

Amyloidosis Involving Only Periodontium: Report of a Case

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Abstract:

Amyloidosis includes a wide range of disorders characterized by deposition of amyloid in different organs and tissues. Localized gingival enlargement due to accumulation of amyloid without involvement of other organs is a very rare condition. This article describes a 31-year-old male patient with amyloidosis involving only the periodontium of the anterior mandibular teeth with localized severe horizontal alveolar bone loss. To our knowledge this is the first case report that documents this condition.

Key Words: Amyloidosis; Gingival Overgrowth; Periodontium

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INTRODUCTION

Virchow first used the term amyloid in the mid-19th century to describe the starchy extracellular material observed during postmortem examination of livers [1,2]. The deposition of amyloid, an insoluble proteinaceous fibrillar material, occurs in different tissues and several disorders which are collectively termed "Amyloidosis" [2].

An incidence of 12 cases per million has been reported annually for Amyloidosis. A number of diseases that demonstrate amyloid precipitation are serious and may have a fatal outcome. Amyloidosis along with Alzheimer's disease, Creutzfeldt-Jakob disease and transmissible spongiform encephalopathies are included in the spectrum of conformational diseases. These disorders result from self-aggregation and tissue deposition of constituent proteins due to alterations in size or shape [3].

Amyloid deposits may be either localized or systemic. The type and source of fibrillar pro-

teins can determine the specific arrangement of amyloid precipitation and ultimately the prognosis of the disease [4-6]. The most common types of amyloidosis are primary (amyloid light chain), secondary (amyloid A) and familial, which are categorized according to the source and biochemical composition of the precursor proteins that form the fibrillar deposits [4-7].

Amyloid precipitations may be observed in the skin, synovia and mucous membranes of the oral cavity and gut. The prevalence of primary amyloidosis is higher in males [8]. Several articles have been published reporting the deposition of amyloid in the oral cavity [9-17].

Approximately 45% of cases with systemic precipitation of amyloid are secondary amyloidosis. Various chronic inflammatory conditions such as rheumatoid arthritis, ulcerative colitis and tuberculosis have been reported to induce the secondary or reactive type of this disease [18]. The most commonly involved



Fig 1. Clinical appearance of gingival enlargement in anterior mandibular teeth.

organs are the spleen, liver and kidneys [2,18]. Familial amyloidosis is caused by the mutation of different proteins leading to amyloid deposition and may occur in a group of inherited autosomal-dominant disorders.

CASE REPORT

A 31-year old male presented to the Department of Periodontics, Faculty of Dentistry, Tehran University of Medical Sciences in March 2005, with a one year history of gingival enlargement. A large proportion of the anterior mandibular teeth including the canines and incisors were covered by the excessive gingival tissue (Fig 1).

The enlargement was bluish-red, sessile, elastic and painful with a tendency to bleed spontaneously or while brushing. According to the patient, the mass was initially located between the mandibular central incisors and had gradually increased in size during the last year. The patient's oral hygiene was poor, but he was otherwise healthy and reported no recent history of using medication. A panoramic radiograph revealed localized severe horizontal bone loss in the anterior mandibular region (Fig 2).

Due to poor oral hygiene, heavy calculus had built up under the mass and the patient was scheduled for scaling and root planning. After

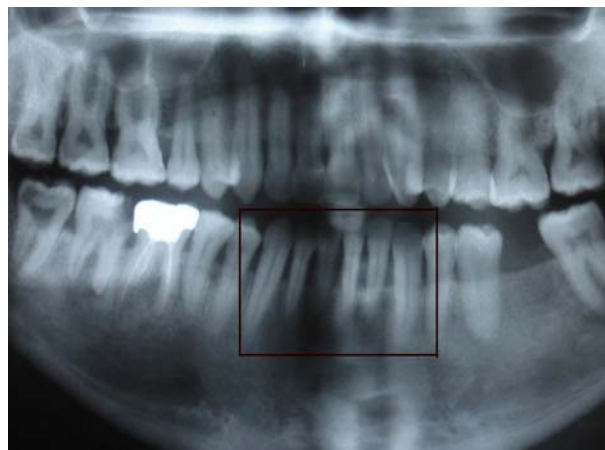


Fig 2. Radiograph showing localized severe alveolar bone loss in anterior mandibular.

completion of phase 1 periodontal therapy, the size of the mass reduced slightly but surgical intervention seemed necessary.

Excisional biopsy was performed following administration of local anesthesia. The mass was removed completely through a horizontal bevel incision placed at the base of the lesion and the specimen was sent for histopathologic examination. A pocket elimination flap was made and granulation tissues were carefully removed. After suturing and dressing, Ibuprofen (400 mg, as required up to 4 times daily) and Amoxicillin (500mg three times per day-every 8 hours) along with 0.2% chlorhexidine digluconate solution (two times daily) were prescribed for 7 days.

