CASE REPORT

Primary Cutaneous Amyloidosis of the auricular concha: Two case report

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ABSTRACT

Primary cutaneous amyloidosis of the auricular concha is considered as an entity of unknown etiology and a rare variant of primary localized cutaneous amyloidosis (PLCA) that predominates among women in the fourth and seventh life decade. There are a few clinical and dermoscopic reports in medical literature. However, a characteristic patron composed by globular structures has been identified. These structures, which vary in size, are localized on a brown or pink network referred to as cobblestone pattern.

The following article introduces two cases of female patients that presented with hyperpigmented papules on the auricular region. These were clinically diagnosed as cutaneous amyloidosis of the auricular concha and confirmed through histopathology study. No other body parts manifested PLCA.

INTRODUCTION

The cutaneous amyloid deposit with no systemic involvement is recognized as primary localized cutaneous amyloidosis (PLCA).[1,2] Medical literature recognizes 4 types of PLCA: Lichenoid, macular, nodular and familiar. Auricular amyloidosis is an unusual type of PCA that lies on the auricular concha or external auditory canal.[3,4]

Amyloidosis was first described in 1983 by Sanchez and cols. Up to date, there is no consensus about its actual origin, given that some authors consider this condition as a new entity of cutaneous amyloidosis, and some others voice that it is a subtype of lichen amyloidosis.[5] Medical literature presents very few reports of this condition, and little to know dermoscopic reports.

CLINICAL CASES

Case 1

47-year-old female patient with history of melasma in the middle portion of the face presents with multiple clustered hyperpigmented, well-delimited papules of smooth surface located on the auricular concha, left helix (Figure 1) and right side (minor presence), measuring 1-2mm in diameter, leaving eroded areas associated with constant manipulation. There are no accompanying symptoms. Dermoscopy revealed well-delimited lesions, brown and erythematous globular lesions, as well as a small whitish central area. Such globules come together to form a cobblestone pattern. Histopathology showed epidermis with atrophy foci within elongated interpapillary ridges surrounding the

subjacent papillae, along with necrotic keratinocytes, some eliminated through the stratum corneum (Figure 2). The dermis showed superficial perivascular infiltrate and a presence of melanophagi and melanin pigment in the interstitium, along with amyloid material deposit, making this lesion compatible with amyloidosis of the auricular concha. Curettage was advised as treatment.

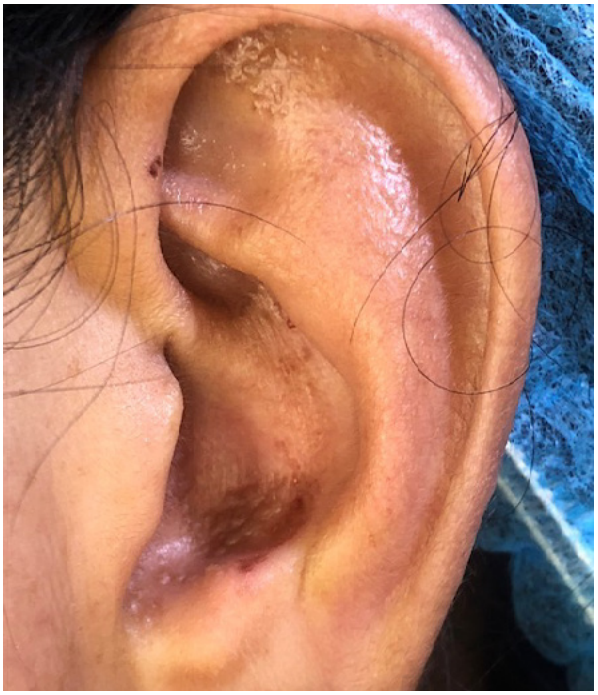

Figure 1: Hyperpigmented papules with eroded surface on auricular concha and left helix.

