Original Research Paper

Volume - 13 | Issue - 01 | January - 2023 | PRINT ISSN No. 2249 - 555X | DOI : 10.36106/ijar

Radiology

PARRY ROMBERG SYNDROME - A RARE CASE REPORT

Dr. Kavan Parikh* B.J MEDICAL COLLEGE (3rd-year Resident)*Corresponding Author

Dr. Vasant Rathod B.J Medical College Assistant Professor

Dr. Ankit Agarwal B.J Medical College 1st year Resident

ABSTRACT The purpose of this report is to present a rare case of Parrv-Romberg syndrome (PRS), also called "progressive hemifacial atrophy". This rare degenerative condition is characterized by atrophic changes affecting one side of the face. The etiology of these changes remains idiopathic. Definitive diagnosis of Parry-Romberg syndrome is based on thorough clinical and radiological examination. Treatment is usually conservative while others include using alloplastic implants to improve facial disfigurement was suggested to the patient.

KEYWORDS : Parry-Romberg syndrome; Romberg's disease; Romberg hemifacial atrophy

INTRODUCTION

The PRS was first introduced & reported by Parry in 1825 but named as a syndrome by Romberg in 1846, is a rare slowly progressive, and self-limiting disease characterized by atrophy of the skin, subcutaneous tissue, cartilage, bone & muscles on one side of the face. This syndrome is insidious in onset, more common in girls with age of presentation predominantly in the first to the second decade of life, and progresses rapidly following 2-10 years of onset followed by stabilization. Etiology remains idiopathic. Still, different etiologic factors have been proposed similar as viral infections, genetics, trauma, supplemental and trigeminal neuritis, localized scleroderma and, endocrine diseases, although, none have been substantiated. Some authors believe this syndrome is an overlap of linear scleroderma “en coup de saber.” In some cases, its signs can be detected at birth, in others from 5 years, and in a few cases after 15 years of age. It can involve the ocular structures & most commonly manifests as enophthalmos. The earlier the onset the more severe the disfigurement, because of the high rate of growth at these periods. In 50 percent of the patients, neurologic involvement can be seen such as neuropathic pain, epilepsy, and intracerebral dysplasia.

In this report, we have reported a case of Parry-Romberg syndrome, with no familial history of any syndromes. We have discussed clinical presentation, radiological approach & role of cross-sectional & radiological pointers for diagnosing this rare entity which helps the clinician for further management & improving the quality of life for affected patients.

Written informed consent was taken from the case to use her medical records and photos for this report. CARE criteria compatibility was considered & matched.

CASE STUDY:

A 22 years old young Indian female presented to our Department of Radiology with complaints of facial asymmetry & deformity involving the left side of her face & difficulty opening her mouth & chewing since she was in standard 6th. The patient had no familial history of similar complaints. Also, the patient had no learning difficulties. She was ordered an MRI of the Temporo-Mandibular joint for further evaluation of suspected internal disc derangement by the dental department.

Figure 1: Showing the facial asymmetry, left malar hypoplasia & restricted mouth opening with facial deformity.

Sources: Department of Radiology, B.J Medical College, Ahmedabad

IMAGING FINDINGS:

1.5 tesla Philips MRI machine was used & contrast Magnetic resonance (MR) imaging was performed using . The left parotid gland appeared atrophic & showed altered signal intensity -T2 hypointense signal as compared to the right side. Atrophy of the left masseter muscle, subcutaneous fat & retro-antral fat was noted as compared to the right side. The left mandibular condyle appeared altered in shape with resultant anteroposterior flattening. The open-mouth study noted reduced anterior translation of the mandibular condyle on both sides. In the closed-mouth study, the Intra-articular disc appeared normal in position & morphology in the bilateral TM joint.

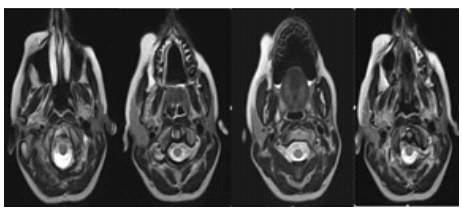

Figure 2: Atrophy of left-sided facial subcutaneous fat with associated atrophy of left parotid gland & left masseter muscle

Source: Department of Radiology, B.J Medical College, Ahmedabad

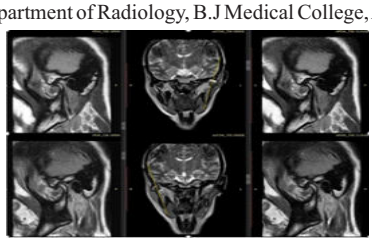

Figure 3: Mild hypoplasia of left mandibular condyle with reduced anterior translation (on open mouth view) on both sides.

