Bad taste as symptom at onset of IgM-related AL amyloidosis

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1 INTRODUCTION

An IgM clone is responsible for light chain amyloidosis (AL) in 4 to 7% of cases. While up to 35% of patients manifest polyneuropathies, cranial neuropathies and taste disturbances are rare.

We describe a patient with taste changes and oral cavity sensory symptoms as clinical onset of IgM-related AL in Waldenström macroglobulinemia (WM).

2 MATERIALS AND METHODS

A 68-year-old man presented with dysgeusia, sensory alteration in tongue, gingiva and lips since 2021. In 2023 four-limb paresthesias and distal weakness occurred, associated with mild weight loss.

He was in hematological follow-up for an IgM-k monoclonal gammopathy. Neurological examination revealed hypoesthesia of the anterior half of the tongue, gingiva and lips, bilateral stepping gait, distal limbs dysesthesias and areflexia. The patient needed support to walk (INCAT 2).

A wide diagnostic workup was performed.

3 RESULTS

Laboratory tests showed an IgM/k monoclonal immunoglobulin (9.30 g/L), increased pro-B-type natriuretic peptide, serum free light chain ratio of 15.14 mg/L, proteinuria with negative Bence-Jones. Bone marrow biopsy showed WM. Neurophysiological evaluation detected a sensory-motor axonal polyneuropathy (left fibular nerve DML 3.3 ms, MCV 43 m/s, cMAP amplitude 1.6 mV; right fibular nerve DML 3.3 ms, MCV 44 m/s, cMAP amplitude 2.2 mV; left sural nerve SCV 45 m/s, SNAP 2.7 uV) and larger CSA at ultrasound of left median and ulnar nerve, bilateral sciatic and tibial nerve (Figure 2). *Transthyretin* gene mutations tested negative. Echocardiography and cardiac MRI disclosed a left ventricular hypertrophy. Total-body CT scan revealed bilateral pleural effusion. Periumbilical fat and gum biopsies were consistent with AL kappa amyloidosis (Figure 1).

Treatment with rituximab, dexamethasone and cyclophosphamide had minimal efficacy, so rituximab-bendamustine was started with good hematological (undetectable monoclonal component and Bence Jones proteinuria) and neurological response. Currently, the patient can walk independently (INCAT 1), reports a resolution of sensory symptoms at the hands (he resumed playing the guitar) despite oral cavity symptoms persist. The patient reported subjective improvement in taste perception confirmed at the Taste Bande Strips test which resulted normal.

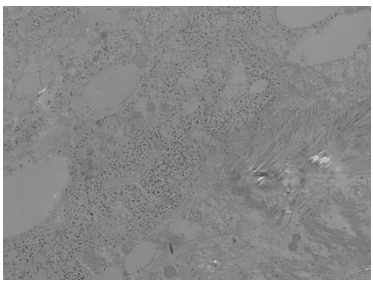
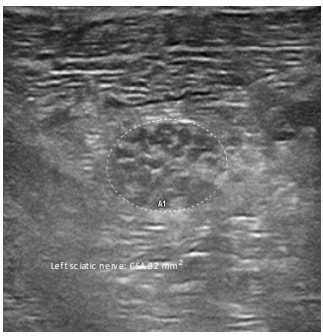


Figure 1. Gum biopsy: *immunolabeling positive for light chains Kappa by means of postembedding immunoelectron microscopy*



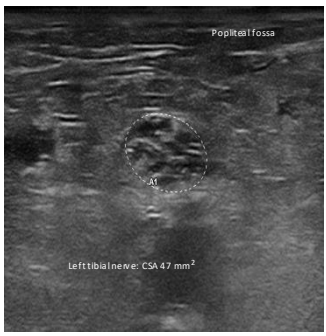


Figure 2. *Group images of larger CSA at nerve ultrasound*

4 CONCLUSION

Dysgeusia, though uncommon as an initial symptom, may represent an early clinical marker of AL amyloidosis. Amyloid recognition is important for a correct diagnosis and a proper therapeutic approach. Our patient had hematological and neurological improvement, but taste symptoms remained unchanged, as described in a previous patient (JNNP 1987;50:111-2).

Taste symptoms were the first to occur in our patient and the delayed therapy response may be due to the longer duration of the damage to the tongue, which is a common site of amyloid deposition.

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