Scalloped Tongue in Primary Amyloidosis

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A 53-year-old man presented with fatigue, weight loss, dyspnea on exertion, and frothy urine. He also reported recently diagnosed hypertension, painful lower extremity edema, abdominal distension, and anasarca. Physical examination revealed a slightly enlarged tongue with scalloping (Fig. 1). Labs revealed heavy proteinuria, alkaline phosphatase of 1600 U/L, and SPEP with abnormal IgM lambda and kappa light chains. Given clinical suspicion for an infiltrative process, an abdominal MRI was ordered, which showed hepatomegaly of 32cm and ascites (Fig. 2). A transjugular liver biopsy was performed for definitive diagnosis, which showed strong staining with Congo red and associated apple-green birefringence.

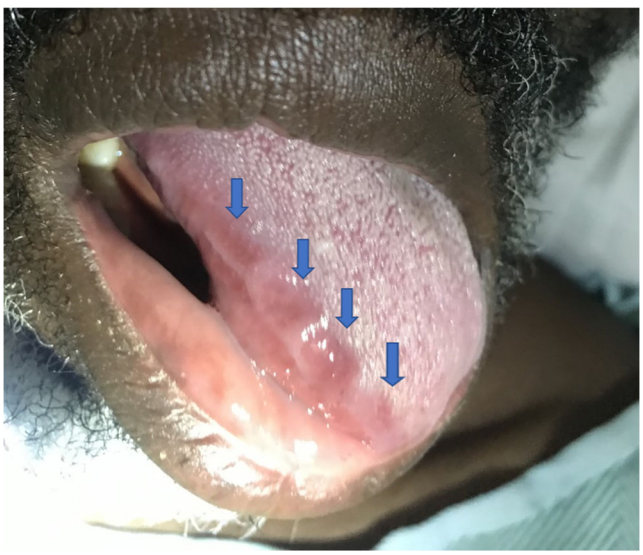

Figure 1 Tongue scalloping due to deposition of amyloid in the tongue, subsequent enlargement, and resulting pressure of the tongue against the teeth (arrows).

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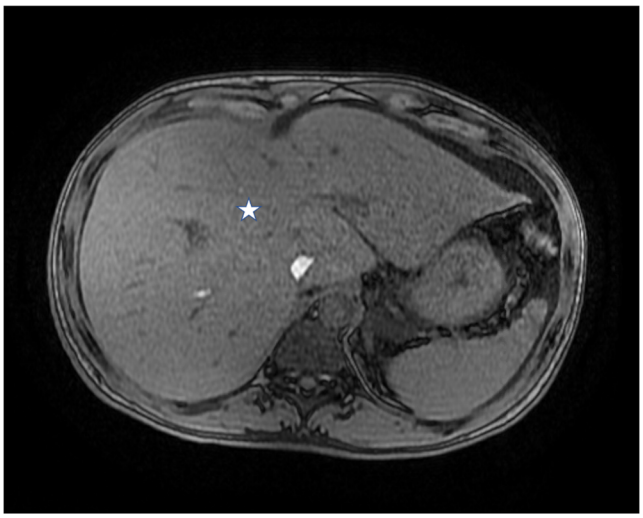

Figure 2 Massive hepatomegaly (star).

Primary AL amyloidosis is a systemic disease characterized by deposition of protein fibrils composed of monoclonal light chains in tissue. Identification of an enlarged, scalloped tongue during physical examination raised clinical suspicion for amyloidosis. Macroglossia with scalloping is a manifestation of light chain deposition found in 15% of AL amyloidosis cases and can be present even without evidence of systemic disease[1,2]. Diagnosis is confirmed with biopsy of an abdominal fat pad, bone marrow, or affected organ. This patient achieved a partial response to chemotherapy treatment [3]. Prognosis of primary AL amyloidosis is poor, with median survival of approximately 18 months[4,5].

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Declarations:

Conflict of Interest: *The authors declare that they do not have a conflict of interest.*

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