Source: Department of Radiology, B.J Medical College, Ahmedabad

DISCUSSION:

Parry Romberg syndrome can lead to several progressive congenital and developmental deformities. It results in atrophy of facial muscles, bones and skin but predominantly it involves muscles restricted to unilateral side as is seen in our case. Also, the skin of the affected side may become dry and can have hyperpigmentation such as port wine stains. This syndrome can be classified according to its severity: - Mild Parry Romberg syndrome: Atrophy of soft tissues at the trigeminal area of lower face without rotation of the occlusal plane.

Moderate Parry Romberg syndrome: Involvement of nostrils and lip commissures in addition to the trigeminal dermatome, no atrophy of skeleton and relatively horizontal occlusal plane.

Severe Parry Romberg syndrome: Rotation and transverse cant of occlusal plane, atrophy of maxilla, mandible and zygoma, deviation of chin and nose to the affected side.

To summarize, PRS is a rare, self-limiting syndrome with slow progressive hemifacial atrophy. Its exact etiology and pathophysiology are still doubtful. It may cause aesthetic, functional and psychological issues, which require a well-planned, specific and multidisciplinary treatment. This paper, discussed a classic case of PRS . A noninvasive approach was taken for the clinical suspicion & diagnosis with Magnetic resonance & Computed tomography correlation. Imaging techniques were quite helpful for early diagnosis & appropriate guidance of both the patient & clinicians to plan further management & outcome with high satisfaction of the patient. So, this report is a satisfactory sample for aiding diagnosis for this rare entity (PRS) through thorough clinical suspicion combined with radiological aspects.

The pathophysiology of the syndrome remains unknown. There is no definitive treatment for this condition but an attempt to improve cosmetic appearance of facial structures by using restorative plastic & facial surgery which includes fat or silicone implants, flap/pedicle grafts, or bone implants can be done to improve facial disfigurement.

CONCLUSIONS:

A high index of clinical suspicion, dedicated history & thorough clinical examination is the key to aiding the differential diagnosis of PRS. Radiological & other supportive investigations can however help in early diagnosis & better management of the patient. Patients of PRS should be screened for other presentations such as enophthalmos, epilepsies, cranial nerve palsies & other complications related to ocular & neurological involvement.

REFERENCES:

1. Parry CH. Collections from the Unpublished Medical Writings of the Late Caleb Hillier Parry. Med Chir Rev. 1825;
2. 124-147. PubMed: https://www.ncbi.nlm.nih.gov/pubmed/29917749 2. Moore T, Southard KA, Casko JS, Qian F, Southard TE. Buccal corridors and smile esthetics. American Journal of Orthodontics and Dentofacial Orthopedics. 2005; 127: 208-213. PubMed: https://www.ncbi.nlm.nih.gov/pubmed/15750540
3. Al-Khenaizan S, Al-Watban L. Parry-Romberg syndrome: Overlap with linear morphea. Saudi medical journal. 2005; 26: 317-319. PubMed: https://www.ncbi.nlm.nih.gov/pubmed/15770315
4. Hickman JW, Sheils WS. Progressive facial hemiatrophy: report of a case with marked homolateral involvement. Arch Intern Med. 1964; 113: 716-720. PubMed: https://www.ncbi.nlm.nih.gov/pubmed/14120599
5. Tolkachjov SN, Patel NG, Tollefson MM. Progressive hemifacial atrophy: A review. Orphanet J Rare Dis. 2015; 10: 39. PubMed: https://www.ncbi.nlm.nih.gov/pubmed/25881068
6. Wolf SM. Verity, Neurological complications of progressive facial hemiatrophy. Journal of Neurology, Neurosurgery & Psychiatry. 1974: 37: 997-1004.
7. Van der Cruyssen F, Van der Cruyssen F, Schoenaersb MJ, Politis C. Parry Romberg syndrome: A long-term retrospective cohort study of 10 patients. Oral and Maxillofacial Surgery Cases. 2018; 4: 73-83.