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Case Report

Amyloid deposition in bilateral mandibular condyles and buccal mucosa

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ABSTRACT

We report a very rare case of systemic amyloidosis occurring in bilateral mandibular condyles and showing condylar bone destruction and lesions in the buccal mucosa bilaterally. The patient was a 61-year-old man complaining of malocclusion in which the lower teeth extended abnormally over the upper teeth. He first noticed this about 1 year earlier. Medical history included renal cancer, rheumatic myalgia in multiple joints, osteoporosis, hypothyroidism, chronic hepatitis, malignant lymphoma and hemodialysis for renal failure. Panoramic radiography and computed tomography showed bone destruction in bilateral mandibular condyles and a soft tissue mass lesion around the condyle. Slightly raised mass lesions with areas of whitish and purplish coloration were also observed on the buccal mucosa bilaterally. Histopathological examination revealed amyloid deposition in these lesions, and AA and/or Aβ2M amyloidosis was diagnosed. However, we were informed that the patient died within 3 months from when he had a last check-up at our clinic due to systemic problems.

1. Introduction

Amyloidosis can be divided into systemic amyloidosis, in which proteins with a fiber structure referred to as amyloid are deposited in organs systemically, and localized amyloidosis with deposition only into specific organs. Localized amyloidosis is further classified into clinical types corresponding to the various amyloid proteins involved. We report a case of systemic amyloidosis presenting bilaterally in the mandibular condyles and buccal mucosa.

2. Case report

A 61-year-old man visited a local general hospital with otalgia and tinnitus of the left ear and malocclusion. Medical history included renal cancer, rheumatic myalgia in multiple joints, osteoporosis, hypothyroidism, chronic hepatitis, malignant lymphoma and hemodialysis to treat renal failure.

Computed tomography (CT) and magnetic resonance imaging (MRI) showed a neoplastic lesion in the superior articular cavity on the left side of the temporomandibular joint (TMJ), leading to a suspicion of synovial chondromatosis and pseudogout. Biopsy was scheduled, but was canceled because his general condition was insufficient and symptoms did not appear to be deteriorating.

After 6 months, the neoplastic lesion had expanded to the temporal bone and a new lesion was detected in the contralateral TMJ. Dialysis amyloidosis was suspected and the patient was subsequently referred to our hospital. The progress of malignant lymphoma is observed because that is seemed to be low-grade. Therefore the metastasis of malignant lymphoma toward the TMJ and buccal mucosa is undeniable. Besides, the operation was already performed due to renal cancer and medications are on for other diseases.

Open bite and slight swelling around the right TMJ were observed (Fig. 1), and panoramic radiography and CT showed bone destruction in bilateral mandibular condyles. MRI also showed a soft-tissue neoplasm around the condyle on T1-weighted imaging. Upper and lower joint compartments are included in this lesion. The lesion of left is infiltrated to the temporal bone, whereas that of

Abbreviations: AL, amyloid light-chain; AA, amyloid A; AF, familial amyloidosis; ATTRwt, transthyretin-related amyloidosis wild-type; AH, amyloid A hereditary; NT-proBNP, N-terminal proBrain natriuretic peptide.

☆ Asian AOMS: Asian Association of Oral and Maxillofacial Surgeons; ASOMP: Asian Society of Oral and Maxillofacial Pathology; JSOP: Japanese Society of Oral Pathology; JSOMS: Japanese Society of Oral and Maxillofacial Surgeons; JSOM: Japanese Society of Oral Medicine; JAMI: Japanese Academy of Maxillofacial Implants.

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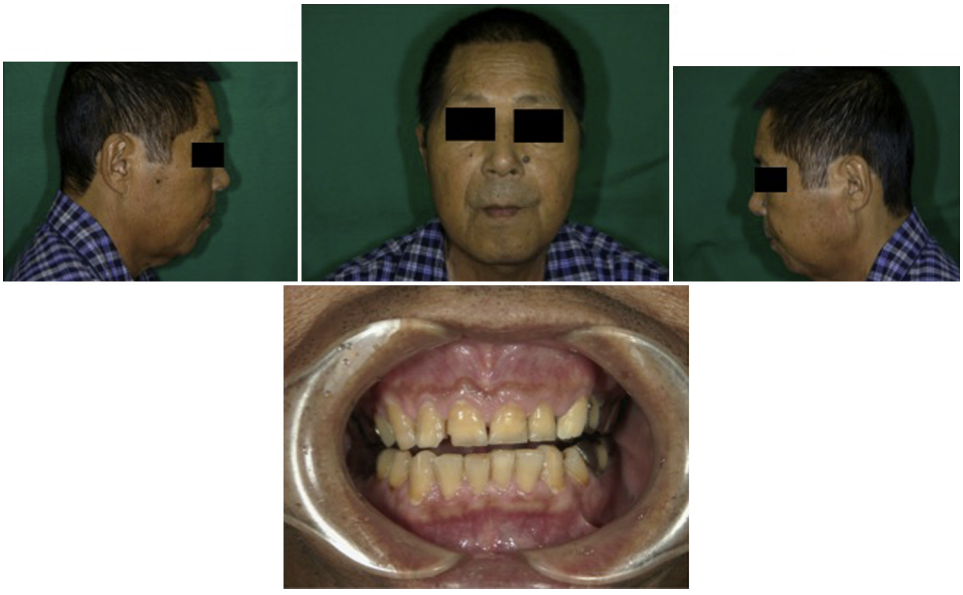

