

Swelling Of The Tongue As The First Symptom For Multiple Myeloma With Lingual Amyloidosis: A Case Report

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Abstract

Background: Swelling of the tongue is a common oral manifestation due to variety of causes. In addition to trauma, infection, allergy, tumor and other causes, a rare cause which is the amyloidosis should be identified. The amyloidosis is a group of rare diseases caused by extracellular accumulation of amyloid. Amyloid deposits progressively disrupt tissue structure and function, and amyloidosis is commonly associated with complex systemic diseases as well.

Case presentation: This case report describes a rare case of the patient with progressively swelling of the tongue as the first symptom for multiple myeloma with lingual amyloidosis. With the treatment of multiple myeloma, the condition of swelling of the tongue stabilized. And the diagnosis and causes of oral amyloidosis were discussed.

Conclusions: The clinicians need to distinguish the oral amyloidosis from other oral diseases to avoid misdiagnosis and incorrect treatment. Multidisciplinary consultation is necessary to find out the underlying systemic diseases of the patients with oral amyloidosis and to achieve early accurate diagnosis and treatment.

Keywords: Swelling of the tongue; Oral amyloidosis; Multiple myeloma; Case Report

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I. Background

Swelling of the tongue is a common symptom and there are variety of reasons for it. In addition to usual causes such as trauma, infection, tumor, angiolymphatic malformation, a rare cause which is lingual amyloidosis should be identified. The amyloidosis is a group of rare diseases caused by extracellular accumulation of amyloid, leading to destruction of organ structure and function. Presentation of oral amyloidosis is generally a rare phenomenon, and tongue being the most common site to manifest the disease process [1]. Amyloidosis can be

classified as primary, secondary, familial and localized [2-3]. The causes of primary amyloidosis are unknown. Secondary amyloidosis is usually associated with systemic diseases, such as osteomyelitis, tuberculosis, rheumatoid arthritis and tumors. Familial amyloidosis associated with genetic genes. Localized amyloidosis is characterized by the deposition of amyloid in a tissue, without other systemic manifestations. Histological demonstration of amyloidosis is essential for confirmation of the diagnosis. Amyloid is identified by the histopathological examination with Congo red stain positive and apple-green birefringence noted under polarized light [1-3].

This case report describes a rare case that the 37-year-old male patient with progressively swelling of the tongue as the first symptom for multiple myeloma with lingual amyloidosis. And the diagnosis and causes of oral amyloidosis were discussed.

II. Case Presentation

A 37-year-old man presented the stomatology department with about 6 months history of progressively swelling of the tongue. And the speaking, eating and sleep of the patient were affected with the enlargement of the tongue. The patient described the burning sensation of his tongue and generalized fatigue over the past several months. The patient denied history of diabetes, tuberculosis, drug allergy history and family cancer.

Oral examination showed that the tongue was obviously enlarged and swollen, with indentation in margin of the tongue. The movement of the tongue was severely restricted and it was difficult to stick out the tongue. Figure 1 and 2 showed the manifestations of the tongue.

The complete blood count (CBC) revealed that the number of red blood cell (CBC) was reduced to $3.63 \times 10^{12}/L$ (normal $4.3 \sim 5.8 \times 10^{12}/L$), the hematocrit (HCT) was reduced to 36.7% (normal 40%~50%), the hemoglobin concentration (HGB) was reduced to 123g/L (normal 130~175g/L). His urine routine test revealed that the PH of the urine was increased to 8.0 (normal 5.0~7.0), urobilinogen (UBG) was positive. The serum immunoglobulin G (IgG) was decreased to 4.67g/L (normal 7.0~16.0g/L), IgA was decreased to <0.26g/L (normal 0.70~4.0G/L), IgM was decreased to <0.18g/L (normal 0.40~2.30/L). HIV testing were negative. The fungal smear of the oral mucosa and the lesions tested negative.

