Amyloidosis Presenting as a Mass Abutting the Tympanic Membrane

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Figure 1. Computed tomography of the temporal bone without contrast. Coronal image (A) shows a mass in the external ear canal abutting the tympanic membrane. Coronal (B) and axial (C) images of this mass indicate an anterior inferior erosion of the bony external auditory canal into the temporomandibular joint. (D) Noncontrast axial magnetic resonance image of the brain without contrast demonstrates left external canal lesion abutting the temporomandibular joint.

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An 80-year-old female presented to our tertiary care center with a 2-year history of unilateral aural fullness and hearing loss. On review of symptoms, she denied tinnitus, dizziness, trismus, and arthralgia of the temporomandibular joint (TMJ). Her past medical history was significant for end-stage renal disease (ESRD) secondary to polycystic kidney disease, for which she has been receiving dialysis for the past 10 years, in addition to hyperlipidemia and hypothyroidism. Her past surgical and social history were noncontributory.

Otomicroscopy revealed a tan-brown doughnut-shaped lesion on the anterior auditory canal adjacent to the tympanic membrane (TM) with a core of desquamated skin. The remainder of the canal and TM were normal without retraction, fluid, or obvious extension of the mass into the middle ear. Behavioral audiometry demonstrated bilateral mild symmetric high-frequency sensorineural hearing loss consistent with presbycusis. Noncontrast computerized tomography of the temporal bones showed a 5-mm soft tissue mass in the inferoanterior external ear canal adjacent to the TM with bone erosion into the left TMJ, confirmed on magnetic resonance imaging (Figure 1). An excisional biopsy was performed under local anesthesia and pathologic analysis confirmed amyloidosis (Figure 2). She has been followed conservatively without recurrence.

The head and neck region is a frequent site of presentation for systemic amyloid deposition with up to 20% of amyloidosis cases involving this region.1 Patients with systemic amyloidosis have been noted to have amyloid accumulation in the upper and lower respiratory tracts, parotid gland, tongue, larynx, and nasopharynx.1 While rare, amyloid may also present in the external auditory canal (EAC).2

Amyloidosis refers to diseases in which aberrant low-molecular-weight proteins fold improperly and aggregate into \( \beta \)-pleated sheets, the etiology of which is largely unknown.3 These sheets become insoluble and form fibrils that deposit extracellularly in tissues and organs. The location and severity of deposition is highly dependent on fibril type. To be considered amyloid, deposits must bind Congo red dye to show orange, yellow, or green birefringence under polarized light.3 Identification of specific amyloid proteins usually requires the use of mass spectrometry or immunochemistry staining.4

Amyloid diseases are classified into systemic, familial, or localized subtypes, the latter of which typically has a better prognosis.3,5 Signs of systemic amyloidosis include macroglossia, restrictive cardiomyopathy, carpal tunnel syndrome, periorbital purpura, hepatomegaly, and nephrotic syndrome.6 Systemic immunoglobulin light chain (AL-type) amyloidosis is often associated with diseases such as multiple myeloma or other plasma cell dyscrasias with renal or cardiac involvement. Systemic serum amyloid A-type amyloidosis is seen with chronic inflammatory states such as rheumatoid arthritis.1 Amyloidosis can also be related to hemodialysis (dialysis-related amyloidosis, DRA) and is more common in patients with ESRD with 10 or more years of dialysis. Dialysis-related amyloidosis results in deposition of amyloid protein in osteoarticular structures secondary to ESRD impairment of proximal tubular catabolism.4 Conversely, conditions such as primary localized cutaneous amyloidosis (PLCA) usually do not involve the aforementioned systemic symptoms. Primary localized cutaneous amyloidosis has 3 forms: lichen, macular, and nodular types.7 The nodular form of PLCA involves raised, non-itchy bumps that can be brown, red, or pink.5 The etiology is likely due to local infiltration of plasma cells into the dermis.7

To date, only 16 cases of amyloidosis involving the EAC have been reported, and of these, 8 were diagnosed with localized cutaneous amyloidosis lacking systemic symptoms.2,6-14 Only 2 were associated with long-term dialysis, as was potentially the case with our patient.5,9 However, no prior cases had documented bone erosion into the TMJ.2,8-14 There have been 3 reported cases of localized unilateral EAC amyloidosis, and Gheriani et al reported the only case that extended into the middle ear.8,12,13

While our patient’s renal status and history of dialysis were significant, she did not present with arrhythmia, cardiomyopathy, hepatomegaly, or osteoarticular dysfunction. Therefore, it is challenging to definitively classify the form of amyloidosis present in this case. While her past medical history of prolonged dialysis may suggest that the lesion is an unusual presentation of DRA, the lack of nonrenal accompanying osteoarticular dysfunction may suggest an unusual...

Figure 2. (A) Biopsy of the external auditory canal reveals amorphous eosinophilic deposits characteristic for amyloid deposition on hematoxylin and eosin (H&E) staining. Amorphous deposits weakly stained positively of Periodic Acid-Schiff (PAS) and Periodic Acid-Schiff-Diastase (PAS-D) with (B) apple green birefringence on Congo red staining under polarized light pathognomonic for amyloidosis.
location for nodular PLCA. At present, local resection is the most common treatment for nonsystemic amyloidosis of the EAC. Factors such as age, associated symptomatology, and comorbidities are crucial considerations that may guide management decision-making. Close observation and multidisciplinary management of systemic amyloidosis may be appropriate in many cases to monitor potential systemic progression. Additional data regarding nonsystemic amyloidosis of the EAC are warranted to establish a consensus regarding treatment of such conditions.

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