Fig. 1. Open bite and slight swelling the right TMJ.

right is also infiltrated to the cranial base. An irregular margin was showed at both lesions and upper and lower joint compartments almost cannot be seen (Fig. 2).

Blood examinations showed low levels of erythrocytes, hemoglobin and platelets and high levels of creatinine (Table 1). Biopsy under general anesthesia was scheduled, but canceled because of the poor general condition of the patient.

Table 1
Blood test results 1.

WBC	$7.5 \times 10^3/\mu l$
RBC	$299 \times 10^4/\mu l$
Hemoglobin	10.8 g/dl
Hematocrit	31.10%
Platelets	10.7×10^4 μl
CRP	0.08 mg/dl
Total protein	6.1 g/dl
Albumin	3.7 g/dl
Creatinine	5.98 mg/dl
GOT	26 mU/ml
GPT	19 mU/ml
CPK	27 mU/ml
Na	140 mmol/l
K	4.0 mmol/l
Cl	101 mmol/l

After 8 months, pain around the left TMJ and slightly raised mass lesions with areas of whitish and purplish coloration were observed on the buccal mucosa bilaterally (Fig. 3). His general condition subsequently improved, so biopsy of the TMJ and buccal mucosa was performed (Fig. 4). The tissue displayed a pink amorphous, eosinophilic structure with hematoxylin and eosin staining, along with positive Congo-red staining and characteristic birefringence under polarized light microscopy (Fig. 5). Amyloid deposition in the TMJ and buccal mucosa was identified and systemic amyloidosis was diagnosed. Blood examination showed high levels of serum amyloid A protein (SAA) and β2-microglobulin (Table 2).

Table 2
Blood test results 2.

Serum amyloid A protein	36.0 μg/ml (normal value: <8.0 μg/ml)
β2-microglobulin	46.4 mg/l (normal value: <2.0 mg/l)

The patient was therefore diagnosed with AA amyloidosis and/or Aβ2M amyloidosis. Good control of symptoms was achieved and occlusal adjustment was continued. Open bite and slight swelling of TMJ were improved. And then the progress of them is observed. However, we were informed that the patient died within 3 months due to systemic problems.

3. Discussion

Systemic amyloidosis is caused by conformational changes and aggregation of autologous proteins that are deposited in tissues in the form of fibrils. Almost 15 forms of systemic amyloidosis are known and classified according to the different amyloidogenic precursor proteins [8]. However, the following 5 types of amyloidosis account for more than 99% of cases encountered as "Classification of the main types of systemic amyloidosis" (Table 3):

Table 3
Classification of the main types of systemic amyloidosis.

Denomination	Clinical syndrome	Amyloidogenic protein
AL	Monoclonal gammopathy	Immunoglobulins/light chains
AA	Sustained, chronic inflammation	Serum amyloid A protein
AF	Familial polyneuropathy	Mutant transthyretin
	Familial cardiomyopathy	A1-apolipoprotein
	Familial nephropathy	Gelsolin
		Fibrinogen
		Lysozyme
ATTRwt	Senile restrictive cardiomyopathy	Wild-type transthyretin
AH	Dialysis-related	β2-Microglobulin

(1) Immunoglobulin light chain (AL) amyloidosis
AL amyloidosis is the most common form of systemic amyloidosis. In AL amyloidosis, the pathogenic protein is a monoclonal light chain produced by a small-sized bone marrow plasma cell clone.
(2) Reactive (AA) amyloidosis
In AA amyloidosis, the amyloid fibrils are formed by proteolytic fragments of the acute-phase protein SAA. Reactive

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