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ORAL DISEASES Leading in Oral, Maxillofacial, Head & Neck Medicine WILEY

CLINICAL IMAGE

Macroglossia associated with multiple nodular lesions of the tongue

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CASE REPORT

An 81-year-old woman referred pain on the left border of the tongue, xerostomia and a "bulky tongue" sensation that appeared few months earlier, associated with difficulty in swallowing.

The patient had never smoked and rarely consumed alcohol (wine and not spirits). The patient reported to be affected by rheumatoid arthritis and she was on therapy with prednisone 25mg/day. No recent blood tests were available. No cervical lymphadenopathy or extraoral swelling were detectable.

The intraoral examination showed a clinical picture of macroglossia and the presence of multiple lobulated irregular lesions on the borders of the tongue (Figure 1). The lesions had a whitish/yellowish colour, a hard and firm consistency and seemed to originate from submucosal layer. Moreover, at the lower labial mucosa, there was a red purplish lesion compatible with ecchymosis.

WHAT IS YOUR DIAGNOSIS?

Based on the patient's history, physical examination, and laboratory findings, which one of the following is the most suspicious diagnosis?

A. Haemangioma or lymphangioma
B. Amyloidosis of the tongue
C. Submucosal squamous cell carcinoma
D. Schwannoma

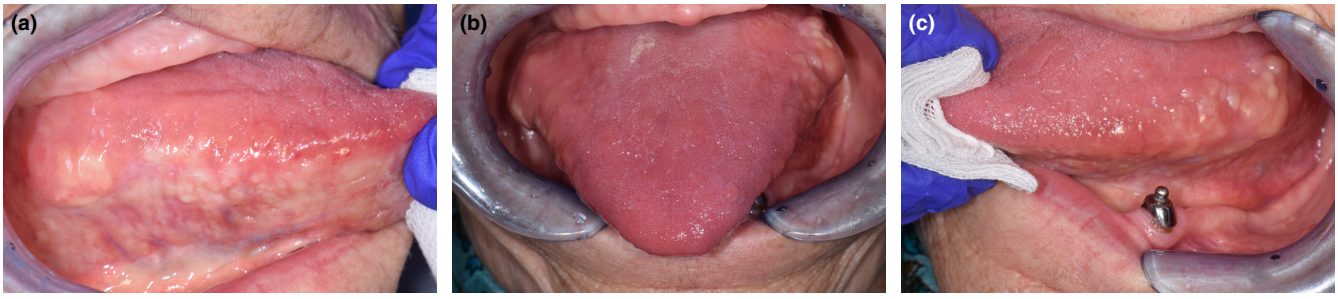

FIGURE 1 Multiple neoformations of the right (a) and left (c) border of the tongue associated to a clinical picture of macroglossia (b)

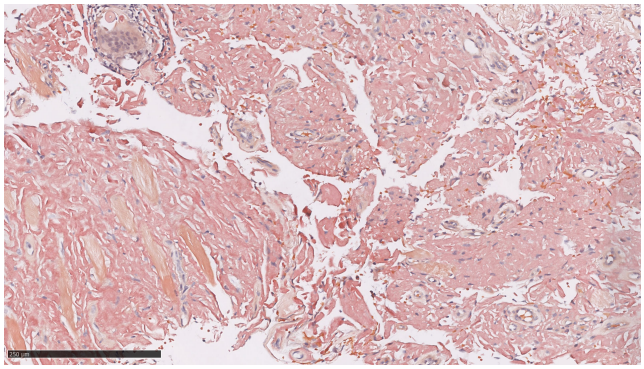

FIGURE 2 Histopathological examination of the specimen with Congo Red stain (15×magnification)

DIAGNOSIS

The right diagnosis is B. Amyloidosis of the tongue. Differential diagnosis may include benign or malignant submucosal lesion, actinomycosis of the tongue, amyloidosis of the tongue (D'Amore et al, 2020; Lombardi et al, 2021). A biopsy was performed on the left border of the tongue, extending deeply to the muscle and submucosal layers. Congo red stain is the gold standard for the demonstration of amyloid in tissue sections, showing amyloid deposits as apple-green birefringence under polarized light. This diagnostic procedure led to the final histological diagnosis of amyloidosis of the tongue (Figure 2). Amyloidosis is characterized by extracellular deposition of insoluble amorphous fibrillar protein. The amyloid deposits may involve few tissues in local forms or various organs, causing a wide range of clinical manifestations (Maturana-Ramírez et al., 2018). The simplified classification of systemic amyloidosis includes: primary amyloidosis (AL), which is the most common type of amyloidosis and it is associated with multiple myeloma; secondary amyloidosis (AA), which is associated with chronic infections or inflammatory diseases, such as rheumatoid arthritis; $A\beta_{2M}$ associated with long-term haemodialysis; familial ATTR amyloidosis, an inherited disease with mutant form of transthyretin (TRR); localized amyloidosis, in which amyloid deposits are usually due to local production of immunoglobulin light chains not originating from the bone marrow (Guijarro-Martínez et al., 2009). Macroglossia is observed in more than 40% of amyloidosis cases and it may be an unique early manifestation of the disease (da Costa et al., 2018; Guijarro-Martínez et al., 2009). However, localized amyloidosis of the tongue is rare and it's often secondary to a systemic form (Guijarro-Martínez et al., 2009). For this reason, it is mandatory to evaluate the systemic involvement of the disease and the presence of other amyloid deposits (Maturana-Ramírez et al., 2018). In case of macroglossia, amyloidosis should be considered in differential diagnosis, since the early diagnosis guarantees appropriate treatment and prevents irreversible organ damages (Maturana-Ramírez et al., 2018).

OUTCOME

Considering the association with rheumatoid arthritis, a provisional diagnosis of secondary amyloidosis was made.

The patient was referred to amyloidosis centre for further diagnostic tests, but she died of pulmonary embolism few weeks later.

CONFLICT OF INTEREST

All authors have no conflicts of interest to disclose.

AUTHOR CONTRIBUTIONS

Niccolò Giancesare Lombardi: Conceptualization; Data curation; Writing – original draft. **Roberto Franchini:** Conceptualization; Writing – review & editing. **Alberto Pispero:** Conceptualization; Writing – review & editing. **Laura Moneghini:** Data curation; Writing – review & editing. **Elena Maria Varoni:** Conceptualization; Data curation; Supervision; Writing – original draft.

PATIENT CONSENT

The patient reported in this manuscript provided written informed consent for the publication of the case details.

PEER REVIEW

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