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Primary Localized Cutaneous Nodular Amyloidosis on Scalp Successfully Treated with Excision

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Primary localized cutaneous nodular amyloidosis (PLCNA) is the rarest form of cutaneous amyloidosis, characterized by nodular deposits of light chain amyloids in the dermis and subcutaneous tissue, without apparent systemic involvement. One or several nodules are preferably located on the extremities, trunk, or face. The most useful stain for detecting amyloid fibrils is Congo red, which, when combined with polarized light, makes amyloid proteins appear apple-green under a microscope. Immunohistochemical staining can help identify the exact type of amyloid proteins. Although the exact etiology of PLCNA is unclear, removal of nodules by shaving or surgical excision has shown good results. To the best of our knowledge, only seven cases of PLCNA have yet been reported in the Korean literature. In three of these cases, the patients had lesions on the scalp. Herein, we present a case of a 34-year-old male with PLCNA on the scalp with all the results of immunohistochemical evaluation.

Keywords: Amyloidosis, Primary localized cutaneous nodular amyloidosis

INTRODUCTION

Amyloid is a material deposited in the skin and other organs, and is eosinophilic, homogeneous, and hyaline in appearance. Primary localized cutaneous amyloidosis occurs when amyloid is deposited only in the skin, without evidence of systemic involvement[1]. Amyloidosis may be classified into three clinical types, namely, macular, lichenoid, and nodular amyloidosis[1-3]. Primary localized cutaneous nodular amyloidosis (PLCNA) is the rarest subtype. In contrast to macular and lichenoid amyloidosis, amyloid fibrils in PLCNA are believed to derive from immunoglobulin light chains produced by monoclonal plasma cells and not from degenerative keratinocytes[3,4].

One or several nodules are typically observed on the limbs, trunk, or face, and occasionally, elsewhere. The nodules are typically tumefactive and waxy, and commonly measure 1 to 3 cm in diameter[2]. To date, only seven cases of PLCNA have been reported in the Korean literature[5-11]. Herein, we report a case of PLCNA on the scalp, along with the results of immunohistochemical evaluation.

CASE REPORT

A 34-year-old Korean male visited our clinic complaining of two firm masses that had been present on his scalp for 7 years. Physical examination revealed salmon-colored, well-circumscribed waxy nodules on the occipital and right temporal areas (Fig. 1A). There was no subjective symptom or previous trauma history.

Biopsy specimens were obtained from the scalp lesions. The epidermis showed atrophic changes. Large deposits of amorphous, fissured, pale, eosinophilic materials were observed throughout the dermis into the subcutaneous fat. Perivascular and interstitial lymphocytic infiltrate admixed with plasma cells, mast cells, and a few eosinophils were observed throughout the dermis (Fig. 2A, B). These deposits were positively stained with Congo red (Fig. 2C). Under the diagnosis of cutaneous amyloidosis, systemic workup was performed to rule out

systemic involvement. Laboratory studies including complete blood cell count, blood chemistry, liver function tests, hepatitis viral markers, and urinalysis showed normal limits. Chest radiography and electrocardiogram showed no abnormality.

In addition to these results, to determine the exact type of amyloid fibrils deposited, immunohistochemical staining was performed using a panel of antibodies including kappa and lambda immunoglobulin light chains, and amyloid A. Immunoperoxidase staining showed positive reaction with both anti-kappa or anti-lambda light chain antibody in amyloid materials and infiltrating plasma cells (Fig. 3). Amyloid A immunostaining showed no response to amyloid materials. Immunohistochemical staining showed that the amyloid fibril was amyloid L (AL) protein, derived from polyclonal immunoglobulin light chain.

The scalp lesions of the patient were surgically removed by excision, and the patient remained stable without recurrence during a follow-up period of 18 months (Fig. 1B). We received the patient's consent form about publishing all photographic materials.

