# **C**ASE **R**EPORT

# Parry Romberg Syndrome: A Rare Case Report

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#### ABSTRACT

**Introduction:** Parry-Romberg syndrome is a rare and self-limiting disease that causes gradual progressive hemifacial atrophy. This condition may manifest itself in a range of prenatal and developmental problems that progressively deteriorate over time.

**Case report:** We present a 54-year-old man who was involved in a road traffic accident and was incidentally diagnosed with Parry Romberg syndrome after undergoing a head CT scan. The accompanying clinical and imaging findings were highlighted, with a focus on the disease's radiographic features.

**Conclusion:** Parry Romberg syndrome is a rare disorder in men, and radiological imaging can aid in the exclusion of other possible causes and the monitoring of disease development.

Keywords: Parry-Romberg Syndrome, Hemifacial Atrophy, Head CT, Radiological Imaging, Rare Disorder in Men

## INTRODUCTION

Parry Romberg syndrome (PRS) is a rare, insidious onset disease that leads to facial shrinkage, commonly known as progressive facial hemi-atrophy. In 1825 Parry introduced it first, and in 1846 Romberg called it a syndrome.<sup>1-4</sup> It mainly includes the skin, subcutaneous binding tissues and may subsequently advance involving the underlying muscles, cartilage and osseous structures.<sup>5-8</sup> Bilaterally occurring atrophy was noted in 5-10% of cases.<sup>9</sup> However in more than 85% of the instances, the left side of the face was involved.

Although none were substantiated, many etiological variables such as viral infections, genetics, trauma, peripheral and trigeminal neuritis, localised scleroderma, and endocrine problem were hypothesised.<sup>10-12</sup> The diagnosis is based on clinical symptoms. Small injuries can be observed in clinical examinations above the eyebrows and the frontal paramedian flap, which grow progressively atrophic with disease progression and separate healthy and atrophic tissue borders, stated en coup de sabre.<sup>13-15</sup> Deforming face asymmetry is also seen, which affects the oral functions by the implication of chewing muscles.<sup>16,17</sup> Sometimes vitiligo, hyperpigmentation and/or port stains occur at the local level. Also ears can be deformed or smaller than normal.<sup>18,19</sup>

The bottom portion of the face, including the mandible and chin may shift to the affected side. Lip deviation can result in an increase in tooth show on the affected side compared to the opposing side.<sup>20</sup> Intraoral binders can result in hemiatrophy of the affected side's lips, tongue, and soft palate. Additionally, malformations and congenital tooth loss can be observed, as well as delayed eruption, root underdevelopment, or root resorption.<sup>21-23</sup> On the affected side, the alveolar ridge, mandible body, ramus, and gonial angle may be smaller and undeveloped, resulting in a deviation of dental and skeletal midlines.<sup>24,25</sup> This syndrome can have psychological consequences, impairing the patient's daily functioning and impairing his or her looks. Although changes in cutaneous PRS may appear to be mild on imaging investigations, 20% of these patients will develop cerebral manifestations that may be unrelated to the level of soft-tissue involvement or neurologic symptoms.<sup>6,26,27</sup> Radiologic studies can help uncover clinically asymptomatic cerebral involvement, rule out other possible differential diagnoses, and track disease progression.<sup>15</sup>

## **CASE REPORT**

A 52-year-old male met with a road accident while driving a two-wheeler. He was bought to our hospital casualty and further sent to radiology department for a CT brain scan and the same was done. The physical examination revealed multiple lacerations over the left side eyebrow, infraorbital and zygomaticomaxillary region. The routine clinical laboratory parameters were done and no remarkable findings were observed. CT brain concluded atrophy of the right temporalis (Figure 2), bilateral right pterygoids (Figure 3) and right masseter muscle (Figure 1). MRI was also taken after analysing CT findings and the former revealed fatty

C53



**Figure-1:** Atrophy of right masseter muscle is seen in both images (blue arrows)



Figure-2: Atrophy of right temporalis muscle (yellow arrows)



**Figure-3:** Atrophy of right lateral pterygoid (red arrow) and right medial pterygoid muscle (green arrow).

atrophy of right temporalis (Figure 4b), bilateral right pterygoids (Figure 5) and right master muscle (Figure 4a), consistent with CT findings. The interesting observation of this case was that the patient came to our department due to a road accident and PRS was incidentally diagnosed. The patient was given appropriate treatment for his pain.

# DISCUSSION

Parry Romberg syndrome is a sporadic illness that has been shown to be more prevalent in females<sup>6,29,30</sup> without any obvious regional or ethnic predisposition.<sup>29</sup> The disease often manifests in early and second decades of life, as an initially subtle but increasing hemi-atrophy of the face over a period of 2–20 years, with a little preference for the left side.<sup>31</sup> Without apparent cause, the evolution abruptly halts and stabilises, eventually reaching a "burned out" phase.<sup>30</sup> The clinical history, examination, and elimination of alternative



Figure-4: T2w axial image shows fatty atrophy of right masseter muscle (blue arrow) and right temporalis muscle (yellow arrow).



Figure-5: T2w axial image shows fatty atrophy of right medial pterygoid (green arrow) and mild fatty atrophy of right lateral pterygoid muscle (red arrow).



**Figure-6:** Dimpling of the right side of the face (zygomatic region) and laceration is noted at left zygomaticomaxillary region due to RTA.

options are used to make the diagnosis of PRS, which is supported by histopathologic and imaging investigations.<sup>32</sup> The condition causes atrophy of the epidermal and subcutaneous tissues, as well as muscular, cartilaginous, osseous, and glandular components.<sup>6</sup> Beginning in the maxillary or periorbital region, the disease can spread to the forehead, perioral region, teeth, jaw, and neck.<sup>30</sup> Teeth involvement may help determine age of onset in uncertain cases, as PRS has been linked to smaller teeth with short roots.<sup>33</sup> Early start and lengthy persistence of PRS have been linked to illness severity.<sup>30</sup>

Many hypotheses have evolved over time to explain this odd condition. Our understanding of the underlying pathophysiology is still inadequate, and no single

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Parry Romberg Syndrome

explanation adequately characterises and predicts PRS.<sup>3</sup> One theory proposes that the condition is caused by trauma or hereditary susceptibility. Another suggests that it is caused by sympathetic cervical ganglion malfunction.<sup>30</sup> The strongest current data supports an inflammatory autoimmune illness with or without vasculopathy.<sup>30</sup> Immunosuppressive medication during active illness improves clinical outcomes.<sup>34</sup> Enophthalmos due to retrobulbar fat loss is a common radiologic finding.<sup>30</sup>

### CONCLUSION

Parry Romberg Syndrome is an uncommon, self-limiting and gradually progressing facial hemi-atrophy that generally affects the skin and subcutaneous tissues but may also affect deeper tissues such as muscle, cartilage, and osseous structures. Although neurologic and ophthalmologic symptoms are frequently encountered, the underlying aetiology remains unknown. Along with determining the amount of disease, radiologic examinations may aid in the exclusion of other possible diagnoses, aid in disease progression monitoring, and evaluate post-treatment responses.

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