

Tongue Involvement in Parry-Romberg Syndrome

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Parry-Romberg syndrome (PRS) is an uncommon disorder characterized by progressive hemiatrophy of the skin and soft tissue of the face. It can also involve atrophy of deeper structures, such as muscles, cartilage, and bones¹. Intraoral involvement may occur with hemiatrophy of the mandible, teeth, and tongue². We describe striking mucosal involvement of the tongue in 2 girls with PRS. Cases were from the British Columbia Children's Hospital.

A 4-year-old female presented with hyperpigmentation over right chin for 6 months. Examination revealed hyperpigmented plaques above the right ear, behind the right ear extending to the jaw, and over the mental prominence. The right ear lobe and helix were atrophic. Oral examination revealed rightward deviation of the tongue and hemiatrophy with loss of papillae (Figure 1, left panel). Tests for anti-nuclear antibodies, anti-dsDNA, and *Borrelia* were negative. Treatment with oral prednisone 5 mg twice daily and 15 mg weekly oral methotrexate (MTX) resulted in no progression of the lesions over 2 years.

A 15-year-old female presented with hypopigmentation and induration of the right paranasal skin. Physical examination revealed an indurated plaque along the right paranasal skin with atrophic plaques extending from the right cheek to the chin and neck. Oral examination revealed hemiatrophy and loss of dermal papillae over the right side of the tongue (Figure 1, right panel). Treatment with 15 mg twice daily oral prednisone and 20 mg weekly oral MTX was initiated with softening and partial resolution of the facial and neck lesions. The oral lesions showed minimal change over the 4-year followup.

REFERENCES

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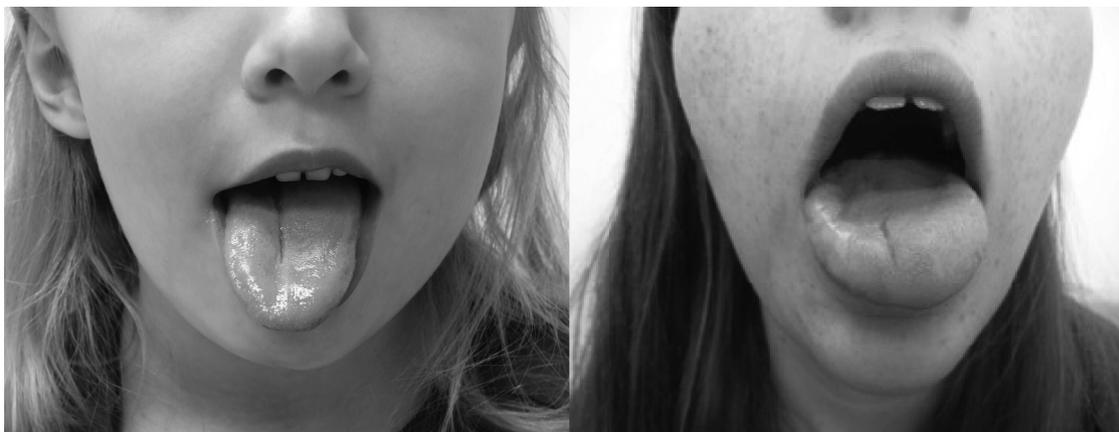


Figure 1. Left panel: hemiatrophy of the right side of the tongue in a 4-year-old girl. Right panel: hemiatrophy and loss of papillae of the right side of the tongue in a 15-year-old female.