Amyloidosis of the external auditory canal and middle ear: Unusual ear tumor

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Abstract
Amyloidosis of the ear is rare. We describe the case of a 41-year-old man who had localized amyloidosis that involved the external auditory canal and middle ear. To the best of our knowledge, only 4 other cases of amyloidosis involving the external auditory canal have been previously reported; in none of these cases was the middle ear involved. We also discuss the clinical importance of this condition and its treatment.

Introduction

Many cases of cutaneous amyloidosis have been reported in the literature, but to the best of our knowledge, only 4 cases have involved the external auditory canal.[1-4] In this article, we report a new case. Moreover, as far as we know, ours is the first reported case of amyloidosis that also involved the middle ear.

Case report

A 41-year-old man who worked as a roofer was referred to us by his general practitioner for evaluation of a 3-month history of right ear irritation, discomfort, and hearing loss. The patient had no other ear complaint.

Examination of the right ear revealed that a polypoid mass had filled the external auditory canal. Debris and keratin deposition were also present. Pure-tone audiometry showed a right-sided mixed hearing loss and a bilateral and symmetrical decrease in high-frequency sensorineural threshold that was consistent with noise-induced hearing loss.

The patient was administered general anesthesia, and the polyp was removed. The tympanic membrane was intact. The excised specimen was sent for histologic analysis. Hematoxylin and eosin (H&E) staining revealed a diffuse replacement of the dermal subepithelial connective tissues by extracellular, amorphous eosinophilic material (figure 1). Congo red staining showed birefringence under polarized light, characteristic of amyloidosis. Based on these findings, computed tomography (CT) of the temporal bone was obtained (figure 2). CT revealed the presence of a soft-tissue mass that involved the external auditory canal; no evidence of bony erosion was seen. CT also showed that abnormal soft tissue occupied the area around the bony ossicles of the right middle ear. The density of this tissue was similar to that of the abnormal soft-tissue mass that involved the external auditory canal.

The patient was treated with regular cleansing and debridement of the ear canal. Because of the nature of his job as a roofer, which required significant reliance on the vestibular system, we decided against exploring his right middle ear to obtain tissues for histologic analysis lest we provoke postoperative dizziness or tinnitus.

We also looked for evidence of systemic disease by obtaining various hematologic and imaging studies, including a full blood count, renal and liver function tests, serum immunoglobulin assays, protein electrophoresis, electrocardiography, chest x-ray, and ultrasonography of the abdomen. All findings were normal. Therefore, we declared a diagnosis of primary amyloidosis of the right external auditory canal and middle ear. Fourteen months after diagnosis, the disease remained confined to the right ear.

Discussion

Amyloidosis is not a single disease entity but a spectrum of diseases that have in common the extracellular deposition of insoluble protein fibrils in tissue or organs in a beta-pleated sheet configuration. These protein fibrils are diverse and unrelated, but they all produce amyloid deposits with a common beta fibrillar structure.[5] These protein subunits are derived from normally occurring serum proteins, such as light-chain (AL) immunoglobulins lambda and kappa, the acute-phase reactant protein A, prealbumin, transthyre-

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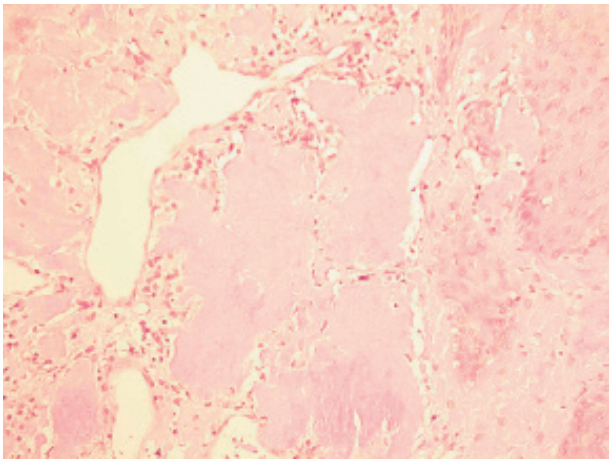

Figure 1. H&E staining shows the diffuse subepithelial replacement of connective tissue by amorphous, extracellular eosinophilic deposits.

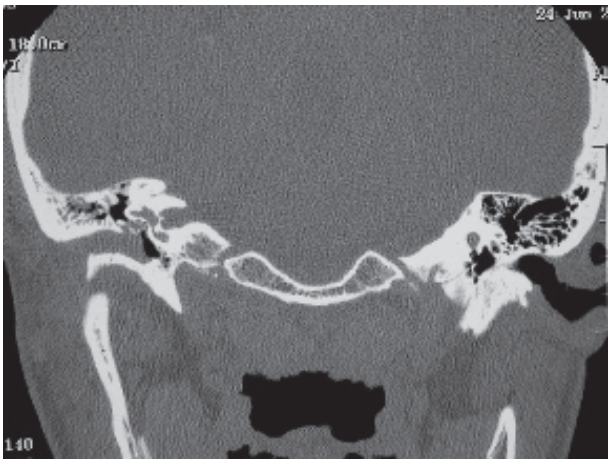

Figure 2. CT of the temporal bone shows the soft-tissue mass involving the right external auditory canal and middle ear.

tin, and other rare types (table). The reason these fibrils accumulate is not fully understood, although the process appears to involve a chronically elevated serum precursor level coinciding with a predilection, possibly genetic, for amyloid deposition.[5]

Over the years, amyloidosis has been classified in several different ways. First, amyloidosis is classified as localized or systemic. Systemic disease is subclassified according to the nature of the precursor plasma proteins that form the fibril deposits (table).

The diagnosis of amyloidosis is based on clinical suspicion and confirmed by identifying tissue that is positive for amyloid deposition.[5,6] Treatment is directed at the underlying cause, so the choice of drug varies.[7] Local excision is usually sufficient for the treatment of the localized form.[8] Close observation is necessary because of the possibility of local reccurence.[9] Unfortunately, it is difficult to mobilize amyloid deposits once they have developed. A new drug, anthracycline iododoxorubicin, has been shown to bind to AL amyloid in vivo and to promote resorption.[10]

Table. Classification of systemic amyloidosis

AL (light-chain immunoglobulin)	
	Plasma-cell/immunocyte dyscrasia
	Multiple myeloma
AA (serum protein A)	
	Acute-phase protein produced in response to inflammation
	Rheumatoid arthritis, inflammatory bowel disease, osteomyelitis, tuberculosis, familial Mediterranean fever
	Gastric carcinoma, renal cell carcinoma, Hodgkin's disease
Aß2M ($ß_2$ microglobulin)	
	Chronic hemodialysis
Familial	
	AF (prealbumin protein)
	ATTR (transthyretin protein)
	Other rare familial types

The prognosis for patients with amyloidosis is variable and dependent on the underlying cause. Patients with the localized form have a good prognosis.[8] The worst prognosis is for patients with systemic amyloidosis secondary to AL amyloid deposition; death within 1 to 2 years is common.[11] Death from systemic amyloidosis is usually caused by cardiac involvement, particularly cardiac arrhythmia and heart failure.

The localization of amyloid deposition specifically to the ear canal has raised the suggestion that physical or chemical factors, such as tissue pH or hydration forces on cellular surfaces, might be responsible.[11,12] However, the rarity of this condition in the ear makes it difficult to study this theory. The treatment of ear amyloidosis should include local excision and debridement to prevent irritation, deafness, and secondary infection. Topical steroid therapy can also help limit the local inflammatory process.[1,3,4] The possibility of an underlying cause should always be considered and treated as soon as possible.

References

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