Case Report

rare tumors

Rare Tumors
Volume 15: 1–5

DOI: 10.1177/20363613231166540
journals.sagepub.com/home/rtu
SAGE

Solitary amyloid tumor of the palate: A case report and literature review

Farnoosh Razmara[1], Samira Derakhshan[2], Nazanin Mahdavi[2] and Saba Mohammadi[3]

Abstract

Amyloidosis is often caused by the abnormal extracellular accumulation of amyloid in organs and tissues. This condition, affecting the head and neck region, is typically localized, and may also involve the oral cavity, particularly the tongue and buccal mucosa. As a solitary manifestation, the localized amyloidosis occurring intraosseous is highly infrequent. In addition, localized amyloidosis has a great rate of recurrence. In this paper, a 50-year-old female patient with the chief complaint of pain in the anterior of the maxilla is reported. According to clinical examination, no significant pathologic lesion was seen. The radiographic image showed a radiolucent lesion around teeth four and five. The treatment of choice for the patient was an excisional biopsy. As amyloidosis diagnosis is clinically challenging, biopsy and histologic examination of lesions are necessary in this regard. Accordingly, it is concluded that long-term follow-up is mandatory in case of localized amyloidosis because late recurrence can occur in some cases.

Keywords

Amyloidosis, amyloidogenic proteins, palate, hard, rare diseases, congo red

Date received: 2 January 2023; accepted: 13 March 2023

Introduction

Amyloidosis, as a rare progressive metabolic disorder, is identified by the abnormal accumulation of amyloid in extracellular matrices (ECMs). In addition, amyloid is an insoluble fibrillar protein that is misfolded, and ultimately, leads to functional disability.[1,2] Accordingly, this metabolic disorder can be categorized into localized and systemic types. Amyloidosis is subdivided by deposited proteins, including immunoglobulin light chain amyloidosis (AL) and serum amyloid A (SAA), as the most prevalent subtypes. In this case study, a literature review was initially performed on localized amyloidosis, limited to the oral cavity, followed by presenting an unusual localized amyloid tumor on the hard palate. Amyloidosis occurring in the head and the neck region is normally of localized form; however, it may be a manifestation of underlying systemic diseases.[3,4] Primary systemic amyloidosis is also typically related to plasma cell proliferative disorders (PCPDs) and multiple myeloma (MM). In this sense, body organs such as the kidneys, liver, heart, nervous system, and gastrointestinal (GI) tract are the most common sites of deposition. On the other hand, AA form, as the secondary or reactive systemic amyloidosis, is commonly associated with several other chronic inflammatory or infectious diseases, including osteomyelitis (OM), tuberculosis (TB), rheumatoid arthritis, and Crohn's disease.[1,5] Localized amyloidosis is also defined as a single organ involvement, and the most frequently involved sites are the lungs, brain, and skin. Contrary to

[1]Craniomaxillofacial Research Center, Oral and Maxillofacial Surgery Department, School of Dentistry, Tehran University of Medical Sciences, Tehran, Iran
[2]Oral and Maxillofacial Pathology Department, School of Dentistry, Tehran University of Medical Sciences, Tehran, Iran
[3]School of Dentistry, Tehran University of Medical Sciences, Tehran, Iran

Corresponding author:
Saba Mohammadi, School of Dentistry, Tehran University of Medical Sciences, End of North Kargar st, Tehran 1439955991, Iran.
Email: Mohammadi.saba98@gmail.com