One week later, the patient returned for suture removal and follow-up. Clinical examination revealed uneventful healing. Microscopic analysis showed extensive extracellular deposition of an eosinophilic amorphous material in the lamina propria and deep connective tissue, covered by hyperparakeratinized stratified squamous epithelium. Fibroblasts, chronic inflammatory cell infiltration and congested blood vessels were also seen in the lamina propria. The deposits stained positive with Congo red and produced characteristic apple green birefringence under polarized light, confirming the presence of amyloid deposits (Fig

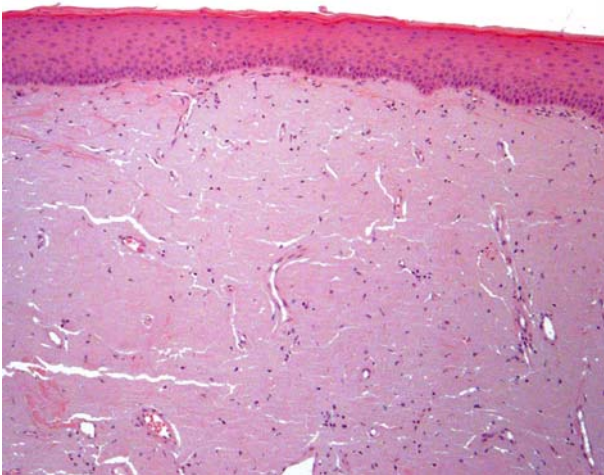


Fig 3. Hematoxylin and Eosin staining of the biopsy specimen; note the extracellular deposits of eosinophilic material (Original magnification, $\times 20$).



Fig 4. The Congo red positive material appears apple green (white arrows) when viewed under polarized light (Original magnification, $\times 20$).

3 and 4). The patient was referred to the hospital for further evaluation. In order to rule out multiple myeloma, bone marrow aspiration was performed and demonstrated normocellular marrow.

The tuberculin test was negative and all urine, blood biochemistry, serologic and hematologic tests were within normal range. Urine protein electrophoresis had a normal pattern.

According to the chest X-ray, the heart and lungs were unremarkable. Upper endoscopic findings showed a normal esophagus, stomach and duodenum. The liver, biliary ducts, gall bladder, pancreas, spleen, kidneys and bladder were also reported normal in the sonographic report. Neck MRI revealed no abnormalities in the airways, sternocleidomastoid muscle, scalene, paraspinal muscles, thyroid gland, vascular structures, parotid and submandibular salivary glands. There was no evidence of an abnormal mass or adenopathy in the cervical spaces. Based on the clinical, radiographic and histopathologic findings, the final diagnosis was primary amyloidosis involving gingiva.

DISCUSSION

Gingival hyperplasia is a common finding in periodontal patients. It may be genetic as in

familial gingival fibromatosis, or acquired following exposure to drugs like phenytoin, cyclosporin and calcium channel blockers [19]. Gingival hyperplasia commonly occurs in the absence of systemic lesions and disorders. However, it can be an insignificant feature of a more serious disease produced by precipitation of abnormal materials. Amyloid deposition may occur in gingival tissues and its diagnosis is confirmed by biopsy [8], especially when there are no clinical symptoms in the patient [21].

Several studies have considered microscopic evaluation to be the most reliable method for diagnosing amyloidosis [2,5,6,22,23]. Histologic diagnosis is highly dependant on factors like the site and sufficiency of the biopsy sample [1]. Congo red staining is employed to confirm the presence of amyloid in tissue specimens and clinicopathologic features can be additionally used to establish the specific type of amyloidosis [1,2].

Urine and blood analyses, tests for liver function and bone marrow biopsies should be obtained for monitoring patients with amyloidosis. Electrocardiography and echocardiography may be required if cardiac symptoms develop [1]. Numerous clinical and paraclinical exami-

nations can help exclude systemic involvement of other organs.

To diagnose the case as primary amyloidosis, secondary amyloidosis should be ruled out. Chronic inflammatory conditions such as tuberculosis, chronic osteomyelitis and rheumatoid arthritis have been associated with secondary amyloidosis. Differentiation between primary amyloidosis and multiple myeloma is sometimes difficult because both disorders belong to the spectrum of the same fundamental disease process. However the lack of Bence-Jones protein in the urine and serum, should point toward primary amyloidosis [24]. Several reports have documented that amyloid deposition in the oral cavity is associated with systemic amyloidosis [9-17].

To our knowledge, the deposition of amyloid within the periodontium without systemic involvement of other organs has not been reported in the English literature. Therefore, based on clinical and histopathologic descriptions, it seems that this is the first case report to describe this finding.

CONCLUSION

The present case report provides further documentation on the deposition of amyloid within the periodontium. Histologic examination of gingival tissues is strongly recommended for patients presenting with gingival enlargements that suggest any kind of systemic disease.

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