Biopsy of the tongue on Hematoxylin and eosin staining sections showed the pink and homogenous amyloid deposits, and fibrosis with hyalinization. On special stain with Congo Red was positive to indicate the presence of amyloid. Figure 3 showed the histological manifestations of amyloid deposits and Figure 4 showed the manifestations of Congo red staining positive.

Due to the abnormal findings of IgG, A, M and CBC, the patient was referred to the department of hematology for further medical examination. The diagnosis of multiple myeloma was confirmed by the specialists

of hematology according to the findings of bone marrow examination, bone marrow cells by flow cytometry and so on.

III. Diagnosis

The diagnosis of multiple myeloma with lingual amyloidosis was made according to the clinical manifestation, serological test results, bone marrow examination findings and histopathological demonstration.

IV. Treatment

The patient was treated in the department of hematology and under regular multidisciplinary consultation including oral medicine. With the treatment of multiple myeloma, the condition of swelling of the tongue stabilized.

V. Follow-up and outcomes

During the patient's 1-year follow up, we found that the swelling of the tongue did not continue to progress, but there was no significant reduction either. And the follow-up was continued.

VI. Discussion

Amyloidosis of the tongue is characterized by enlarged tongue with indentation in margin of the tongue. Sometimes accompanying infection may lead to congestion, suppuration and necrosis. The speaking, swallowing, breathing and sleeping of the patients are affected as the progress of the disease. Pale yellow amyloid deposits can also be scattered in the mucosa of margin of the tongue, the floor of the mouth, buccal and lips [1-3].

In addition to the tongue, oral amyloidosis can also occur at the floor of the mouth, buccal mucosa, soft palate, gingiva, hard palate, lower lip, and even invade the salivary glands [2-3]. The biopsy of a clinically suspected organ and histological analysis are extremely important to differentiate the amyloidosis from other oral mucosal diseases. Amyloid is identified by the histopathological examination Congo red stain positive and apple-green birefringence noted under polarized light [1-3].

The secondary amyloidosis is commonly associate with underlying systemic diseases, such as multiple myeloma, rheumatoid disease, pulmonary tuberculosis, syphilis, etc [4-6]. Therefore, the patients suspected to be oral amyloidosis subjected to through history taking and clinical examination. Age, occupation, history of smoking, alcohol intake, history of drug intake, family history, blood pressure, diabetes and other systemic diseases, duration of disease, and so on should be recorded. Multidisciplinary consultation and trying to discover and treat the underlying systemic disease is important part of the management to achieve early accurate diagnosis and

treatment.

VII. Conclusions

The clinicians need to distinguish the oral amyloidosis from other oral diseases to avoid misdiagnosis and incorrect treatment. Multidisciplinary consultation is necessary to find out the underlying systemic diseases of the patients with oral amyloidosis and to achieve early accurate diagnosis and treatment.

VIII. List of abbreviations

CBC: complete blood count

HCT: hematocrit

HGB: hemoglobin concentration

UBG: urobilinogen

IgG: serum immunoglobulin G

IgA: serum immunoglobulin A

IgM: serum immunoglobulin M

IX. Declarations

1. Ethics approval and consent to participate

Not applicable

2. Consent for publication

Written informed consent was obtained from the patient for publication of this case report and any accompanying images. A copy of the written consent is available for review by the Editor of this journal.

3. Availability of data and materials

The datasets used and/or analyzed during the current study available from the corresponding author on reasonable request.

4. Competing interests

The author has declared that no competing interest exists.

5. Funding

No funding was received in association with this article.

6. Authors' contributions

Min Zhao performed the physical examination, oral cavity examination, follow-up visits and

multidisciplinary consultation. Min Zhao was the contributor in the biopsy of the tongue and writing the manuscript.