localized amyloidosis, multiple organs can be affected in the systemic form.[5] Amyloidosis affecting the head and neck region can similarly occur in the larynx, pharynx, salivary glands, orbit, sinuses, and oral cavity.[6] The oral cavity amyloidosis is less prevalent, whereas it typically has a tendency to involve the tongue or buccal mucosa.[2,3] Most cases with amyloidosis are adult patients, between 50 and 70 years old, with the gender ratio (female/male) of 1:2.[5,7] As amyloid is deposited at different sites, patients have various symptoms, such as arrhythmia, angina, proteinuria, renal failure, myalgia, edema, urinary retention, paresthesia, carpal tunnel syndrome, dyspnea, anorexia nervosa, orthostatic hypotension, and hyposplenism.[8] It should be noted that amyloidosis has infrequent oral manifestations, and may present as nodules, ulcers, papules, petechiae, ecchymoses, hemorrhagic bullae, and macroglossia. In addition, the color of the mucosa may be in a spectrum of yellow to purple.[5] Although amyloid deposits are rare in the oral cavity, once they occur, the tongue is primarily affected, followed by the lip.[5] Localized amyloidosis of the head and neck region is typically a benign condition, which is often uncommon. Moreover, the localized amyloidosis of the hard palate is considerably scarce. The gold standard for amyloidosis diagnosis is thus the histopathological examination of a tissue biopsy from the lesions.[2] The histology of amyloid, using the routine hematoxylin & eosin (H&E) staining, has further demonstrated amyloid in the form of a homogeneous, eosinophilic, and amorphous deposit, representing metachromasia with crystal violet.[5] Once stained with the Congo red dye, amyloid also displays a characteristic apple-green structure, under polarized light microscopy. Furthermore, it reveals purple-red when stained with metachromatic stain (namely, crystal violet).[1,2] Upon the diagnosis of oral cavity amyloidosis, examinations should include the evaluation of systemic involvement. In this sense, abdominal fat tissue aspiration is the most specific test available for this purpose.[6]

Despite localized amyloidosis has an excellent prognosis, appearance of the lesion in the jaw is very rare. Accordingly, long duration of monitoring patient with amyloidosis is highly recommended. In this paper we report a rare case of unusual localized amyloidosis in the hard palate with recurrence after 10 years. We highlight the importance of extensive evaluation of patients with localized amyloidosis to rule-out the systemic involvement, as well as long term follow-up for early detection of reccurence.

Case presentation

A 50-year-old woman with chief complaint of pain in anterior of maxilla for 3 months, was referred to the Department of Oral and Maxillofacial Surgery, affiliated to Tehran University of Medical Sciences, Tehran, Iran. Clinical examination revealed no significant pathologic lesion in the painful area. Gingiva of the tooth six had mild recession with caries in CEJ area. Also, tooth eight had mobility grade III (Figure 1). Although the past medical history (PMH) was not significant but the patients documents revealed history of amyloidosis 10 years ago. According to panoramic radiography, patient had a radiolucent lesion around teeth four and five on the anterior side of the hard palate. The lesion resulted to tooth mobility upon displacement. (Figure 2(a)). Cone-beam computed tomography (CBCT) image showed an intrabony, ill-defined radiolucency in anterior region of the maxillary bone with perforation of hard palate (Figure 2(b)) Radiologic differential diagnosis includes inflammatory process due to periodontal lesion of the involved tooth, and odontogenic tumor.

Therefore, an excisional biopsy was obtained, and the macroscopic examinations represented that the specimen consisted of several pieces of white, creamy, soft to elastic tissues (2 × 1.2 × 0.5 cm in diameter). The histologic examinations correspondingly indicated a fragment of the mucosa with hyperplastic spongiotic stratified squamous epithelium and severely impaired exocytosis in the surface and the adjacent connective tissue, having mixed inflammatory infiltrates and dense collagen bundles (Figure 3(a) and (b)) The foci of scattered, amorphous, and eosinophilic globular structures were also detectable (Figure 3(c)). Histopathologic differential diagnosis are composed of: 1. Inflammatory lesions such as; chronic osteomyelitis, 2. The lesions with prominent inflammatory background, including eosinophilic granuloma, and some rare tumors with amyloid deposition, including: Calcifying epithelial odontogenic tumor (Pindborg tumor). Round to oval-shape accumulation of eosinophilic materials mimicking amyloid material and the absence of eosinophil cells among the inflammation and histiocytes can help to limit the differential diagnosis.

Based on the absence of odontogenic cells, the significant presence of inflammation, and the lack of clear cell shape, and according to the patient's PMH, we excluded pindborg tumor and amyloidosis was the most possible diagnosis. The Congo red staining was further conducted and amyloid deposits showed an dark green structure, confirming the amyloid material in polarized light microscopic picture (Figure 3(d)). The Amyloid diagnosis was additionally established after histopathological staining. According to the blood test results, there were no significant changes in the serum. As any hematological or immunological disorders were excluded in the blood tests, there was no other focus of amyloid deposition and systemic involvement. Follow-up was conducted each 6 months and after 2 years no evidence of recurrence or any significant problem was observed.

