# Raynaud Phenomenon of the Tongue in Juvenile Systemic Lupus Erythematosus

Diana Rita Oliveira<sup>1,2</sup>, Graça Barros Loureiro<sup>2,3</sup>, Mariana Rodrigues<sup>2</sup>, Iva Brito<sup>2</sup>

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A 12-year-old girl was referred to pediatric rheumatology for heliosensitive butterfly-malar rash. She complained of fatigue, mild alopecia, and the appearance of large and painful oral ulcers. No weight loss, recurrent fever, dysphagia, arthralgia, morning stiffness, ocular symptoms, or extremity changes. Laboratory testing showed Coombs positive hemolytic anemia, mild lymphopenia, without thrombocytopenia, and an erythrocyte sedimentation rate of 36 mm/h. Complement C3 and C4 were within the normal ranges. Antibody panel showed positive antinuclear antibodies (ANA) (1/320 speckled) and anti-Sjögren-syndrome-related antigen A (SSa), with negative double stranded deoxyribonucleic acid (dsDNA) and antiphospholipid antibodies. Urinary sediment and urinary protein/creatinine ratio were normal. Juvenile systemic lupus erythematosus (SLE) was diagnosed, and she started treatment with lowdose prednisolone and hydroxychloroquine.

Three years after the diagnosis, she started complaining of an occasional tingling sensation on the tongue, without obvious precipitating factors, which was never present during the observation period. This was without recurrent mumps or other sicca symptoms. The patient then documented her complaints with a picture that showed white discoloration of the tongue (Fig. 1), associated with the tingling sensation, which minutes later had returned to its normal red appearance. A diagnosis of Raynaud phenomenon of the tongue was made. In the following year, the patient started to exhibit mild Raynaud phenomenon in her hands.

Raynaud phenomenon is a recurrent vasospasm triggered by cold conditions or emotional stress, causing pallor, cyanosis, and reactive hyperemia of extremities. It can appear in up to 10%-45% of systemic lupus erythematosus. It is manifested as dramatic color changes of fingers and toes and, less frequently, the tip of the nose, nipples, or the earlobes. Raynaud phenomenon of the tongue is rare given the existence of

collateral circulation, but has been previously reported in association with radiotherapy, scleroderma, and other autoimmune conditions. This case highlights the need to thoroughly assess any patient with connective tissue disease who presents with unexplained oral symptoms.

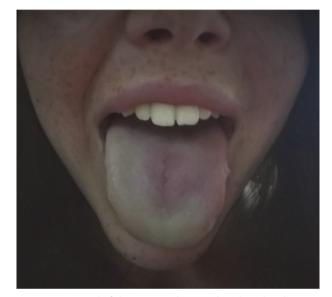


Figure 1. Raynaud of the tongue, showing the white discoloration of the tongue.

**Keywords:** Adolescent; Lupus Erythematosus, Systemic; Raynaud Disease; Tongue Diseases

#### WHAT THIS REPORT ADDS

• Patients with systemic lupus erythematosus may experience Raynaud phenomenon, usually affecting the fingers and toes.

• Raynaud phenomenon of the tongue is a rare symptom in patients with connective tissue disease and should be investigated carefully.

• The diagnosis of Raynaud of the tongue can be difficult given the short period of symptoms and the normalization of the tongue's appearance between episodes.

#### **Conflicts of Interest**

The authors declare that there were no conflicts of interest in conducting this work.

**Corresponding Author** 

- https://orcid.org/0000-0002-3186-6638
- dianarfoliveira@gmail.com

<sup>1.</sup> Pediatrics Department, Hospital de Braga, Braga, Portugal

<sup>2.</sup> Pediatric Rheumatology Unit, Centro Hospitalar e Universitário São João, Porto, Portugal

<sup>3.</sup> Pediatrics Department, Centro Hospitalar Entre-Douro e Vouga, Santa Maria da Feira, Portugal

Diana Rita Oliveira

Serviço de Pediatria, Hospital de Braga, Sete Fontes - S. Vitor, 4710-243 Braga, Portugal

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#### **Consent for publication**

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## **Confidentiality of data**

The authors declare that they have followed the protocols of their work centre on the publication of patient data.

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