published Date: 24 Jan 2022

Citation:

Reshad M, Sasikumar L, Avni SKP, Binesh B, Moideen S. Parry Romberg Syndrome - A Pediatric Case Report. *Ann Clin Case Rep.* 2022; 7: 2105.

ISSN: 2474-1655

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Figure 1: Atrophic lesion on the left side of the face, extending from the trichion (red arrow) down to the chin (white arrow). Madarosis can be seen on the left side (black arrow).



Figure 2: The same child with alopecia noted on the left frontal region.

glandular structures [8]. The etiology of Parry-Romberg syndrome is still undetermined; however, the most accepted hypothesis is the autoimmune etiology [9]. Diagnosis of Parry Romberg syndrome is based on patient history and clinical examination and is supported by imaging and histopathological studies [10,11]. This child was normal till six years of age, when the clinical symptoms began to develop. The anatomic changes of Parry-Romberg syndrome impact the growth potential of hard tissue, preventing an increase in size during active growth periods. The dominant clinical feature noted is atrophy of the facial tissues, usually adipose tissue [12]. Seldom, skin, and other connective tissues, including bone, could be affected. The severity of the atrophy can range from barely noticeable asymmetry to severe disfigurement [13]. The present case showed a clear line of demarcation from the forehead down to the chin on the left side. Around 10% to 35% of patients show ocular involvement; the most encountered manifestation is enophthalmos characterized by sunken



Figure 3: Hyperintense oval area in the caudate nucleus and the lentiform nucleus on the left side on T2 MRI brain image.

eyes due to loss of adipose tissue around the orbit [12]. We did not observe any orbital changes in our case. Around 50% of affected individuals show dental changes [9], though our child did not have any findings. While the most frequent neurological manifestation of Parry-Romberg syndrome is epilepsy (60.5%), focal seizures ipsilateral to brain calcifications are also common (50%). Secondary epilepsy, seen in about 30% of the cases, is very difficult to treat. Headache (44%), behavioral disturbances, and trigeminal neuralgia (8.5%) may also be present [1,14,15]. Our child had seizure episodes which were consistent with the EEG findings. He also had behavioral disturbances. MRI showed hyperintensity areas of the white matter, similar to the description in several previous case studies [14,16-19]. Parry-Romberg syndrome has a significant biopsychosocial impact on the patient's life due to functional and aesthetic limitations [11]. There have been proposals to restore facial asymmetry through surgical approaches. Treatment of mild and moderate cases is performed with silicone fillers, collagen, porous polyethylene implants, and autologous fat grafting. The grafting of cartilage, bones, fat, and skincare treatment are reserved for more severe cases [20,21]. The main aim of psychological treatment is the social reintegration of these patients [21].

Conclusion

Parry-Romberg syndrome is an uncommon condition, which manifests as atrophy of one side of the face. In most cases, Parry-Romberg syndrome appears to occur randomly from unknown etiologies. The pathophysiology of the syndrome remains largely unknown. We present a case of Parry-Romberg syndrome with classical facial features and magnetic resonance imaging findings.

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