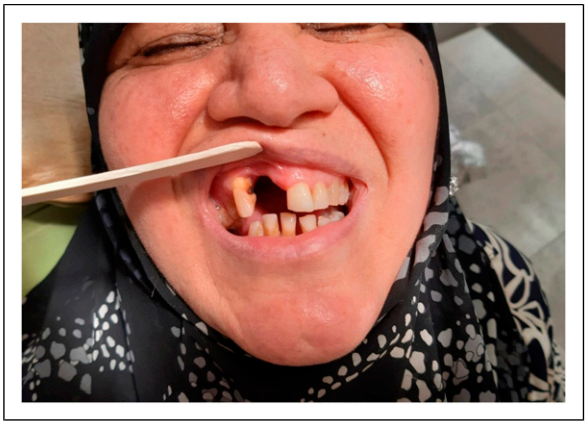

Figure 1. Clinical features.

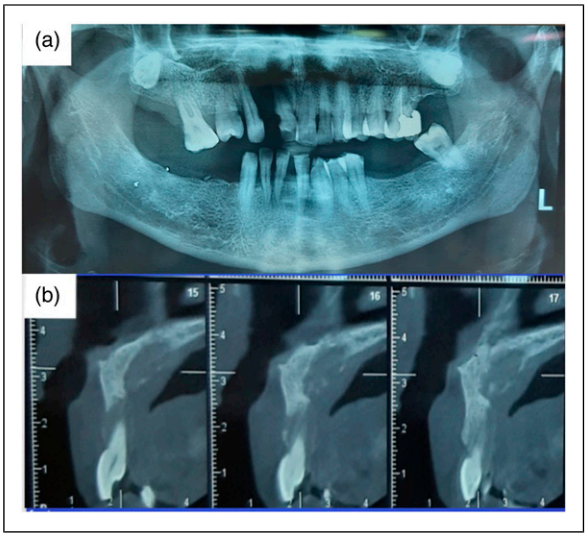

Figure 2. (a) Panoramic view of the lesion: Demonstrates the radiolucent lesion around the teeth four and five with bone destruction, (b) CBCT: ill-defined, radiolucent lesion in anterior of maxillary bone, and palatal perforation.

Discussion

In this paper, a 50 year old female with diagnosis of intraosseous amyloidosis has been reported. Appearance of amyloidosis in mouth, specially in jaw is rare. Furthermore, a literature review was performed on oral cavity amyloidosis.

The accumulation of a specific protein, known as amyloid, in different organs and tissues in the human body, leads to a rare condition called amyloidosis, which was initially devised by Virchow, during the 19th century, characterized by a variety of forms, each one having its own exclusive type of amyloid.[9] The head and neck region is often rare for the amyloid build-up and it usually manifests as the localized form.[10] Oral cavity amyloidosis is extremely rare with a less than 9% prevalence rate.[11] Generally, localized oral amyloidosis is not associated with systemic amyloidosis or associated diseases like multiple myeloma.[12] According to the literature review, most of the patients with oral cavity amyloidosis, represent a localized or organ-limited type, in accordance with Takumi et al.[4] Localized oral cavity amyloidosis is also a rare condition, and the tongue is affected in the first place when it appears.[7] The most involved site in the oral cavity is tongue, followed by the labial and buccal mucosa, hard palate, floor of the mouth, gingiva, lower lip, salivary glands, and soft palate. The manifestations of localized oral cavity amyloidosis usually contain macroglossia, xerostomia, hemorrhagic bullae, mucosal papules, nodules, and petechiae.[10] The most known oral exhibition of amyloidosis is macroglossia. Moreover, macroglossia can be observed in 10–20% of patients with the systemic form.[11] The disease is more common in the group of patients in their 60s and there is a clear prevalence towards the female gender.[13] A biopsy of a tissue specimen is also required to identify amyloidosis. As well, amyloid displays as an orange-pink, homogenous, shapeless, and hyaline-like material under a light microscope using the H&E staining. Moreover, it shows an apple-green structure under a polarizing microscope with the Congo red dye.[10] After the diagnosis of oral cavity amyloidosis, additional examinations to reject related systemic diseases are mandatory. This should include abdominal fat tissue aspiration and bone marrow examination. To rule out plasma cell dyscrasias, bone marrow biopsy is further carried out.[6,14] Generally, systemic amyloidosis has a poor prognosis, which is often associated with the type of tissue affected and the extent of harm occurring in the organs. Contrarily, organ-limited amyloidosis is not a serious and threatening condition and does not normally progress to the systemic form. The prognosis of the localized amyloidosis is influenced by the location and dysfunction of the organs involved.[11] Treatment alternatives for the localized form may thus include follow-ups or surgeries. It is noteworthy that radiotherapy or a wide local excision (WLE) is the treatment of choice if the localized amyloidosis is symptomatic.[11] Localized amyloidosis has a good prognosis and is not life-threatening. It usually does not progress to the systemic form. Symptom management or palliative care along with local therapy is accordingly practiced to manage the localized amyloidosis. Besides, clinical observations and follow-ups are sufficient for patients without symptoms and organ disabilities. The best treatment once the lesions cause pain, and are persistent or enlarged, is to excise it surgically.[11] The treatment of choice for the majority of amyloidosis cases is follow-up. According to papers, the lesion had been thus surgically removed in less than 50% of the patients from. Of note, amyloidosis has an extensive differential diagnosis that ought to be taken into

