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症　　例

Primary Localization Amyloidosis of the Sublingual Gland

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Abstract We here in present a very rare case of primary localized amyloidosis with amyloid A protein of the sublingual gland. It presented a tumorous appearance on the left oral floor. Pretreatment with potassium premanganate made biopsy specimens unstained by Congo red. Immunohistochemical staining for AA protein was positive. Systemic amyloidosis was ruled out based on clinical and laboratory examinations. The gastric and the labial salivary glands biopsy showed no amyloid deposits. As far as we know, this is the first case of primary localized amyloidosis with amyloid A protein of the oral cavity, and tumor-formed amyloidosis of the sublingual gland.

Key words: sublingual gland, localized amyloidosis, protein AA

Introduction

Primary localized amyloidosis in the oral cavity is a rare, disease, while oral manifestations arising from systemic amyloidosis are reportted relatively frequently reported. The oral amyloid deposits usually occur in the tongue. Amyloidosis of the salivary glands are not common. The majority of such cases occur in the parotid gland. Salivary gland amyloidosis is usually associated with an enlarged gland. However, a nodular or a tumorous appearance, which is also called amyloid tumor or amyloidoma, is very rare. Histologically, in both primary and localized amyloidosis, most amyloid fibrils have been reported to consist of immunogloblin light chain fragments (AL protein).

We here in present the case of a tumorous appearance of localized primary amyloidosis with amyloid A protein (AA protein) of the sublingual gland. To our knowledge, this is the first known case of a sublingual gland amyloidosis and a tumorous appearance of localized AA amyloidosis of the oral cavity in the literature.

Case Report

A 51-year-old man presented at our hospital with a localized painless mass in the left oral floor of 2 months duration. His medical and family history was non-contributory. A clinical examination revealed a film, elastic and mobile mass in the left oral floor, which measured 15×25×10 mm (Fig. 1). Conventional radiographic and ultrasound investigations provided no diagnostic information regarding this lesion. A computed tomographic examination also showed no diagnostic information on the mass but it did show the mass lesion to press

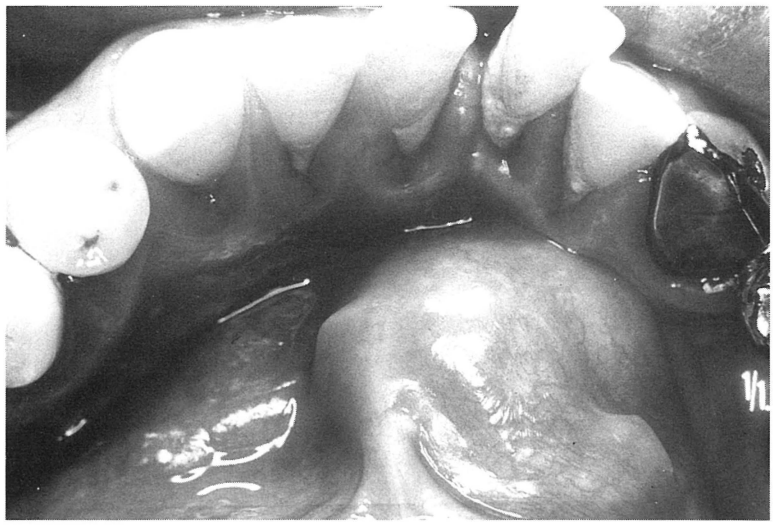

Fig 1. Clinical appearance of the mass of the oral floor.

against the surrounding muscles while not invading them. No clear findings of the lymph nodes were observed. A tentative diagnosis of monomorphic adenoma was thus suggested. The laboratory finding were also unremarkable.

A tumorectomy was performed in April 1992. The tumor was easily removed from the surrounding soft tissue but the tumoral bed consisted of a sublingual gland. The histologic examination demonstrated amyloid deposits in the sublingual gland. The laboratory examination findings for systemic amyloidosis including a gastric biopsy, were negative. Bone marrow aspiration was negative for multiple myelome, which is often complicated amyloidosis. In addition, no evidence of underlyning disease for secondary amyloidosis, rheumatoid disease, chronic infections or inflammatory disease. Serum amyloid A, identified as a precursor of AA protein, was within normal range. Furthermore, we performed a biopsy of the labial salivary glands and it showed no amyloid deposits and no atrophic acini.

