Orofacial and ophthalmological Manifestations of Long-Standing Parry-Romberg Syndrome

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Background

Parry-Romberg Syndrome (PRS) is a rare acquired disorder characterized by progressive hemiatrophy of soft and sometimes hard facial tissues typically starting before 20. The extent of facial deformity is a function of age of onset and duration of the condition. Although aetiology is unknown, some experts consider it a variant of localized scleroderma and limited evidence for regression of disease activity with immunomodulator therapy support a possible autoimmune or inflammatory nature.

Diagnosis is primarily clinical and PRS has highly variable course and severity including serious neurological, ophthalmological and orofacial sequela. We report a young male with long-standing PRS and multitude of complications.

Case

A 24-year-old male presented for evaluation of facial asymmetry starting with appearance of a shiny skin patch under his right eye during adolescence. He was subsequently diagnosed with morphea and treated with topical steroids to no avail. Considering his condition merely a skin disease, he had not sought additional care ever since. He denied history of trauma, burn, extracranial anomalies, or neurological problems. PMH was non-contributory and he was not on any medications.

Extraoral exam revealed sunken-in right face with a focal shiny plaque below the orbit and only a thin skin covering the depressed temporal, zygomatic and mandibular region. Other notable findings were right-sided ear protrusion, enophthalmos, lower lid ectropion, sparse facial hair, limited mouth opening and upward deviation of lip corner with ipsilateral exposure of teeth.

Intraorally, right tongue was atrophic, depapillated, had poor muscle tone and deviated ipsilaterally. Proprioception of dental instruments was also diminished on the affected side of his mouth. His maxillary midline was shifted causing Class III occlusion with posterior crossbite and open bite on the right side. Panoramic radiograph revealed right-sided dental crowding, delayed eruption and shorter roots as well as ipsilateral reduction in ramus height, body size, gonial prominence and mental tubercle.

Differential also included facial lipoatrophy and hemifacial microsomia but history, clinical, radiographic and laboratory (ANA:1:80, nucleolar pattern associated with systemic sclerosis) findings supported diagnosis of Parry-Romberg Syndrome with complications and he was referred to specialists for management.

Figure 1: Clinical presentation of patient illustrating a focal shiny plaque below the right orbit together with enophthamos, sunken-in appearance, upward deviation of lip commissure and ear protrusion on the right side.

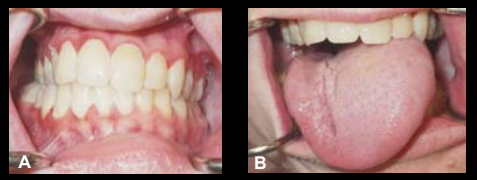

Figure 2 : Clinical presentation of D) atrophic, depapillated right tongue; E) right-sided shift of maxillary mid line & posterior cross bite.

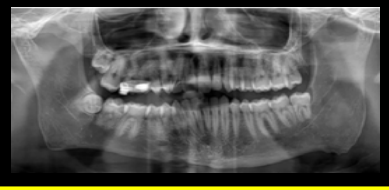

Figure 3: Panoramic radiograph illustrating right-sided dental crowding, shorter roots, ipsilateral reduction in ramus height, body size, gonial prominence and mental tubercle.

Discussion

Reported Clinical & Radiological features of Perry Romberg Syndrome
• Facial Asymmetry
• Coup de sabre
• Alopecia, sparse facial hair, thinning of eyebrows
• Enophthalmus; +/- ocular problems
• Deviation of facial midlines (ipsilateral deviation of mouth & nose)
• Unilateral lip atrophy, unilateral tooth exposure
• Ipsilateral tongue atrophy and loss of lingual papilla
• Shortening of mandibular body and/or ramus
• Short roots & delayed eruption on the affected side
• +/- ipsilateral body atrophy
• +/- neurological problems (seizures, TN,..)

Our patient had many of the features reported for PRS in the literature.

Etiology of PRS is unclear but it may be related to localized scleroderma.

Esthetic, functional and psychological challenges of PRS mandates multidisciplinary collaboration for management. This includes immunosuppressive therapy to arrest active disease, symptomatic treatment of neurological sequela and cosmetic interventions when the condition is stabilized.

Conclusion

Clinicians should include PRS in the differential diagnosis of patients with facial asymmetry. Timely diagnosis, patient education and early multidisciplinary interventions are necessary to halt progression of active disease, ameliorate symptoms and prevent serious sequela.