Case Report

Macroglossia as a presenting feature of multiple myeloma

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Abstract

Macroglossia has been very rarely reported as a first clinical sign of multiple myeloma.

Keywords: Amyloidosis, multiple myeloma, tongue growth

Background

The term "macroglossia" defines a painless and long-term tongue enlargement, which can be recognized by observation of a resting tongue protruding over the dentoalveolar structures.[1]

Macroglossia is classified into two major categories: true- and pseudo-macroglossia. True macroglossia is associated with definitive histological changes in the tongue and it is frequently observed due to hypertrophy of the tongue muscles or due to infiltration of normal tissue with abnormal proteins or glycogen.[2] Pseudomacroglossia refers to a normal-sized but large-appearing tongue. This condition is usually a result of the anatomical abnormalities of the oral cavity.[3-5]

Case Report

A 54-year-old female presented with tongue enlargement. She had first recognized enlargement of her tongue about 6 months ago. Soon after that, red-purple spots emerged on both her arms, and she began to feel fatigued all the time. Her physical examination revealed an enlarged tongue with teeth indentation marks on both lateral borders [Figure 1]. Her arms were covered with numerous erythematous macules with a diameter of 1 mm [Figure 1]. Bilateral pretibial three positive pitting edema was also observed. Her liver and renal function tests were in normal range, but her complete blood count was consistent with anemia of chronic disease. Her urinalysis revealed proteinuria consistent with nephrotic syndrome. The patient was consulted to internal medicine department with preliminary diagnosis of amyloidosis, hypothyroidism, and lipoid proteinosis. Multiple biopsies from kidneys and bone marrow were performed. Biopsy specimens were stained positive with Congo red and crystal violet in vascular walls and interstitium. According to histopathological findings, a diagnosis of amyloid light type amyloidosis was established. Subsequent bone marrow biopsy revealed an increased percentage (30%) of plasma cells [Figure 2]. The patient was diagnosed with multiple myeloma and underwent chemotherapy treatment. However, she was lost due to multiorgan involvement and loss of function in 6 months.

Conclusion

Amyloidoses are rare diseases characterized by extracellular aggregation of at least 31 different amyloid proteins.[6] Diagnosis

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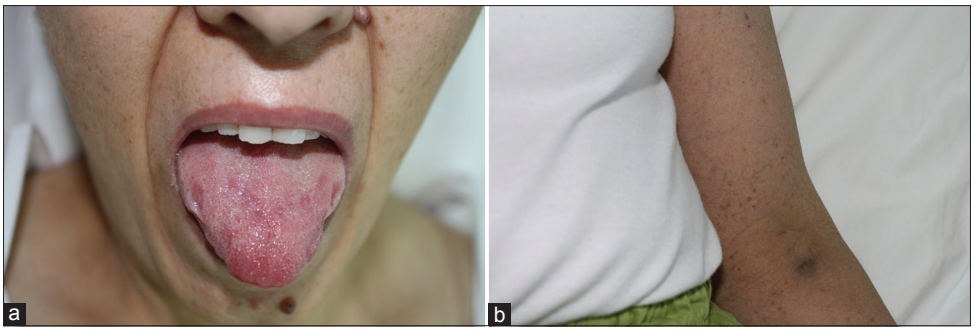

Figure 1: (a) Dental indentations located on lateral surfaces of the tongue as the result of macroglossia (b) Petechiae on the antecubital fossa (pinch purpura)

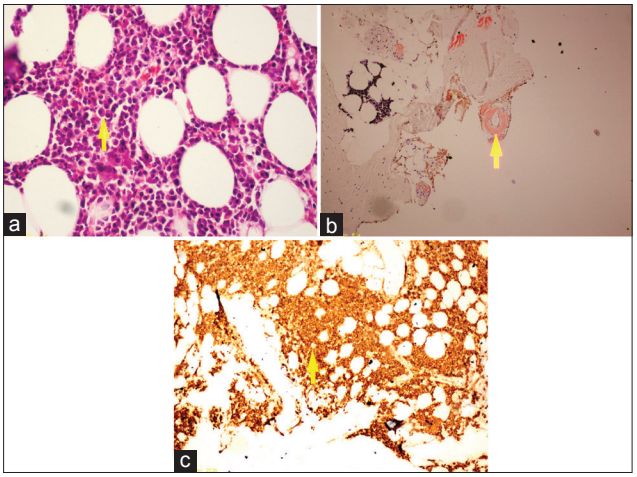

Figure 2: (a) Plasma cells in the bone marrow (H and E, ×400) (b) bone marrow (Congo red, ×100) (c) plasma cells in the bone marrow (CD138, ×100)

of amyloidosis depends on histopathological examination. In routine hematoxylin-eosin staining, homogeneous eosinophilia is observed. Amyloid protiens also react with Congo red stain and yield reflection under polarized microscopy.[7] Amyloid-proteinaceous materials have various effects on cellular level. They may aggregate between cells and cause atrophy. Furthermore, it has a direct toxic effect on cells leading to eventual cell death.[8] Amyloid proteins may accumulate in various internal organs, particularly in the heart and kidneys and also in the tongue.

Macroglossia has been very rarely reported as a first clinical sign of multiple myeloma. In making a diagnosis of multiple myeloma-associated amyloidosis, it is possible for dermatological and venereal diseases' specialist to contribute to early diagnosis by considering the condition seriously when a patient applies with a complaint of tongue enlargement.

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Conflicts of interest

There are no conflicts of interest.

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