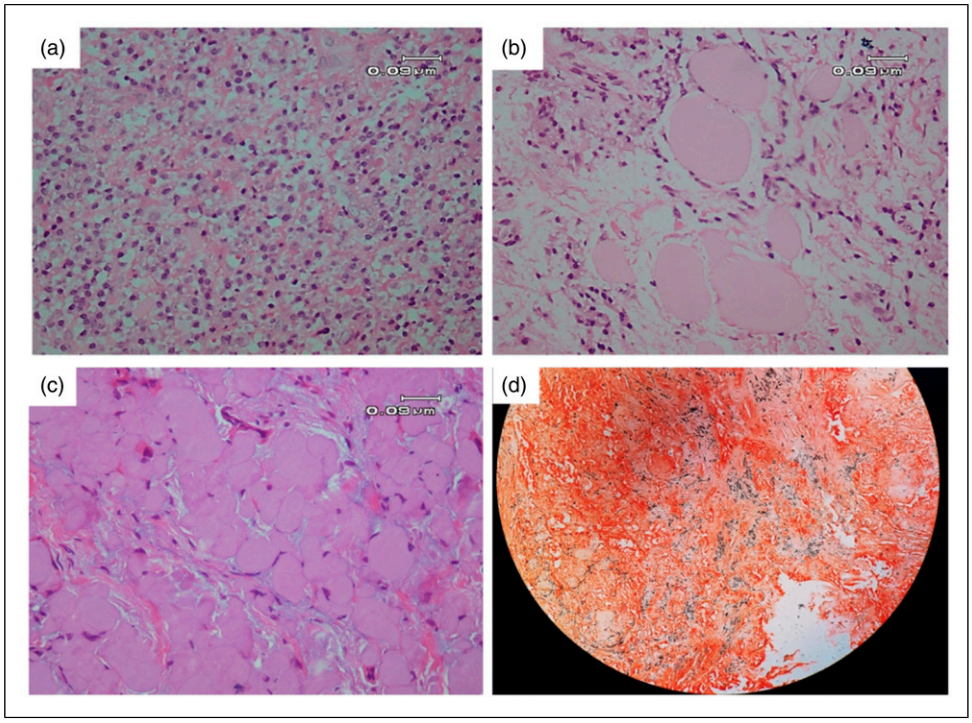

Figure 3. Histopathological view of the lesion; (a) Non-Specific inflamed connective tissue with prominent histiocyte and plasma cell infiltration, (b) Scattered area of round eosinophilic globular structures among the inflamed connective tissue, (c) Aggregation of Amyloid material, (d) Congo red staining was positive for amorphous material and showed an dark-green structure under polarized light.

consideration. The differential diagnosis for nodular lesions includes lipomas, fibromas, granular cell tumors, and salivary gland tumors for nodular lesions. Relapsing polychondritis, sarcoidosis, and Wegener's granulomatosis are also considered in the differential diagnosis list of inflammatory diseases.[4] The primary localized amyloidosis of the hard palate is often uncommon and its management is generally symptomatic. The reported patient had tooth mobility upon displacement, and even some painful symptoms, causing discomfort, so an excisional biopsy was performed. Recurrence of the amyloidosis after 10 years points out that follow-up the patients with this lesion is mandatory. There was no significant limitation in management of our case. In diagnostic process, laboratory tests, treatment plan and all steps of management the patient there was not any limitations and all of the radiographic images, histopathology results, and laboratory tests are available.

