Primary Localized Amyloidosis of the Angle of Mouth: Report of a case

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A case of primary localized amyloidosis of the angle of mouth is reported. The patient was a 64-year-old woman, and physical examination revealed a lesion about the size of a grain of rice on the left angle of mouth. Diagnosis was made by histopathologic study of surgical specimen. Immunohistochemical study of the infiltrated plasma cells suggested that the origin of amyloid protein to be amyloid L protein. Systemic amyloidosis was ruled out based on clinical and laboratory examination. At follow up 15 months later, no recurrence was observed and no symptoms were present.

Key words: localized amyloidosis, AL protein, angle of mouth
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Introduction
Amyloidosis is a metabolic disease resulting from the extracellular accumulation of abnormal protein materials in various types of body tissue. Clinically, amyloidosis is categorized into two main forms, localized and systemic (1). The localized form of amyloidosis is a rare disease, in which amyloid deposits are limited to a single organ or tissue without involvement of any other sites in the body. The deposits may produce grossly detectable nodular masses or be evident only on microscopic examination. Nodular (tumor-forming) amyloid deposits are most often encountered in the lung, larynx, skin, urinary bladder, tongue, and the region of the eye (2-5).

We report a case of primary localized amyloidosis with amyloid L protein of the angle of mouth, and also review the literature, especially emphasizing the clinicopathologic and immunohistochemical aspects.

Case report
A 64-year-old woman visited our hospital on 26th September 2000. She had noticed a localized painless mass on the left corner of her mouth 6 months earlier. Her medical history included a colon polyp operation at the age of 40. Collagen disease was suspected at the age of 54, and at the age of 57 she was diagnosed by her family doctor as having Sjögren's Syndrome. On examination, there was a grain of rice size, elastic hard, reddish mass on the left angle of mouth (Fig. 1). In addition, the patient had xerostomia and a fissured tongue.

Laboratory findings disclosed the following: white blood cells 4800/µl, red blood cells 450×10⁴/µl, hemoglobin 13.7g/dl, platelets 21.8×10⁵/µl. BUN, creatin, bilirubin, glutamic oxaloacetic transaminase and glutamic piruvic transaminase values were normal. There was no evidence of monoclonal protein (M protein, Bence Jones protein) as shown by serum and urine immunoelectrophoresis. Antibodies to SS-B, rheumatoid factor and se-

Fig. 1: Clinical appearance of the mass in the left angle of mouth.
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rum amyloid A protein were negative or within the normal range. The titer of antibody against SS-A was 4:1. Antinuclear antibody was positive. Schirmer test and Rose Bengal stain were positive. Though the lip biopsy was not examined, these results applied Sjögren’s Syndrome using European criteria.

The clinical diagnosis was granuloma. Under local anesthesia, the mass lesion was excised with surrounding normal tissues. Histopathologically, the mass consisted of homogeneous eosinophilic clumpy material with monomorphic infiltration by mature plasma cells (Fig 2). Eosinophilic material was stained by alkaline congo red staining and showed apple-green birefringence under polarized light microscopy (Fig 3). The eosinophilic material was diagnosed as amyloid deposits. Furthermore, immunohistochemical staining with anti IgG, M, A, chain, and chain antibodies was performed. Immunohistochemical evaluation was performed with the avidin-biotin-peroxidase complex (ABC) method using the following antibodies: IgG, IgM, IgA (Dako, Carpinteria, CA; monoclonal, working dilution, 1:5000), chain (Novocastra, Newcastle, UK; monoclonal, 1:600), and chain (Novocastra, Newcastle, UK; monoclonal, 1:800).

Anti IgG and chain antibodies revealed monoclonal infiltration of plasma cells (Fig 4). Anti IgM, A, and chain antibodies were negative. Amyloid deposits reacted with anti chain but not with anti chain antibody (Fig 5).

Further possibility of systemic amyloidosis was investigated. However, there was no evidence of systemic amyloidosis and multiple myeloma because biochemical and radiologic screening investigation failed to demonstrate any involvement of liver, kidney, heart and bone with amyloidosis. Biopsy of the gastrointestinal tract (duodenal mucosa) showed no amyloid deposits. These results suggest that the present case is primary localized amyloidosis of the left angle of mouth.

At follow-up 15 months later, no recurrence was observed and no symptoms were present.

Discussion

Including our case, 16 cases of localized amyloidosis in the oral cavity have been reported in Japan (6-17). Seven patients were male, and nine were female, with ages ranging from 38 to 78 years (median, 58.9 years). Five cases involved the buccal mucosa; four cases the lip; two cases the oral floor, tongue and sublingual gland; and
one case the palate and angle of mouth. Amyloidosis of only the angle of mouth has not previously been reported. Four cases had a history of chronic inflammatory disorders: three cases were chronic hepatitis, and one case was secondary Sjögren’s Syndrome (with rheumatoid arthritis). In terms of clinical features, all cases were tumorous in appearance. In immunohistochemical classifications, primary amyloidosis (AL) was detected in seven cases, secondary amyloidosis (AA) in two cases, and non-AA,AL in one case; six cases were not examined.

Systemic amyloidosis occurs in several different settings, including a primary form that occurs sporadically and a secondary form that is often associated with chronic inflammatory disease such as rheumatoid arthritis, tuberculosis, inflammatory disease and so on (1, 2). Each of these clinical forms generalizes with distinctly different amyloid proteins. The amyloid protein characteristically found in AL is of immunoglobulin origin, derived specifically from the variable region of immunoglobulin light chains. The protein in AA is derived from an acute phase reactant produced in the liver (2). In the present case, the immunohistochemical examination of amyloid protein revealed AL protein. There was no evidence of systemic amyloidosis for several clinical examinations, which included a biopsy of the gastrointestinal tract. We, therefore, ruled out systemic amyloidosis. Imamura et al. (11) reported that localized tumoruous forms of amyloidosis are associated with local chronic inflammation. Our present case had frequent angular cheilitis due to xerostomia from Sjögren’s Syndrome. We suggest that the mass in the angle of mouth may be caused by chronic inflammation. As a result of pathology, there was monotonous infiltration of plasma cells. Furthermore immunohistochemical staining against chain and IgG revealed clonality of plasma cells, and amyloid deposits reacted with anti chain but not with anti chain antibodies, thus indicating that the amyloid protein is derived from immunoglobulin light chain fragments. These results lead us to conclude that the present case is primary localized amyloidosis of the left angle of mouth. However, since the patient in the present case has Sjögren’s Syndrome, we cannot exclude the possibility that the monoclonal infiltrations of plasma cells were neoplastic lesions.

Local surgical excision is the best choice of treatment for localized amyloidosis in the head and neck, and
since surgery, the present case has been free of recurrence for 15 months. Because the literature also contains a report of multiple recurrences of tumorous amyloidosis for more than 10 years after initial treatment (18), careful long-term follow-up is needed.

References

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