The scalp nodules as the presenting symptom of primary localized cutaneous nodular amyloidosis: A rare case report

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ABSTRACT:

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Primary localized cutaneous nodular amyloidosis (PLCNA) is a rare variant of primary cutaneous amyloidosis which is characterized by a single or multiple yellowish waxy nodule with telangiectasias. The predilection site is the acral area but there are some reports presenting on the head and neck regions. Amyloid protein deposition in nodular amyloidosis originates from immunoglobulin light chains secreted by plasma c ells infiltrating the skin. We hereby report a case of 68-year-old women presented with multiple

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waxy skin-colored to yellowish nodules on the scalp for 2 years which prior suspected of nevus sebaceous. The incisional biopsy was done.

Histopathologic findings revealed nodular deposits of amorphous eosinophilic material in the entire dermis. The Congo red stain was positive for amyloid. The patient underwent exhaustive systemic evaluations, which were all negative. According to these finding, the diagnosis of primary localized cutaneous nodular amyloidosis was made. The patient was advised to undergo long term follow-up to monitor for potential progression to systemic disease.

Key words: nodular amyloidosis, primary localized cutaneous amyloidosis, scalp nodules

A 68-year-old Thai female presented with a 2-year duration of asymptomatic multiple waxy skin-colored to yellowish nodules on the scalp. The lesions were gradually increasing in size over time. There was no history of dry eyes or dry mouth. No other systemic symptoms such as fever, night sweat, weight loss, dyspnea or chest pain. She denied antecedent trauma to the scalp. No underlying medical conditions have been documented. No family member was affected with similar skin lesions. A nevus sebaceous was initially suspected. Physical examination revealed multiple ill-defined indurated waxy skin-colored to yellowish nodules confluent to a plaque of 4x3 cm on the vertex of scalp with telangiectasias. These nodules ranged from 0.5 to 2 cm in diameter (Fig. 1). Incisional biopsy revealed thinning of epidermis with massive nodular deposits of amorphous eosinophilic material throughout the whole dermis extended beyond the limits of resections (Fig. 2).

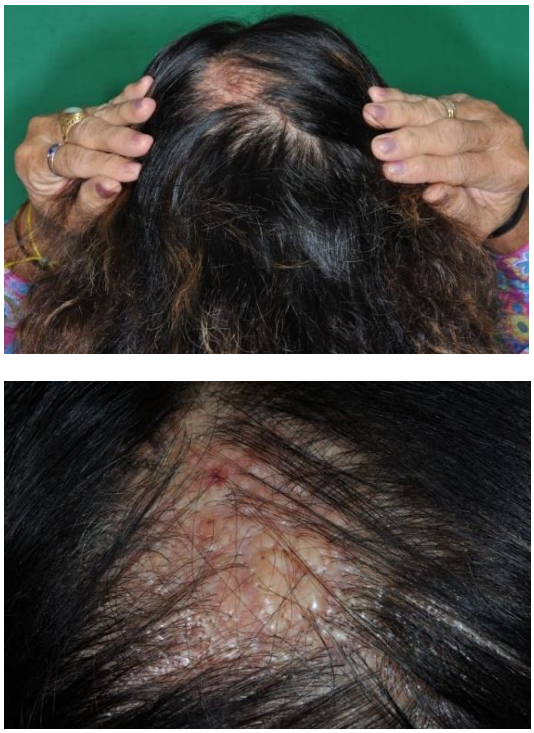

Figure 1 Localized group of indurated waxy skin-colored to yellowish nodules confluent to plaque on the vertex of scalp with overlying telangiectasia.

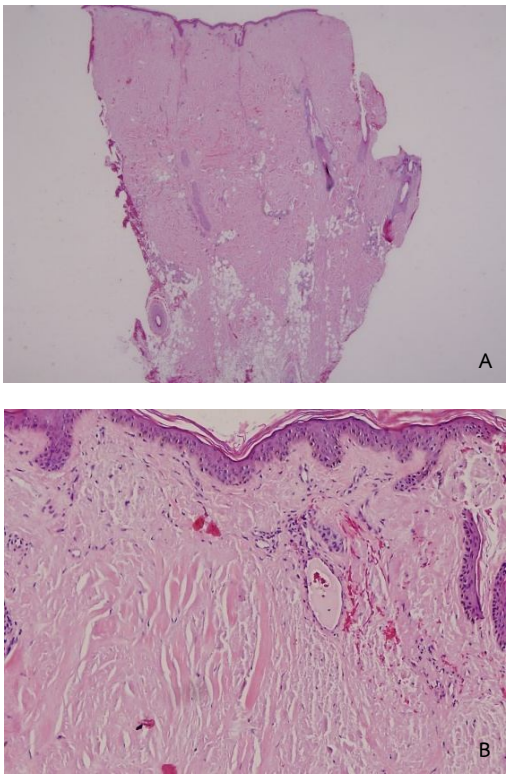

Figure 2 A biopsy specimen from a plaque on the scalp demonstrated a thinning of epidermis overlying a massive nodular deposits of amorphous eosinophilic material entire dermis. (H&E, original magnification X2 (A), X40 (B))