For the following 6 months, the patient had no symptoms and no clinical or laboratory evidence of either systemic amyloidosis or any other diseases.

The tumor consisted of extracellular homogenous eosinophilic clumpy material and salivary elements. The eosinophilic material was seen with destruction of both ducts and acini (Fig. 2). A foreign body reaction to the material was seen and the accumulation of plasma cells was noted to be scattered throughout the gland parenchyma (Fig. 3). The eosinophilic material densely stained for alkaline Congo red and also showed typical apple-green birefringence under polarized light. However, after pretreatment with potassium permanganate, it was unstained by Congo red, thus indicating that the amyloid to be AA type. Furthermore, immunohistochemical staining for AA protein was also positive. Staining for light and heavy chains revealed polyclonal infiltration of plasma cells. κ : λ ratio was about 4 : 1, thus indicating the plasma cells to demonstrate inflammation.

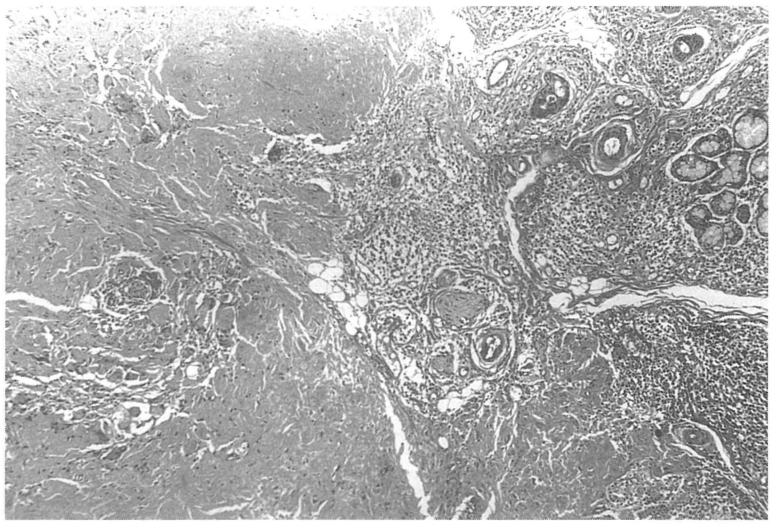

Fig 2. Salivary parenchyma destroied and replaced by homogenous eosinophilic material. (H. E.×4)

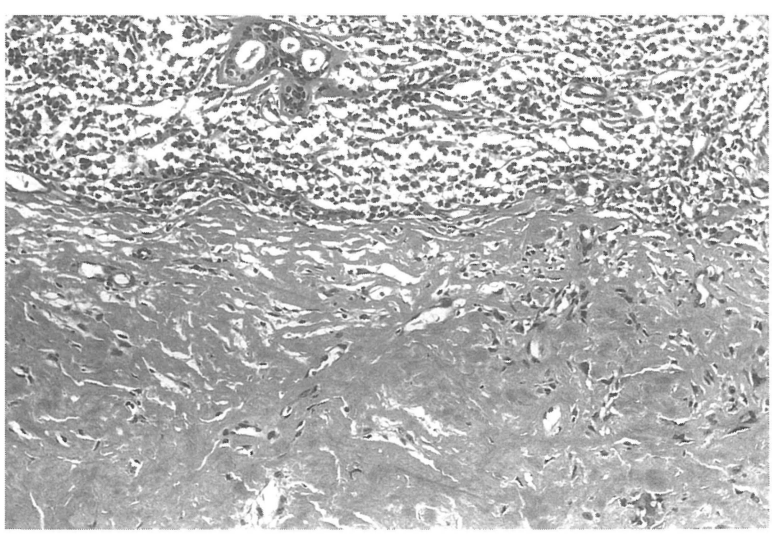

Fig 3. Infiltrations of plasma cells and foreign body reaction to the material. (H. E. ×20)

