Unilateral Isolated Primary Cutaneous Amyloidosis of the External Auditory Canal

Giuseppe Magliulo, Ludovica de Vincentiis, Annalisa Pace, Irene Claudia Visconti, Francesco Le Foche, Mara Riminucci, Alessandro Corsi

INTRODUCTION
Primary cutaneous amyloidosis (PCA) refers to the deposition of amyloid either at the dermal–epidermal junction or within the dermis in otherwise normal skin and in the absence of any underlying systemic illness [1]. Three main clinico-pathological types have been distinguished. Lichen amyloidosis (LA) and macular amyloidosis (MA) commonly occur sporadically and can concur (biphasic amyloidosis) in the same patient [1]. Because LA and MA have been related to keratinocytes' degeneration and High Molecular Weight-Cytokeratins (HMW-CKs) have been demonstrated within the amyloid deposits, they are considered variants of the same disease and classified as keratin-type amyloidosis [1]. Conversely, nodular amyloidosis is due to the deposition of immunoglobulin light-chains, associated with plasma cell infiltration, and, sometimes, may progress to systemic disease [1, 2].

External ear skin involvement in PCA is uncommon and typically not associated with other concurrent skin lesions [3]. It presents with normochromic, yellowish-whitish or erythematous, usually pruritic, either unilateral or bilateral, variably sized papules mainly in the auricular concha [3]. Because isolated PCA of the external auditory canal (EAC) has been very rarely described [2-10], we report here a patient with unilateral localized PCA of the left EAC and review the pertinent literature.

CASE PRESENTATION
A 65-year-old female presented for itching within the left EAC. She denied any history of local trauma and was not under dialytic treatment. Her history was negative. Otoscopy revealed a smooth, firm, white papule, 3 mm in maximum dimension, in the cartilaginous portion of the left EAC. The lesion was excised. Histologic examination revealed nodular deposits of an amorphous, eosinophilic, and homogeneous material in the papillary dermis (Figures 1a and 1b). The deposits contained some clefts and were partially encircled by epidermal collarettes. The overlying epidermis was thin and hyperkeratotic. Inflammatory cells and melanophages were absent. The deposits stained with Congo Red and, under polarized light, revealed the apple green birefringence characteristic of amyloid (Figure 1c). In addition,
The diagnosis of amyloidosis of the external ear may cause concern for physicians for raising the possibility of underlying systemic amyloidosis. The demonstration of HMW-Cks in the amyloid deposits could represent a simple and reproducible tool to establish their epidermal origin and exclude an underlying associated condition such as plasma cell dyscrasia or chronic disease with secondary involvement of external ear. As a consequence, it seems reasonable that physicians do not need to pursue extensive systemic clinical workup for these patients, except dermatologic ex-
amination since, even though rarely, amyloidosis of the external ear can concur with the involvement of other skin regions in the context of LA and MA[2, 15]. However, because of its rarity, further cases should be examined to exclude definitely a relationship between amyloidosis of the EAC, and more in general of the external ear, and systemic amyloidosis.

Finally, on the basis of the data from the literature, resection is the treatment of choice for isolated PCA of the EAC[3-5, 7, 9, 10]. In the cases in which follow-up was available[3, 9], recurrences have not been reported.

CONCLUSION
We describe a case of unilateral localized keratinic PCA of the EAC. The clinico-pathologic findings observed in our case, in combination with those described in the other few cases reported in the literature, support the absence of an association with systemic disease and a role of local chronic stimulation/irritation in its pathogenesis.

Informed Consent: Written informed consent was obtained from the patient who participated in this study.

Peer-review: Externally peer-reviewed.

Author Contributions: Concept - G.M., A.C.; Design - G.M., A.C.; Supervision - G.M., A.C.; Data Collection and/or Processing - G.M., L.d.V., A.P., I.C.V., F.L.F.; Analysis and/or Interpretation - G.M., L.d.V., A.P., I.C.V., F.L.F., M.R., A.C.; Literature Search - L.d.V., A.P., I.C.V.; Writing - L.d.V., A.C.; Critical Reviews - G.M., M.R., A.C.

Conflict of Interest: The authors have no conflict of interest to declare.

Financial Disclosure: The authors declared that this study has received no financial support.

REFERENCES
1. Schreml S, Szeimies RM, Vogt T, Landthaler M, Schroeder J, Babilas P. Cutaneous amyloidoses and systemic amyloidoses with cutaneous involvement. Eur J Dermatol 2010; 20: 152-60. [Crossref]
2. Wenson SF, Jessup CJ, Johnson MM, Cohen LM, Mahmooodi M. Primary cutaneous amyloidosis of the external ear: a clinicopathological and immunohistochemical study of 17 cases. J Cutan Pathol 2012; 39: 263-9. [Crossref]
3. Panarese A, Roland NJ, Green B. Primary amyloidosis of the external auditory canal: case report. J Laryngol Otol 1994; 108: 49-50. [Crossref]
4. Zundel RS, Pyle GM, Volytovich M. Head and neck manifestations of amyloidosis. Otolaryngol Head Neck Surg 1999; 120: 533-7.
5. Shintani T, Momoshima N, Himi T. Amyloidosis of the bilateral external auditory canal [in Japanese]. Otolaryngol Head Neck Surg (Tokyo) 2001; 73: 494-549.
6. Ashimori N, Hayashi Y, Hoshino T. Two cases of amyloidosis of the external auditory canals [in Japanese]. Pract Otorhinolaryngol (Basel) 2003; 96: 1049-54. [Crossref]
7. Mozos A, Caballero M, Sole M, Cardesa A. Amyloidosis of the external auditory canal. Acta Otorrinolaringol Esp 2011; 62: 392-4. [Crossref]
8. Park K, Taniyama T, Kabashima K, Miyachi Y. Primary cutaneous lichen amyloidosis of the external auditory canal, possibly due to scratching with a metal nail head for severe pruritus. Eur J Dermatol 2014; 24: 95-6. [Crossref]
9. Wasano K, Saito H, Kanzaki S, Ogawa K. Keratinic amyloidosis of the external auditory canal. Auris Nasus Larynx 2014; 41: 97-100. [Crossref]
10. Shiiya C, Watanabe K, Ota M. Image Gallery: Lichen amyloidosis on the external auditory canal. Br J Dermatol 2016; 175: e149. [Crossref]