Conclusion

As amyloidosis diagnosis is clinically difficult, biopsy and histologic examinations of lesions are obligatory. Deposition of amyloid intra-bony specially in palate is extremely rare. The treatment of localized amyloidosis affecting the hard palate is local surgical or laser excision. Treatment of amyloidosis can lead to reduction in functional disability. Despite the fact that recurrence may arise, follow-up is mandatory for the localized form of amyloidosis.

Acknowledgements

The authors are grateful to the patient who kindly gave consent for this case to be presented in this paper.

Author contributions

FR Contributed to conception and design, drafted and critically revised the manuscript. SM Contributed to conception, design, data acquisition and interpretation, performed all statistical analyses, drafted and critically revised the manuscript. NM Contributed to conception and design and critically revised the manuscript. SD Contributed to conception and design, drafted and critically revised the manuscript. All authors read and approved the final manuscript.

Declaration of conflicting interests

The author(s) declared no potential conflicts of interest with respect to the research, authorship, and/or publication of this article.

Funding

The author(s) received no financial support for the research, authorship, and/or publication of this article.

Ethics approval

This study protocol was approved by the local ethics committee of the Tehran University of Medical Sciences.

Consent to participate

Written informed consent was obtained from the patient for participate in this study.

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-in-Chief of this journal.

Data availability

The information used to support the findings of this study are accessible from the corresponding author upon request.

ORCID iD

Saba Mohammadi https://orcid.org/0000-0002-6957-1392

References

1. Alshehri SA and Hussein MRA. Primary localized amyloidosis of the intestine: a pathologist viewpoint. *Gastroenterology Res* 2020; 13(4): 129–137.
2. Mesolella M, Petruzzi G, Buono S, et al. Focus on localized laryngeal amyloidosis: management of five cases. *Open Med* 2020; 15(1): 327–332.
3. Bucci T, Bucci E, Rullan AMP, et al. Localized amyloidosis of the upper gingiva: a case report. *J Med Case Rep* 2014; 8(1): 198–204.
4. Takumi K, Staziaki PV, Hito R, et al. Amyloidosis in the head and neck: CT findings with clinicopathological correlation. *Eur J Radiol* 2020; 128: 109034.
5. Silva WPP, Wastner BF, Bohn JC, et al. Unusual presentation of oral amyloidosis. *Contemp Clin Dent* 2015; 6(Suppl 1): S282–S284.
6. Serdar A, Basak D, Sercan G, et al. Solitary amyloid tumor of the tongue base. *Int J Otolaryngol* 2009; 2009: 515068.
7. Andreadis D, Poulopoulos A, Papadopoulos P, et al. Localized tongue amyloidosis in a patient with neurofibromatosis type II. *Head Neck Pathol* 2011; 5(3): 302–305.
8. Muchtar E, Dispenzieri A, Magen H, et al. Systemic amyloidosis from A (AA) to T (ATTR): a review. *J Intern Med* 2021; 289(3): 268–292.
9. Neville BW, Damm DD, Allen CM, et al. *Oral and maxillofacial pathology*. Amsterdam, Netherlands: Elsevier Health Sciences, 2015.
10. Balatsouras DG, Eliopoulos P, Assimakopoulos D, et al. Primary local amyloidosis of the palate. *Otolaryngol Head Neck Surg* 2007; 137(2): 348–349.
11. Adamo D, Gasparro R, Marenzi G, et al. Amyloidoma of the tongue: case report, surgical management, and review of the literature. *J Oral Maxillofac Surg* 2020; 78: 1572–1582.
12. Gouvêa AF, Ribeiro ACP, León JE, et al. Head and neck amyloidosis: clinicopathological features and immunohistochemical analysis of 14 cases. *J Oral Pathol Med* 2012; 41(2): 178–185.
13. Liu X, Zhou P and Hua H. Characteristics of orofacial amyloidosis: a case series. *Ann Clin Case Rep* 2016; 1: 1133.
14. Wisniowski B and Wechalekar A. Confirming the diagnosis of amyloidosis. *Acta Haematol* 2020; 143(4): 312–321.