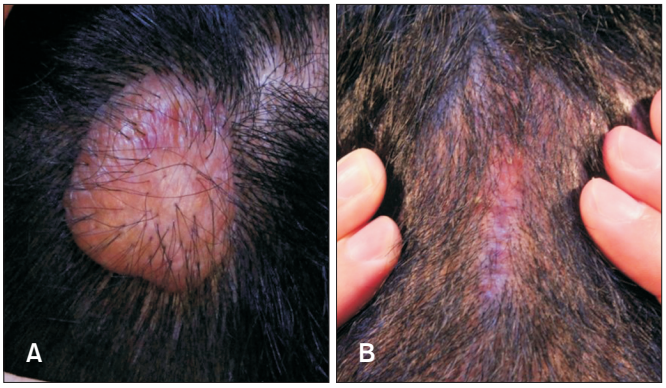

Fig. 1. Clinical features of the patient. (A) About 5 cm×3 cm sized, well-circumscribed, salmon-colored, waxy nodules on the occipital and right temporal scalp. (B) Occipital and temporal scalp without recurrence during a follow-up period of 18 months after surgical excision.

DISCUSSION

Amyloidosis is a disease characterized by the abnormal build-up of amyloid, abnormal non-branching fibrillary β-pleated sheet proteins, resulting in localized or systemic organ dys-

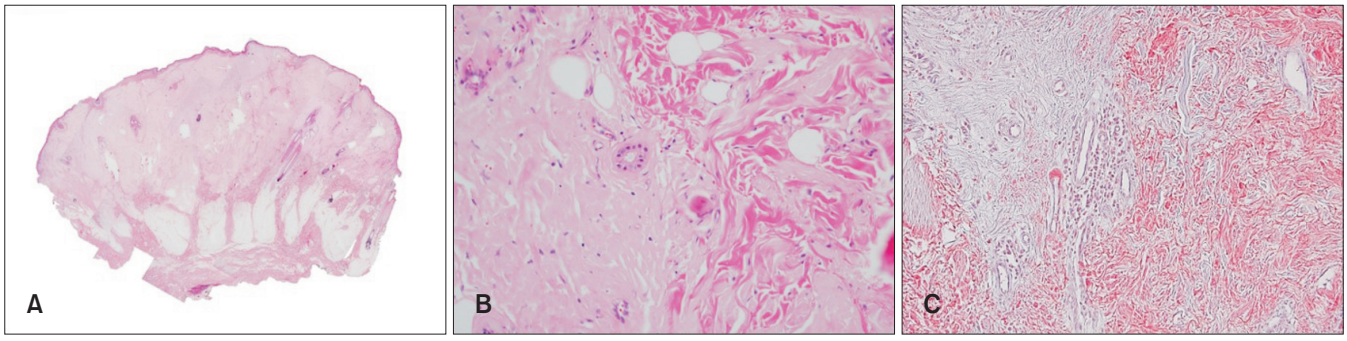

Fig. 2. (A) Deposition of massive amorphous, eosinophilic materials was observed throughout the entire dermis (H&E, ×40). (B) Amorphous, eosinophilic deposits can be seen next to normal dermal collagen (H&E, ×200). (C) The deposits were positively stained with Congo red (Congo red, ×200).

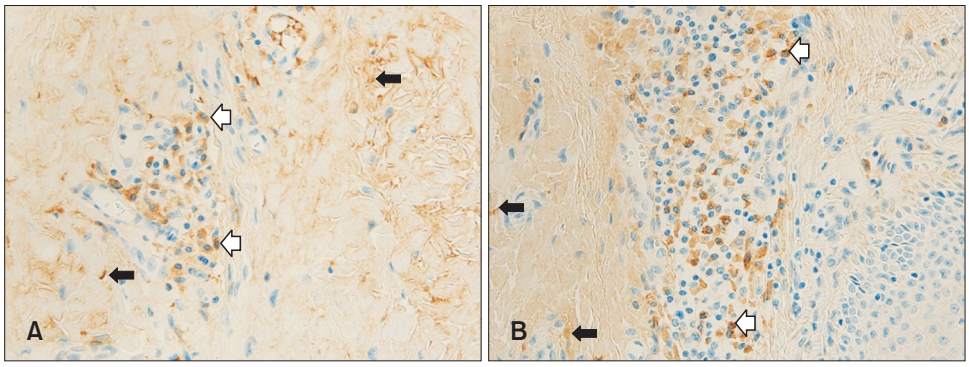

Fig. 3. Immunohistochemical staining with anti-kappa (A) and anti-lambda (B) antibody showed positive staining with both antibodies in the amyloid (black arrows) and the cytoplasm of the infiltrating plasma cells (white arrows) (×200).

function. Amyloidosis can be classified into systemic and localized, depending on the presence or absence of systemic involvement, and into primary and secondary, depending on the cause[1,10]. If primary localized amyloidosis affects the skin, the condition is termed as primary cutaneous amyloidosis.