Discussion

Amyloidosis is a metabolic disease resulting from the extracellular accumulation of abnormal protein materials in various types of body tissue. Clinically, amyloidosis is classified according to whether it is primary or secondary and also whether it is systemic or localized. Biochemically, it is classified based on the type of amyloid fibril proteins, of which there are predominantly two types: (1) AL in which the fibrils consist of immunogloblin light chain fragments and (2) AA in which the fibrils consist primarily of protein AA. The amyloid

protein found in the majority of primary amyloidosis was AL type and AA protein has been identified in secondary amyloidosis. A few cases of primary systemic AA amyloidosis have been observed but few reports of primary localized amyloidosis with AA protein have appeared in the literature.

In the present case, the amyloid protein was AA type. However, the results of clinical and laboratory examinations ruled out systemic amyloidosis. In addition, no evidence of any predisposing condition for secondary amyloidosis was observed. To establish a diagnosis of systemic amyloidosis, gastrointestinal tract biopsy may be the best, and clinically, a gastric biopsy is both useful and easy to perform. Recently, a labial salivary gland biopsy is also recommended as a safe and highly sensitive method. In the present case, no amyloid deposits were seen in the both biopsy specimens. We, therefore, confirmed that systemic amyloidosis could thus be ruled out. Mufarrij et al., reviewed some reports of localized primary amyloidosis of different organs. All cases were histologically associated with local chronic inflammation while a neoplasm was simulated, that same as in our case. However, the amyloid was AL type in all the reported cases, while ours was AA type. To our knowledge, this the first report of localized primary AA amyloidosis in the oral cavity.

In the oral cavity, the tongue is a commonly affected organ in amyloidosis. Only a few cases have shown amyloid deposits in the salivary gland, the majority of such cases are found in the palotid gland while some are also found in the submandibular gland, and most such patients complain of swollen glands. The localized tumorous form of amyloidosis is rare. This form of amyloidosis in a sublingual gland has not been previously reported. One case, with an amyloid tumor of the oral floor, which is the same as in our case, has been reported, however, this case was not proposed of the original tissue or organ. Our case is also the first such description in the sublingual gland.

We here in presented a very rare case. This is the first such case to be reported in the English literature regarding the following, 1) it was a localized primary AA amyloidosis, 2) it was a tumorous form of amyloidosis (amyloid tumor) of the sublingual gland.

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(和文抄録)

舌下腺に発現した限局性アミロイドーシスの一例

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アミロイド症は，アミロイド細線維を主成分とするアミロイド蛋白が，種々の組織間隙に沈着する原因不明の代謝性疾患である．口腔内に症状を認めるものは，そのほとんどが全身性アミロイド症の一分症であり全身に異常がなく口腔内に限局性に腫瘤を形成するアミロイド症はきわめてまれである．口腔内に発現する場合，舌に高頻度にみられる．

唾液腺では耳下腺に好発するが，その頻度は少なく，唾液腺に発症した場合，腺組織の増大を示すが，結節状もしくは腫瘍状に発現した限局性腫瘤形成アミロイド症はきわめてまれな疾患である．

免疫組織学的にアミロイド蛋白の同定が行われるようになり，アミロイド細線維は前駆物質が免疫グロブリンL鎖であり原発性アミロイド症や多発性骨髄腫に伴うアミロイド症にみられるAL蛋白，免疫グロブリン由来でない続発性アミロイド症にみられるAA蛋白，遺伝性アミロイド症の一部にみられるAF蛋白とにわけられている．限局性アミロイド症ではAE蛋白，AS蛋白，AD蛋白などが知られている．

われわれは，51歳，男性の舌下線に限局性，結節性に発現し，そのアミロイド細線維がAA蛋白により構成されたきわめてまれな症例を経験したので若干の文献的考察を加えて報告した．