A CASE OF MULTIPLE MYELOMA AND AMYLOIDOSIS OF THE TONGUE

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SUMMARY

Multiple myeloma is a clonal plasma cell proliferative disorder. Ten to fifteen per cent of patients with multiple myeloma have associated primary amyloidosis. We describe a case of oral amyloidosis presented with macroglossia and characteristic nodular lesions which developed as a complication of multiple myeloma. Pathogenesis, diagnosis and treatment of oral amyloidosis are also discussed.

Key Words: Amyloidosis, Tongue, Multiple Myeloma.

MULTIPLE MYELOMA AND DILDE AMYLOIDOZIS OLGUSU

Multiple myeloma klonal plazma hücre proliferasyona ile karakterize bir hastalıktır ve multipl myelom hastalarının %10-15’inde primer amiloidozis gelişmektedir. Bu makalede multipl myelomunun komplikasyonu olarak dilde makroglossi ve karakteristik nodüler amiloidosis lezyonları meydana gelen bir olgu sunulmuş ve oral amiloidozis patogenezi, tanısı ve tedavisi kısaca gözden geçirilmişdir.

Anahtar Kelimeler: Amiloidozis, Dil, Multipl Myeloma

Multiple myeloma is a malignant disorder which is characterized by an uncontrolled proliferation of plasma cells in bone marrow. Primary amyloidosis can either arise idioopathically or can be associated with plasma cell discrasia (1,2). Here we present a patient with amyloidosis of the tongue which developed as a complication of multiple myeloma.

Case Report

A 73-year-old man admitted to our clinic with macroglossia and asymptomatic multiple ulcerated nodular lesions on his tongue which first appeared 4 months ago and enlarged gradually. He complained of difficulty in speech and swallowing solid foods. His past medical history revealed multiple myeloma which was diagnosed one year ago and he was still being treated with pulsed courses of vincristine, adriamycine, dexamethasone and pamidronate disodium.

His dermatologic examination revealed slight macroglossia and multiple shiny, reddish-purple, ulcerated nodular lesions on the lateral borders of his tongue (Figure 1).

Apart from slight anemia and high erythrocyte sedimentation rate, laboratory examinations including complete blood count, serum biochemistry, urine analysis and protein electrophoresis were all normal.

An incisional biopsy was made from one of the nodular lesions under the diagnostic possibilities of hemangiomia, lymphangiomia, plasmasitoma and amyloidosis. Dermatopathological examination of the biopsy

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material showed massive eosinophilic amorphous material located in the reticular dermis and stained characteristically positive with Congo-red confirming amyloid deposition (Figure 2).

On the basis of these clinical and dermatopathological data the diagnosis of primary amyloidosis due to multiple myeloma was made. Since the lesions were not causing any significant oral disfunction surgical excision was not performed but regular control visits were planned for a close follow-up.

Discussion

Amyloidosis is a rare, fatal metabolic disorder that leads to extracellular deposition of a sulphated mucopolysaccharide in various tissues and organs (1,3). Systemic amyloidosis is subdivided into immunocyte dyscrasia with amyloidosis (AL-fibril type), reactive systemic amyloidosis (AA-fibril type) and familial systemic amyloidosis. Primary systemic amyloidosis belongs to AL-type amyloidosis. It usually occurs in the setting of multiple myeloma, monoclonal gammopathies and macroglobulinemia.

Figure 1. Macroglossia and multiple shiny, red-purple ulcerated nodular lesions on the lateral border of the tongue.

Figure 2. a) Massive eosinophilic amorphous material in the reticular dermis (Congo Red X 50). b) Amyloid deposits typically stained faint red (Congo Red X 100).
Secondary amyloidosis on the other hand mostly associates chronic inflammatory diseases or chronic infections and usually does not produce skin lesions (4,5). In our patient, primary oral amyloidosis was the result of multiple myeloma which was diagnosed 1 year ago.

Amyloid deposition in multiple myeloma associated systemic amyloidosis occurs as a result of plasma cell dyscrasia and is characterized by the presence of amyloid light chain in which the major protein component is the variable portion of immunoglobulin molecule (5,6). The abnormal monoclonal immunoglobulins are produced by the neoplastic cells. Amyloidosis occurring in multiple myeloma is characterized by the elaboration of light chains (Bence-Jones proteins) by the host. These light chains are converted to amyloid fibrils by proteolytic enzymes in macrophages and secreted to tissues. They can be deposited in connective tissues anywhere in the body and extensive deposition may cause disfunction (7).

Oral manifestations occur in nearly 39% of primary amyloidosis patients in which multiple myeloma associated lesions consist a small portion (1,6,8). Rarely oral amyloidosis may be the first symptom of multiple myeloma (9-11). The amyloid deposits in oral mucosa of primary amyloidosis patients presents as papules, nodules, plaques and macroglossia (1,2,6-8). These lesions may interfere with speech, chewing, swallowing and ability to close mouth. Amyloid deposition in the salivary glands may cause xerostomia. In late stages, lesions may even lead to oropharyngeal blockage (5). Even though macroglossia is known to be the most common manifestation, mucosal nodules are considered to be more specific signs indicative of amyloidosis of the tongue since tongue enlargement can also occur in the absence of amyloidosis (2).

Presence of amyloidosis in multiple myeloma patients is usually associated with poor survival. The median survival time in these patients is assumed to be about 4 months and death usually occurs as a complication of amyloidosis effecting major organ systems (12). We followed up our patient for 1 year and during this period his general status worsened although the size of the oral nodular lesions and macroglossia did not show significant difference. His survival time was relatively longer than the expected.

Since the presence of amyloid deposition in multiple myeloma patients is evaluated as a grave factor and since there are no biochemical or hematologic parameters that associates amyloidosis in these patients, a routine histopathological examination is essential for every multiple myeloma patient with suspected oral lesions (2). Pyogenic granuloma, plasmacytoma and oral tumoral lesions such as lymphangioma, hemangioma and squamous cell carcinoma may also cause similar nodules in the oral mucosa but the diagnosis of amyloidosis can easily be made by typical histopathological findings. Light microscopic examination characteristically shows amorphous eosinophilic material which typically stains pale pink with Congo-red. The material also gives apple-green bi-refrignence under polarised light (1,2,4).

Treatment of oral amyloidosis lesions is nonspecific. Since multiple myeloma is a malignant neoplasm and development of primary amyloidosis shortens the survival, noninvasive and conservative treatments are primarily recommended for localized lesions, but surgical interventions can be inevitable for severe cases with extensive lesions compromising vital functions (1,13,14). In our case, we preferred to follow-up our patient since the lesions were not hindering vital functions.

Eventhough involvement of the oral mucosa in primary amyloidosis is a frequent entity; amyloid deposition on the tongue due to multiple myeloma is rare and indicates a poor prognosis. In this report we described a patient who developed macroglossia and characteristic multiple nodular amyloid deposits on his tongue approximately 8 months after the diagnosis of multiple myeloma and had a relatively long survival than the previously reported cases.
REFERENCES