

A male child with parry romberg syndrome

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Abstract

Parry Romberg Syndrome (PRS) is a rare syndrome of unknown etiology. The most important features of this pathology are the one side facial atrophy, enophthalmy, the angle of the mouth and nose were a deviated to the affected side, and hyperpigmented affected side. PRS is mostly misdiagnosed due to problem in recognizing its clinical features and the rarity of the disease. The diagnosis of PRS was made clinical.

We present a nine-year-old boy, with the complaint of gradually progressive right side facial asymmetry with right thinning of face, the eyeballs were asymmetrical positioned and right side enophthalmus, the angle of mouth and nose were a deviated to the right side, and a big cutaneous linear dark pigmented scar was noticed on the right side of forehead, around the eye, nose, cheek, and chin. To our knowledge, there is no published large-scale clinical trial on the Parry Romberg Syndrome, there are few cases were published in the literatures. This is the first case of Parry Romberg Syndrome described in Iraq.

Keywords: parry romberg syndrome, hyperpigmentation facial patches

Introduction

Parry Romberg Syndrome (PRS) was first reported by Parry (1825) and then described as a syndrome by Romberg (1846). PRS is a rare syndrome of unknown etiology, it has a higher prevalence in age of 5 and 15 years [1].

The most important features of this pathology are the one side facial atrophy, enophthalmy, the angle of the mouth and nose were a deviated to the affected side, and hyperpigmented affected side [1]. PRS is mostly misdiagnosed due to problem in recognizing its clinical features and the rarity of the disease [2]. The frequency of PRS was 1 in 250000-700000 [2].

To our knowledge, there is no published large-scale clinical trial on the Parry Romberg Syndrome, there are few cases were published in the literatures. This is the first case of Parry Romberg Syndrome described in Iraq.

Material and Methods

A nine-year-old boy, reported to the department of pediatric surgery with the complaint of gradually progressive right side facial asymmetry with right thinning of face and hyperpigmentation facial patches. It appeared 6 years prior to presentation, initially as a right hyperpigmentation facial patches. We diagnose him as a case of Parry Romberg Syndrome.



Fig 1: Eyeballs were asymmetrical positioned and right side enophthalmus. The angle of mouth and nose were a deviated to the right side.



Fig 2: A big cutaneous linear dark pigmented scar was noticed on the right side of forehead, around the eye, nose, cheek, and chin.

Discussion

Parry-Romberg syndrome (PRS) is characterized by progressive but self-limiting one side facial atrophy mainly involving the skin, subcutaneous tissue, muscle, and bone. In advanced cases, the affected region can become hyperpigmented [1].

The diagnosis of PRS was made clinical, the main feature is hemi atrophy of the facial tissues. Another important features are neurological manifestations, enophthalmos due to loss of adipose tissue around the orbit, deviation of the nose and mouth to the affected side, unilateral exposure of the teeth (if the lips are involved) and unilateral atrophy of the tongue [2]. Mostly it affects the females, on the left side of the face [1, 3]. Depending on the degree of involvement of the skin, nerve, bone, and subcutaneous tissue involvement PRS can be classified as mild, moderate, and severe [3].

The main etiology of this syndrome was unclear, many theories have been included to explain it like autoimmunity, viral infection, trauma, and Possible hereditary etiology is also proposed in few cases [4, 5].

Our child showed clearly the classical clinical manifestations of the Parry-Romberg syndrome. He presented with progressive right side facial hypoplasia with facial cutaneous hyperpigmentation. Physical examination

revealed the right eye was depressed in the socket. The angle of mouth and nose were deviated to the right side. It appeared 6 years prior to presentation. There is no history of trauma to this site. The patient's medical and family history was noncontributory. He had no other underlying illness, no evidence of facial nerve palsy, and normal mastication.

Conclusion

Parry Romberg is a rare syndrome with the diagnosis of it is mainly clinical.

Ethical approval and Consent

We obtained written informed consent from the patients' parents. The Medical Ethics Committee of the University of Anbar, Anbar, Iraq approved this study.

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