CASE STUDY

Amyloidosis of the External Auditory Canal

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Abstract Amyloidosis involving the ear is extremely rare. We present the case of a 76-year-old man who referred unilateral otorrhea and hypoacusis of six months’ duration. The external auditory canal (EAC) was narrowed by a subcutaneous nodule. After its extirpation, unilateral localised amyloidosis of the EAC was diagnosed. In these rare cases, it is crucial to rule out systemic amyloidosis.

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Introduction

Ear involvement in amyloidosis is extremely rare. Very few cases have been reported in the literature to date, 1-12 and only 3 of them were located in the external auditory canal (EAC). 10-12 We report a fourth case of amyloidosis located in the external auditory canal.

Clinical Case

The patient, a 76-year-old male, was referred to the ENT Department at the Hospital Clinic, in Barcelona, for the evaluation of left otorrhea and hearing loss of 6 months’ evolution. The only relevant medical history was a myocardial infarction.

On physical examination, the lumen of the left external auditory canal was reduced due to the presence of a strong, irregular subcutaneous nodule (1 cm in maximum dimension). The surface of the skin was erythematous and focally ulcerated. Computed tomography (Fig. 1) showed left EAC occupation by tissue, with soft tissue density and without bone involvement.
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Figure 1  CT scan of the temporal bone showing a partial occupation of the left EAC, with soft tissue density and without bone involvement.

The lesion was excised under general anaesthesia through a transcanal approach. The histopathological examination revealed diffuse infiltration by eosinophilic material in the dermis, which surrounded the skin appendages (Fig. 2A) and was associated with a discrete inflammatory infiltrate. The material became selectively dyed with Congo red (Fig. 2B and C) and showed apple-green birefringence when examined under polarised light (Fig. 2C). With these findings, we established the diagnosis of amyloidosis.

We carried out immunohistochemical studies of the tissue to evaluate the nature of the amyloid further. These were intensely positive for the immunoglobulin Kappa light chains and slightly so for Lambda chains (Fig. 2D). Tests for amyloid A, transthyretin, lysozyme, apolipoprotein AI, β2 microglobulin, fibrinogen, and cytokeratin 5 were negative.

Neither monoclonal gammopathy nor any data suggestive of systemic amyloidosis were found in subsequent clinical examinations. With these results, the patient was diagnosed with amyloid light-chain (AL) amyloidosis localised in the EAC. The patient is currently asymptomatic, 1 year after the intervention.

Discussion

Amyloidosis represents a spectrum of lesions characterised by the deposition of amorphous hyaline material. It has specific histochemical properties that lead it to become selectively stained with Congo red and show apple-green birefringence when viewed under polarised light. At the ultrastructural level, the amyloid is composed of highly regular fibril bundles that reflect its tertiary structure, composed of protein fibres in a cross-beta sheet configuration. Although all types of amyloid have very similar fibril structures, each type is derived from different source proteins: immunoglobulin light chain (AL amyloidosis), serum amyloid A protein (AA amyloidosis), β2-microglobulin (amyloidosis associated to haemodialysis), transthyretin (familial...
amyloidosis, and keratin (localised cutaneous amyloidosis). Amyloidosis can occur throughout the entire body, but each type affects certain organs preferentially. The head and neck regions are primarily affected by AL amyloidosis, especially in its localised form, and less often by the systemic form. The most common sites of involvement are the larynx, nasopharynx, nasal cavity, parotid, and tongue.11,13

To our knowledge, there are only 3 reported cases of amyloidosis located exclusively in the EAC,10-12 while there are a few more of amyloidosis with auricular shell involvement.2,4-7 There are 3 cases of bilateral amyloid deposition in the EAC in patients affected by multiple myeloma,16,8 and 1 case of amyloidosis related with dialysis.9 In cases of primary involvement of the EAC, the reason for consultation was hearing loss. Exploration revealed a mass, mostly in the EAC cartilage, in the form of a yellowish papule or macula, not ulcerated and generally asymptomatic, although occasionally producing pruritus.10-12 CT findings were available for only 1 of these cases, indicating a mass without bone erosion. Cases of amyloid deposition associated with multiple myeloma also started with hearing loss, although this was bilateral and no bone involvement was observed.

In general, surgical excision is the treatment of choice in localised amyloidosis. Studies reported in the literature on the treatment of amyloidosis in the upper airway showed that the most effective treatment was complete CO2 resection, with which the authors obtained disease-free follow-up of 14 years.14,15 Furthermore, localised radiation therapy is contraindicated because the results are poor and it shows a high rate of secondary effects.16 On the other hand, there is no effective specific treatment in systemic amyloidosis.

In localised amyloidosis, regardless of the type of amyloid, deposits form near the place where the precursor protein (which is abnormal in structure and/or local concentration) is produced. Precursor proteins have to be present for the formation of the fibrils, but the specific factors that determine why they are deposited in certain patients and organs are not well known.17,18 In our case, the presence of an accompanying mild chronic inflammation would be the only possible triggering factor.

For diagnosis, it is important to note that the most cases of amyloidosis located in the head and neck are of the AL type. However, immunohistochemical stains for light chains show a restriction of chains only occasionally.17,18 Therefore, it is recommended to use a panel of specific antibodies for all types of amyloid precursors, as well as to search specifically for an abnormal plasma cell population at the site of deposit. Moreover, a study of monoclonal immunoglobulin production in patient serum must also be carried out.

References