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A Case Of Parry Romberg Syndrome In An Adolescent-Progressive Hemifacial Atrophy

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Abstract

Parry Romberg Syndrome is a rare degenerative disorder characterized by slowly progressive deterioration (atrophy) of the skin and soft tissues of half of the face (hemifacial atrophy). It is more common in females than in males. Over time the soft tissue (muscle and fat) gradually shrinks, the facial bones may change and the skin may become thin. Some patients present with headaches and seizures infrequently. An autoimmune mechanism is suspected and the signs of this syndrome overlap with a condition known as linear scleroderma which is always misdiagnosed as the latter. The cause of Parry Romberg syndrome is unknown. This affects mostly one side of the face. Sometimes it may also affect both sides of the face. It appears between the age group of 5 to 15 years. This present article reports a case of a 15-year-old girl who presented with wasting over the right side of the face and the right thigh region without any neurological changes.

Keywords: Parry Romberg syndrome, atrophy, hemifacial atrophy

Introduction

Parry Romberg Syndrome is a rare disorder whose etiology is not known. It was first coined by Caleb Parry in 1825 and Moritz Romberg in 1846, the term progressive hemifacial atrophy was coined by Eulenberg in 1871(1). This obscure disorder occurs mainly in females and is characterized by a disappearance of fat in the dermal and subcutaneous tissues on one or both sides of the face giving the appearance of facial paresis(2). It usually begins in adolescence or early adulthood and is slowly progressive. In its advanced form the affected side of the face is gaunt and the skin is thin, wrinkled, and rather dark, the hair may turn white and fall out and the sebaceous glands become atrophic, the muscles and bones are not involved as a rule. The condition is a form of lipodystrophy, but the localization within a myotome suggests the operation of some neural factor (possibly a growth factor) of unknown nature. A variegated coloration of the iris and a congenital oculosympathetic paralysis are found in some cases(3). Rarely, certain central nervous system abnormalities referable to the ipsilateral hemisphere (mainly focal seizures, migraine, trigeminal neuralgia and ventricular dilatation), are conjoined(Hosten). The significance of these associations is unclear. Immunosuppressive treatment can stabilize the clinical course.

Case Report:

A 15-Year-old female patient who is a student came with complaints of progressive wasting over the right side of the face for 2 years along with decreased sweating on the right side of the face for 6 months and pain over the right lower limb for 1 year. She was taken to a nearby clinic by a general physician for consultation 6 months back and was advised certain investigations about which the details are not

available and was referred to a neurologist but the patient did not follow up on the advice. But for the past 6 months, she also started noticing sweating on the right side of the face, and pain in the right thigh region had increased gradually so she came to our hospital for further management. The patient also gives a history of swelling over the right cheek for the past 2 years which was firm, painful, and tender and associated with ear pain. There is no history of local redness. She also says that there is waste in the right thigh region associated with pain, wasting of labia minora in the right side perianal region, feeling of loss of contour in the right buttocks region for 1 year, and decreased lacrimation in the right eye since 6 months. There was no history suggestive of any neurological disturbances and vision and hearing were normal. The patient did not suffer from any other systemic illness. History from the mother revealed no similar illness in the family and no history of trauma or any other infection was known to precede the onset of the above symptoms.

Fig 1. Wasting On The Right Side Of The Face Compared To The Left Side Is Normal

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Fig 2. The Left Side Of The Face Is Normal Compared To The Right Side

On Examination patient is conscious and oriented, vitals are stable, and there is facial asymmetry with wasting of muscles involving the right side of the face extending from the right cheek below the eye to the angle of the mouth. There was no discoloration of the overlying skin with any hypo or hyperpigmented patches. Examination of the oral cavity was normal. There was no atrophy or wasting of the tongue. The right zygomatic and the maxillary region appeared more hypoplastic than the left. There is wasting involving the right thigh region, wasting of labia minora in the right side perianal region along with wasting in the right side of the buttocks. Neurological examinations including cranial nerves were all normal. Other systemic examinations were also normal. A clinical diagnosis of Progressive hemifacial atrophy was made before investigations. Routine blood investigations were done and were found to be normal. Genetic testing was done and revealed no chromosomal abnormalities. A Nerve conduction study was done and was found to be normal. Antibody testing like Anti-nuclear antibodies, Anti centromere antibodies, Anti topoisomerase antibodies were done to rule out connective tissue disorders like Scleroderma and were found to be negative. CT scan of the face showed atrophy of the zygomatic bone, the maxillary bone, and its sinus on the right side. CT scan of the brain revealed no significant abnormality. MRI brain was suggested but was not done. Based on the clinical and radiological findings a diagnosis of Parry Romberg Syndrome was made. The patient was suggested to undergo Augmentation surgery. An opinion was obtained from a Plastic surgeon and was advised the same. The patient was then transferred to the surgical ward to undergo the surgery.