Primary cutaneous amyloidosis may be subdivided into three clinical types, namely macular, lichenoid, and nodular amyloidosis[1-3]. PLCNA is the rarest among these subtypes. The mean age of patients with PLCNA is 55 years, and the prevalence does not differ between males and females[10,12,13]. PLCNA was first reported by Gottron[14] in 1950, and it was then referred to as amyloidosis cutis nodularis atrophicans diabetica.

Histopathologic examination showed flattened rete ridges and large deposits of amorphous, sometimes fissured, pale, eosinophilic amyloid material throughout the dermis into the subcutaneous fat. Amyloid can be detected using special stains such as periodic acid-Schiff, Congo red, crystal violet[7,15]. Among these stains, Congo red is the most useful, and it makes the amyloid proteins appear apple-green birefringence under a polarized light microscope[15,16]. However, since systemic amyloidosis has the same cutaneous features, these histological features cannot be distinguished from systemic amyloidosis. Therefore, systemic evaluations including blood tests, quantification of creatinine clearance are necessary to rule out systemic amyloidosis when amyloid deposits are found in the skin.

In nodular amyloidosis, AL proteins are produced by plasma cells, while amyloid fibrils are secreted by keratinocytes in other types of primary cutaneous amyloidosis[17]. As the deposited amyloid fibrils differ depending on the type of amyloidosis, it is important to know the exact type of the amyloid fibril for accurate diagnosis and treatment. Immunohistochemical staining using antibodies including kappa and lambda immunoglobulin light chains and amyloid A can help in recognizing the form of amyloid. Immunoglobulin light chain staining detects the tissue deposition of AL protein. Amyloid A immunostaining detects tissue deposition of serum amyloid A protein deposited in secondary systemic amyloidosis.

Surgical excision is the most frequently used treatment for primary cutaneous amyloidosis, and various methods such as dermabrasion, electrodissection, carbon dioxide laser, and cryotherapy have been attempted. In our case, due to the mass effect, surgical excision was performed. However, recurrence is quite frequent even after total removal[11,18]. In addition, regular monitoring is necessary to prevent its progression to systemic amyloidosis, which may occur in up to 7% of the patients[11,19].

To the best of our knowledge, only eight cases of PLCNA, including our case, have been reported in the Korean literature, and they are summarized in Table 1. Among these, four cases were located on the scalp, two cases on the toe, and one case, each, located on the leg and face. Immunohistochemical studies were performed in four cases[5,7,8]. In one case, AL proteins were positively stained with anti-kappa light chain antibody (cases 1), while other two cases showed positive staining results with anti-lambda light chain antibody (cases 3 and 4). Interestingly, our case showed positive staining for both kappa and lambda light chains, and it is the first case reported in Korea. Furthermore, it is worthy to note that they were completely removed with surgical excision.

Table 1. Cases of nodular amyloidosis reported in Korea

Case	Reference	Sex/age (yr)	Site	Morphology	Size (cm)	Types of amyloid fibril	Treatment	Result	Follow-up period	Systemic involvement
1	5	F/65	Leg	Multiple papules and nodules	2×1	AL (κ)	No	ND	ND	No
2	6	F/44	Toe	Nodule and papule	ND	ND	ND	ND	ND	No
3	7	M/38	Scalp	Three nodules	1×1	AL (λ)	No	Stable	2 years	No
4	8	M/47	Scalp	Nodule	2×3	AL (λ)	Excision	ND	ND	No
5	9	M/68	Cheek	Nodule	2×1	ND	Excision	No recurrence	8 months	No
6	10	M/50	Scalp	Nodule	5×5	ND	No	Stable	1 year	No
7	11	M/54	Toe	Multiple nodules	ND	ND	Excision	No recurrence	6 months	No
8	Our case	M/34	Scalp	Two nodules	5×3	AL (κ, λ)	Excision	No recurrence	18 months	No

F: female, M: male, AL: amyloid L protein, ND: not described.

CONFLICTS OF INTEREST

The authors have nothing to disclose.

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