

Lichen Amyloidosis of the External Ear

Dear Editor,

We report cases of lichen amyloidosis of the external ear, an uncommon type of primary cutaneous amyloidosis, in two women.

Patient 1 [Figure 1] and Patient 2 [Figure 2], both women in their 40s, presented with multiple asymptomatic skin lesions on bilateral ears, noticed one year and four years back, respectively. Physical examination showed multiple closely aggregated hyperpigmented dome-shaped keratotic papules of size 2–4 mm present symmetrically on the conchae of bilateral ears, in both the patients. In addition, patient 1 had ill-defined areas of asymptomatic rippled hyperpigmentation suggestive of macular amyloidosis on the upper back, while patient 2 had pruritic hyperpigmented keratotic papules on bilateral shins suggestive of lichen amyloidosis. Punch biopsies were taken from the conchal papules in both the patients, which showed eosinophilic amorphous fissured aggregates distending the dermal papilla and mild pigment incontinence, with overlying acanthotic epidermis. The eosinophilic deposits stained orangish red with Congo red and demonstrated an apple-green birefringence under polarized light, confirming the diagnosis of cutaneous amyloidosis. Patient 1 did not seek any active intervention after being counselled about the disease, while patient

2 was prescribed a combination of topical steroids and keratolytics with mild improvement.

Amyloidosis of the external ear is an uncommon but distinct variant of primary cutaneous amyloidosis that affects the external ear, first erroneously described by Sanchez *et al.* as collagenous papules on the auricular conchae.^[1] Subsequently, Hicks *et al.* reported four cases and identified the deposits to be of amyloid origin.^[2] Clinically, it presents as largely asymptomatic grouped keratotic or waxy papules in a cobblestoned pattern on the external ear unilaterally or bilaterally, usually on the concha, but other sites such as auditory canal or triangular fossa may also be affected.^[3] Middle-aged women seem to be preferentially affected. Though concurrent macular or lichen amyloidosis at other sites is usually absent in these patients,^[3] both of our patients had primary cutaneous amyloidosis at other areas, which has also been occasionally described.^[4]

There is no consensus on whether it is a distinct type of primary cutaneous amyloidosis or a type of lichen amyloidosis uniquely affecting the external ear. Wenson *et al.*, who reported a large series of this entity in 17 patients, considered it to be a separate entity from lichen amyloidosis due to lack of pruritus or the presence of primary cutaneous amyloidosis at other body sites.^[3] Others have regarded it as a variant of lichen amyloidosis

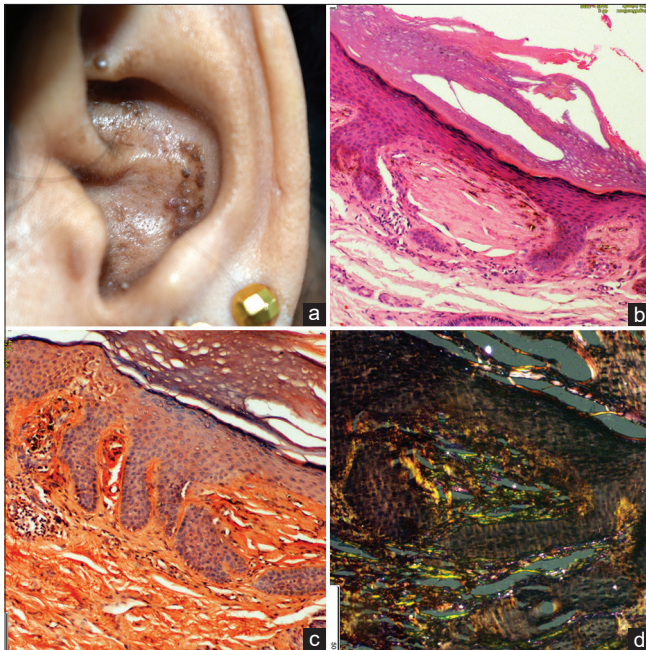


Figure 1: (a) Multiple closely aggregated hyperpigmented dome-shaped keratotic papules present on the ear conchae. (b) Eosinophilic amorphous fissured aggregates distending the dermal papilla (H and E; 200×). (c) Deposits stained orangish red with Congo red (400×). (d) Apple green birefringence under polarized light (400×)

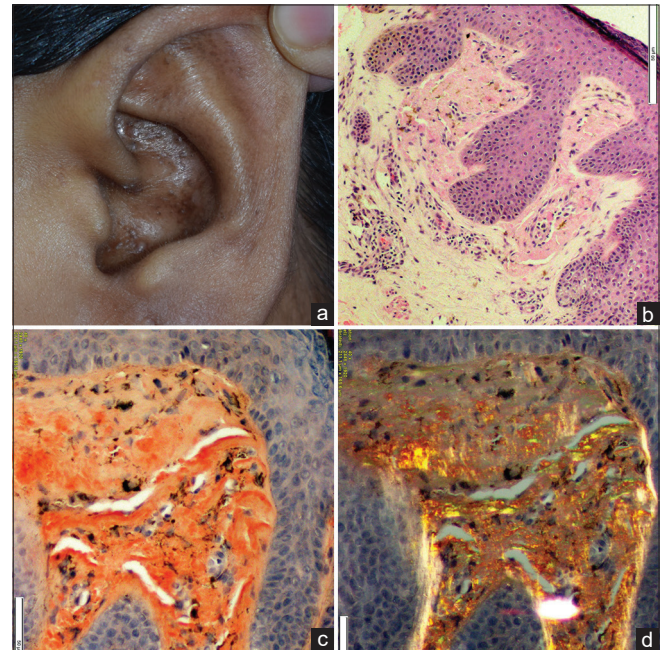


Figure 2: (a) Multiple closely aggregated hyperpigmented dome-shaped keratotic papules present on the ear conchae. (b) Eosinophilic amorphous fissured aggregates distending the dermal papilla (H and E; 200×). (c) Deposits stained orangish red with Congo red (400×). (d) Apple green birefringence under polarized light (400×)

due to clinical and histopathological similarities. Further, immunohistochemical positivity of the amyloid deposits for CK34BE12, a monoclonal antibody against high-molecular-weight keratins including cytokeratins such as CK 1, 5, 10, and 14, has been demonstrated, suggesting an epidermal origin, similar to macular and lichen amyloidosis.

Treatment options include topical keratolytic agents with mild to moderate topical corticosteroids and various physical modalities such as curettage and radiofrequency ablation.^[5]

Lichen amyloidosis of the external ear is a distinctive type of primary cutaneous amyloidosis without any systemic involvement. Differential diagnosis, when isolated conchal papules are present, includes discoid lupus erythematosus, adnexal neoplasms, and seborrheic keratosis.^[5] However, the presence of multiple closely grouped keratotic papules in the concha with the absence of follicular plugging may be a useful clue. Through this report, we wish to increase awareness about this uncommon benign condition.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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