

# Primary cutaneous amyloidosis of the external ear: a clinicopathological and immunohistochemical study of 17 cases

Primary cutaneous amyloidosis includes several forms of localized amyloidosis characterized by superficial amyloid deposits occurring at or near the dermal–epidermal junction in the absence of systemic involvement. Primary cutaneous amyloidosis of the auricular concha and external ear represents a rarely described variant. There have been 27 cases reported in the English language literature, and herein we report 17 additional cases. This article demonstrates that the amyloid observed in this context is generally positive for Congo red, crystal violet and thioflavin T. It also expresses cytokeratin 34 $\beta$ E12 via immunohistochemistry. Our immunohistochemical results and review of the literature suggest that the amyloid in amyloidosis of the external ear is the result of basal keratinocyte degeneration and does not signify deposition from a systemic or generalized process.

**Keywords:** amyloid, amyloidosis, auricular, concha, ear

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Cutaneous amyloidosis refers to the extracellular deposition of amyloid material in the skin. Primary cutaneous amyloidosis comprises several forms of localized cutaneous amyloidosis characterized by superficial amyloid deposits occurring at or near the dermal–epidermal junction, and in the absence of systemic involvement.<sup>1</sup> Primary cutaneous amyloidosis of the auricular concha and external ear (hereafter referred to as amyloidosis of the external ear) is a rarely described variant of cutaneous amyloidosis affecting the external ear, including the pinna and external auditory canal. There have been 27 cases of amyloidosis of the external ear reported in the literature (Table 1).<sup>2–14</sup> However, reports to date have not been helpful in elucidating the etiology or clinicopathological significance of this entity. Specifically, there has been

no substantial correlative data supporting or negating the possible association of amyloidosis of the external ear with underlying conditions such as multiple myeloma, connective tissue disease or trauma. As a result, clinicians often have concerns regarding whether any further work-up or investigation of underlying malignancy or disease is warranted.

In an attempt to better define the etiology and clinical significance of this condition, we examined 17 cases of primary cutaneous amyloidosis of the auricular concha and external ear diagnosed at Caris Life Sciences over a 6-year period (2004–2010). We examined both the cytochemical and immunohistochemical profiles of these lesions. In addition, using a brief questionnaire we further evaluated the patients' clinical presentations and applicable histories. To the best of our knowledge,