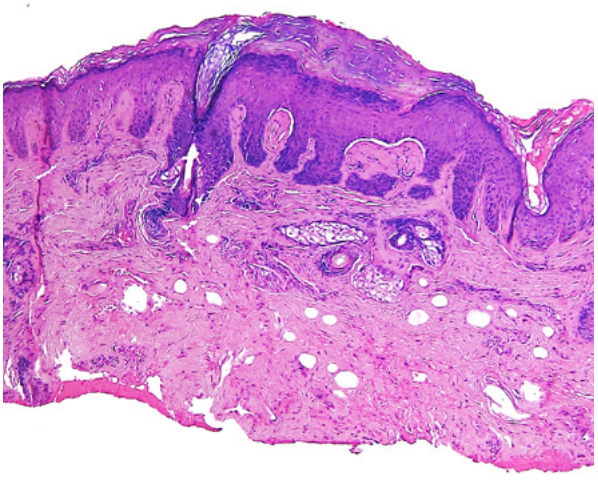

Figure 2: Histopathology evidences epidermis with atrophy foci within elongated interpapillary ridges surrounding the subjacent papillae, along with necrotic keratinocytes, some eliminated through the stratum corneum. The dermis manifests melanophagi and melanin pigment in the interstitium and amyloid deposit.

Case 2

68-year-old female patient presented with brown hyperpigmented papules (20-year-evolution), initially desquamative and located on the left auricular concha (Figure 3). Dermoscopy evidences a brown pigment network with 1mm globules of solid consistency on the left auricular concha, and others at the level of the antitragus, with a whitish central axis. Histopathology examination revealed hyperkeratosis, irregular acanthosis, dense fibrosis of papillary dermis, focal neovascularization and mild perivascular mononuclear inflammatory infiltrate. The papillary dermis also manifests deposits of dense hyaline globules in clusters, confirming a cutaneous amyloidosis diagnosis on the auricular concha (Figure 4).

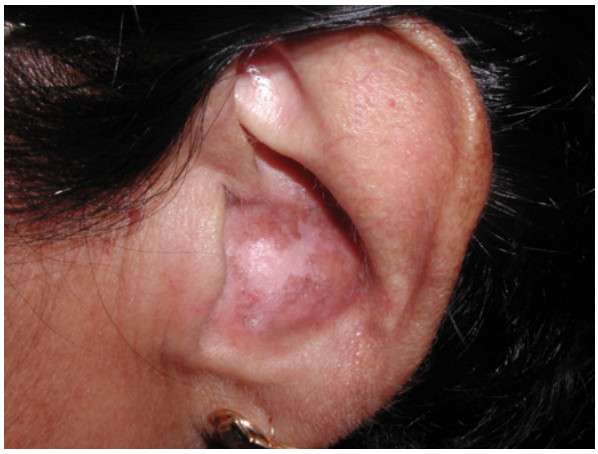

Figure 3: Brown hyperpigmented papules on left auricular concha.

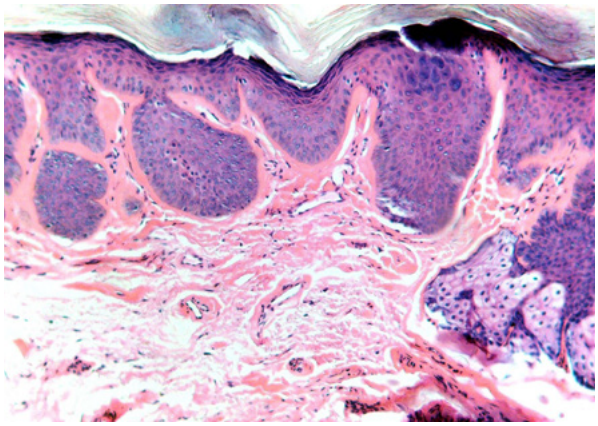

Figure 4: Hyperkeratosis, irregular acanthosis, dense fibrosis of papillary dermis, focal neovascularization and mild perivascular mononuclear inflammatory infiltrate. Papillary dermis also reveals an accumulation of dense hyaline globule deposits.