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Figure legends



Figure1 the enlarged tongue



Figure2 the swollen tongue

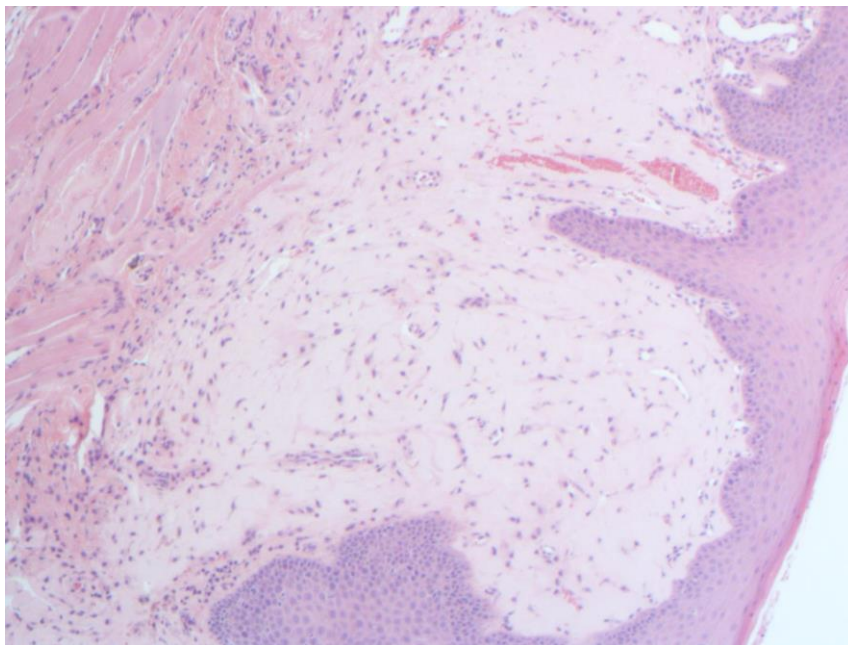


Figure3 Hematoxylin and eosin staining showing the pink and homogenous amyloid deposits

✕ (magnification, ×10)

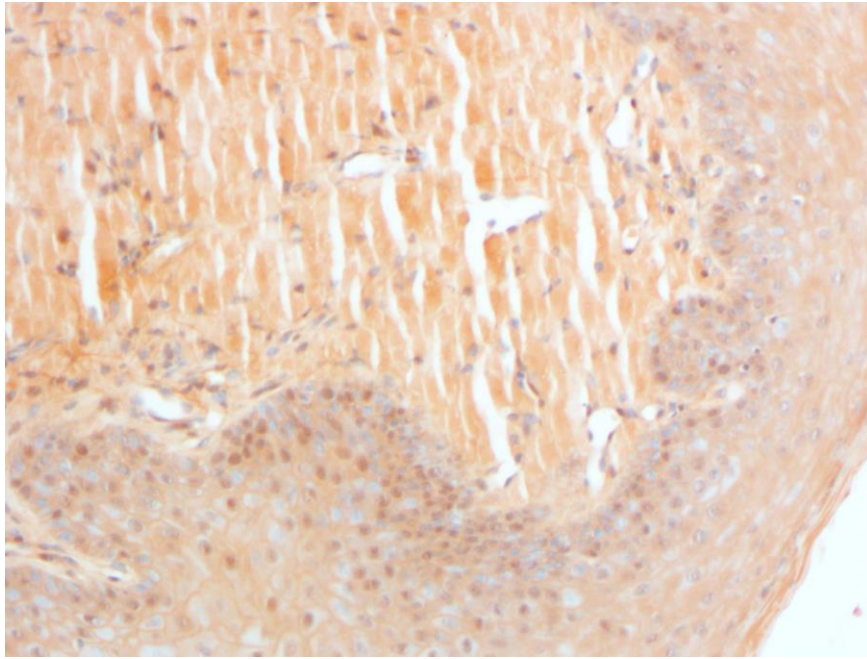


Figure4 Congo red staining positive (magnification, ×40)