Discussion:

Parry Romberg Syndrome is a rare progressive craniofacial disorder also known as progressive facial hemiatrophy which involves atrophy of the skin and soft tissues of half of the face. It is mostly unilateral. Its etiology is not well understood but the involvement of autoimmunity has been suggested(4). It is a disease with a strong female predilection. The etiology of hemifacial atrophy can be subjected to various theories which include hereditary, viral infection like herpes, bacterial infection like Lyme disease, trauma to the face and neck, endocrine disturbances, autoimmunity, sympathetic malfunctions, trigeminal neuritis, and association with a connective tissue disorder particularly scleroderma(5). Parry Romberg Syndrome may be referred to by other names as well such as Progressive hemifacial atrophy, Progressive facial hemiatrophy, Idiopathic hemifacial atrophy, and Romberg's syndrome. In some circumstances, the disease may progress to both sides of the face. It may

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even affect an arm, the midsection, or a leg. Most often Parry Romberg Syndrome is linked to a form of the autoimmune disease scleroderma called linear scleroderma or "en coupe de saber"(ECDS)(6). In linear scleroderma, a localized area of skin and tissues beneath it contain abnormalities similar to those found in Parry Romberg Syndrome(7)Signs and Symptoms may include initial facial changes usually involving the area of the face covered by the temporal or buccinator muscles. The disease progresses and spreads from the initial location resulting in atrophy of the skin and its adnexa as well as the underlying subcutaneous structures such as connective tissue i.e, fat, fascia, cartilage, bones, and also muscles of one side of the face. (8) The mouth and nose may at times be deviated towards the affected side of the face(9). In a few cases, the hair and skin overlying affected areas may become hyperpigmented or hypopigmented with patches of unpigmented skin(7). Neurological abnormalities are common and may include trigeminal neuralgia, migraine headaches that may be accompanied by visual abnormalities, nausea and vomiting, and seizures which are typically Jacksonian and occur contralateral to the affected side of the face(10). Ocular abnormalities may include features suggestive of Horner's syndrome, ophthalmoplegia and other types of strabismus, uveitis, and heterochromia of the iris(11). The tissues of the mouth including the tongue, gingiva, teeth, and soft palate are commonly involved in Parry Romberg Syndrome. Dental abnormalities such as delayed eruption, dental root exposure, and resorption of the dental roots on the affected side(12). Few may have temporomandibular joint disorder and spasm of muscles of mastication on the affected side. Differential diagnoses may include rare syndromes like Goldenhar, which manifests at birth and may be non-progressive, and Barraquer Simons syndrome which resembles PRS and involves both sides of the face. (2)Diagnosis is mainly by history and physical examination who present with facial asymmetry(13). MRI brain may be done for those who present with neurological symptoms(14). Autoantibody testing may be done to rule out its association with other connective tissue disorders. Radiological features may give information on the severity of atrophy and may be useful when surgical procedures are considered. They include maxillary and mandibular hypoplasia and atrophy of facial muscles. (15)Treatment options may be both medical and surgical(16). The initial goal is to slow down the disease progression. Medical management may include immunosuppressive drugs such as methotrexate, corticosteroids, cyclophosphamide, and azathioprine(17). Surgical treatment involves the autologous fat transfer or fat grafts to restore a more normal contour to the face(18). However greater volume defects may require microsurgical reconstructive surgery which may involve the transfer of an island parascapular fasciocutaneous flap or a free flap from the groin, rectus abdominis muscle (Transverse Rectus Abdominis Myocutaneous or "TRAM" flap) or Latissimus dorsi muscle to face. Severe deformities may require additional procedures such as pedicles temporal fascia flaps, cartilage grafts, bone grafts, orthognathic surgery, and bone distraction(19). The timing of surgical intervention is important. It is always better to perform the surgery when the disease is inactive but few surgeons prefer early intervention. (20)

Conclusion:

Parry Romberg Syndrome is a disease known to cause disfigurement of the face and other muscles of the body. It is known to have a major cosmetic concern and surgical treatment modality can be of great benefit as it would cause major social embarrassment and psychosocial issues. This disease mainly presents in the first two decades of life. The present case has been symptomatic for the past 2 years but the diagnosis was made at the age of 15 years. Surgical treatment would be the best option due to social concerns. If diagnosed earlier, the facial disfigurement could have been prevented with early intervention. More research is necessary to find much more treatment options for this incurable disease.

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