DISCUSSION

Thus far, there are thirty two case reports of PCLA located on the auricular concha.[3-12] The first report was completed by Sánchez J. in 1983, who described 4 cases of patients with hard, non-painful papules located symmetrically on both auricular conchae, which he named as "auricular collagenous papules". Nonetheless, in 1988, Hicks recognized the condition as primary cutaneous amyloidosis on the auricular concha because of its hyaline content in the papillary dermis and the presence of intermediate microfilaments in the electronic microscopy.[6]

This condition is a rare entity. As a consequence, the prevalence of PCLA in this particular anatomical area is unknown. However, it generally manifests in Hispanic, Asian and Middle Eastern descendants.[13,14] It predominates in females within the fourth and seventh decade of life.[6,11] Its etiology is unknown, but there is a great correlation between friction and scratching. It's even associated to photoallergic reaction, eczema, dermographism, lichen planus or scabies.[13,15] The deposit is generated by necrotic keranocytes that are liberated in the dermis, where cytokeratin 5 predominates.[8]

Clinically, this condition is presented as papules or yellow, waxy, nacarated patches of regular margins. Unlike on the classic lichen amyloid, they are not pruriginous.[11,12] They are usually bilateral, sometimes desquamative and they are not associated to systemic amyloidosis; although there is a report about an unusual presentation of systemic amyloidosis along with amyloidosis of the auricular concha in a patient with familial Mediterranean fever.[10,16]

Dermoscopy is a useful tool that can help physicians confirm a clinical suspicion regarding cutaneous amyloidosis.[14] There are multiple dermoscopic reports of PCLA. However, the only amyloidosis dermoscopic report on auricular concha was completed by Zhou X, Chen Q and Tian X. It was about a 50-year-old patient that presented with whitish globular structures of variable sizes on a brown and pink network, acknowledged as cobblestone pattern.[12] In contrast to the aforementioned report, dermoscopy of the first case is a lot similar to the one described about lichen amyloid with papules presenting a whitish center surrounded by a halo composed of brown spots simulating a volcanic crater (Figure 5).[14] Multiple hyperpigmented globules are observed on a pink background. However, they do not form a cobblestone pattern. There are some eroded areas due to previous manipulation.

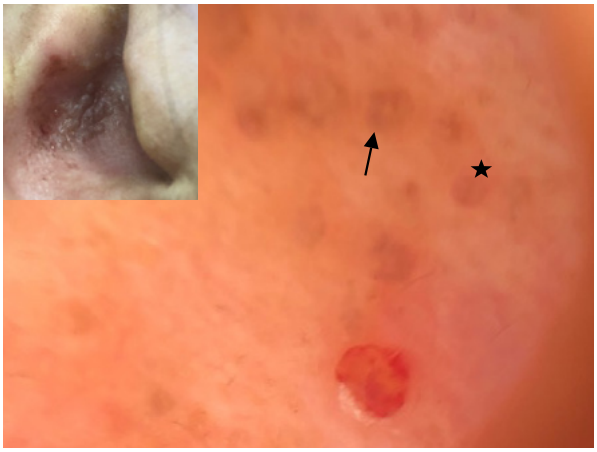

Figure 5. Papules with a whitish center surrounded by a hyperpigmented halo simulating a volcanic crater (arrow), hyperpigmented globule on a pink background (star).

Among histopathology findings, there are hyperkeratosis and deposits of amorphous, homogenous and eosinophilic material on the papillary dermis and atrophic epidermis, where material amyloid can be found as well.[11,12] The stain that is employed to detect amyloid is violet crystal and Congo red. The PAS stain (periodic acid-Shiff), which shows birefringence in polarized light, can also be employed.[11]

Other sorts of differential diagnosis must be considered: seborrheic keratosis, verrucae, helomas, basal cell carcinoma, adnexal carcinoma and erythematous lupus.[11] For patients who require aesthetic treatment, the advised procedures are curettage, excision or electrocoagulation.[11] More invasive modalities have been used in lichen amyloidosis, such as CO2 LASER, Q switched and Nd: YAG, which must be employed as treatment options.[10]

CONCLUSION

Amyloidosis on the auricular concha is a rare presentation of PCLA, which requires a high index of suspicion. Consequently, dermoscopy and biospy enable physicians to rule out other diagnoses. Dermoscopic findings regarding this location are the cobblestone pattern, along with an aspect similar to a volcanic crater, previously described in lichen amyloidosis cases. Nonetheless, dermoscopic reports about this entity must be researched in order to define actual dermoscopic patterns.

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