



Lingual amyloidosis in a long-term hemodialysis patient

Chien-Chin Chen^{a,b}, Kai-Sheng Liao^{a,c*}

^aDepartment of Pathology, Ditmanson Medical Foundation Chia-Yi Christian Hospital, Chiayi, Taiwan, ^bDepartment of Cosmetic Science, Chia Nan University of Pharmacy and Science, Tainan, Taiwan, ^cDepartment of Nursing, Chung-Jen Junior College of Nursing, Health Sciences and Management, Chiayi, Taiwan

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A 55-year-old female had autosomal dominant polycystic kidney disease, chronic hepatitis C virus infection, bilateral carpal tunnel syndrome, renal hyperparathyroidism, and end-stage kidney disease under regular hemodialysis for 26 years. Her family history was unremarkable. She complained of paroxysmal pain in the tongue for several weeks, and she came to our oral surgery department for help. During the physical examination, multiple yellow-to-tan submucosal nodular masses were found in the whole tongue, especially the right side of the tongue [Figure 1a and b]. The consistency was solid and firm. She had no speech disorders or restriction of tongue mobility. On suspicion of abnormal calcification or malignant tumor, the lingual masses were under a diagnostic incisional biopsy. Histological examination revealed abundant deposition of pale pink amorphous substance in the submucosal region, vascular walls, and muscles [Figure 1c, H and E $\times 100$]. Congo red staining demonstrated yellow-green birefringence of the deposition observed by a polarizing microscope [Figure 1d, dark field, Congo red $\times 100$]. Although the clinical doctors did not perform electrophoresis, based on the clinical history and complementary examination, dialysis-related amyloidosis was most likely. The magnetic resonance imaging of lower extremities, 34 months after tongue biopsy, revealed multiple heterogeneous contrast-enhanced lesions over bilateral gluteus muscle, iliacus muscle, and around bilateral hip joints. Although she did not receive biopsy for tissue proof, dialysis-related amyloidosis involving osteoarticular structures and muscles was suspected. She was well after 71 months of postoperative follow-up.

Amyloids are glycoproteins that have a fibrous structure and are not seen in normal metabolism. They consist of various precursors such as immunoglobulin light chains, abnormal prealbumin, and serum proteins [1]. Herein, in each disease, the amyloids have a different composition. Clinically, amyloidosis has two groups: localized amyloidoses and systemic amyloidoses. The systemic amyloidoses consist of AL amyloidosis, AA amyloidosis, familial systemic amyloidosis, age-related amyloidosis, and hemodialysis-related amyloidosis (HRA). For a patient of long-term hemodialysis, β_2 microglobulin gradually

accumulates and results in amyloidosis because it is unable to cross the dialysis filter [2]. HRA predominantly occurs in the osteoarticular structures, but it is also systemically found to appear in the other tissues and organs as well [3]. The clinical manifestations of HRA markedly increase in patients treated with regular dialysis for longer than 10 years, and it is more common in patients who start dialysis when older than 50 years of age [4]. Although HRA is common morbidity of dialysis, dialysis-related lingual amyloidosis is relatively rare. In a reported case series, among 472 patients treated with dialysis for more than 10 years, only eight patients (1.69%) developed lingual amyloidosis while all of them had preceding osteoarticular amyloidosis [5]. Our case also had medical histories of bilateral carpal tunnel syndromes which might be associated with osteoarticular involvement of HRA although they were not histologically proven. In regard to the clinical signs and symptoms, dialysis-related lingual amyloid deposition predominantly starts on the lateral side of the tongue and then develops diffuse nodules probably due to frequent contact with the teeth and related stimulation [5]. The treatment for HRA is difficult and largely unsatisfactory, but successful renal transplantation leads to rapid disappearance of symptoms and no further progression [6]. In addition, biocompatible “high-flux” synthetic membranes are recommended to be used for hemodialysis since they can significantly reduce the prevalence of HRA and thereby improve such symptoms [4,7].

The histological proof is essential for diagnosis, although detection of Bence-Jones proteins in the urine and detection of M proteins by electrophoresis of the serum are helpful for the diagnosis of systemic amyloidosis. The characteristics of amyloid deposits include (a) appearance of amorphous, eosinophilic deposits under light microscope after hematoxylin and eosin staining (b) a bright, apple-green birefringence under polarized light after staining with Congo red, and (c) a β -pleated sheet structure which is demonstrated on X-ray

*Address for correspondence:

Dr. Kai-Sheng Liao,
Department of Pathology, Ditmanson Medical Foundation Chia-Yi Christian Hospital, 539, Zhongxiao Road, East District, Chiayi, Taiwan.
E-mail: carl.liao@gmail.com

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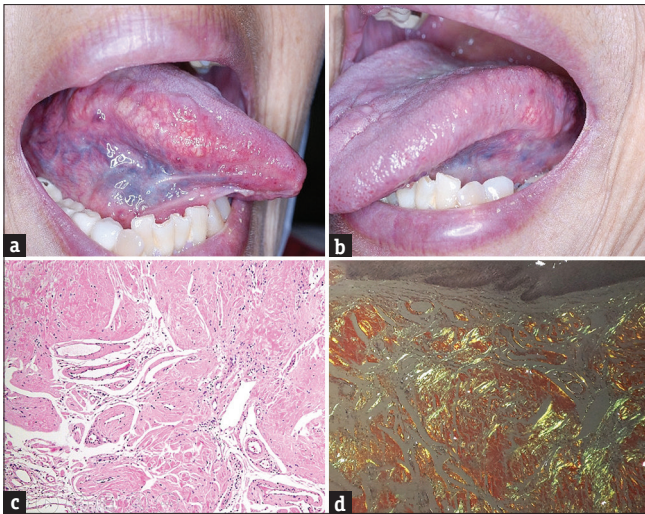


Figure 1: There were whitish-yellow firm nodules in the tongue which appeared in various sizes greater than 5 mm in diameter (a and b). In the light microscopic study, the biopsy revealed abundant deposition of pale pink amorphous substance in the submucosal region, vascular walls, and muscles ([c] H and E, $\times 100$). The deposits were strongly stained for Congo red and showed an apple-green birefringence under crossed polarized light ([d], dark field, Congo red, $\times 100$)

diffraction [8]. Our case fulfilled all these criteria. It is important to rule out systemic from localized amyloidosis, since the etiology, treatment, and outcome of both vary.

In summary, we present a classic case of dialysis-related amyloidosis of tongue, a rare presentation of HRA. While HRA is a major complication of hemodialysis, the severity and symptoms will get worse over time. Early diagnosis and using a high-flux dialyzer could be a therapeutic modality to eliminate serum $\beta 2$ microglobulin and prevent pathological progression.

Declaration of patient consent

The authors certify that the patient has obtained appropriate patient consent form. In the form, the patient has given the consent for her images and other clinical information to be reported in the journal. The patient understands that her name and initial will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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