The Congo red staining revealed the amyloid material staining reddish color under conventional microscopy (Fig. 3) and showed apple-green birefringence under polarized light microscopy (Fig. 4).

Blood examination for complete blood count, blood urea nitrogen, creatinine, liver function tests were normal. The chest radiography showed no active lesions in the lungs. Electrocardiography showed normal sinus rhythms. Urinalysis was unremarkable. Serum protein electrophoresis and urine protein electrophoresis were normal. Sjögren's antibodies, anti-Ro/SSA and anti-La/SSB were all negative.

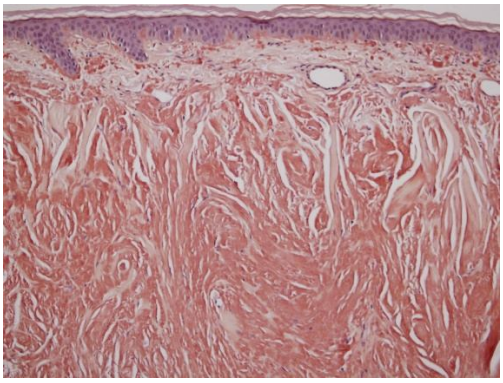

Figure 3 Congo red staining of a biopsy specimen was positive for amyloid. (original magnification X40)

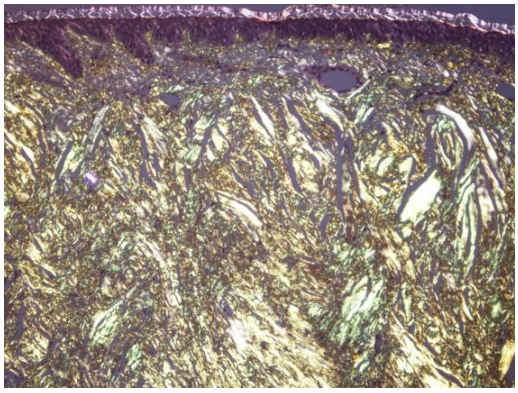

Figure 4 Congo red staining of a biopsy specimen shows apple-green birefringence under polarized light microscopy, consistent with amyloid deposition. (original magnification X40)

According to the clinical and histopathologic findings, the diagnosis of primary localized cutaneous nodular amyloidosis (PLCNA) was made.

Although there are no findings suggestive of systemic amyloidosis at the presentation time. She has been advised a long-term follow-up for surveillance of potential progression to systemic amyloidosis. Our patient did not want to receive treatment as a serial excision due to cosmetic concern. There is no evidence of progression to systemic amyloidosis during a follow-up of 1 year.

Discussion

Amyloidosis is a rare metabolic disease and caused by extracellular deposition of insoluble abnormal amyloid fibrils derived from the aggregation of misfolded proteins[1,2]. Deposits of amyloid can be seen in plasma cell dyscrasias, Alzheimer disease, familial polyneuropathies and primary cutaneous lichen amyloidosis. Amyloidosis can be classified clinically into systemic forms, which involve several organ systems, and localized forms, which are limited to a single organ such as the skin[3]. In this case, we focus mainly on the localized cutaneous amyloidosis which consists of macular amyloidosis, lichen amyloidosis and the rarest type, nodular amyloidosis. The amyloid deposits in macular amyloidosis and lichen amyloidosis are degenerative keratinocyte-derived, whereas those in nodular amyloidosis are mostly composed of immunoglobulin light chains (AL protein) secreted by infiltrating clonal plasma cell population[2,3,4].

PLCNA is the rarest form of cutaneous amyloidosis which is characterized by amyloid material depositing in the dermis, subcutaneous fat, and blood vessel walls. The patient's age is between 50 and 60 years old and both genders are often affected equally[4,5]. Predilection sites are on the acral areas which are traumatic areas but it can also appear on the face or trunk. It can present as a single or multiple pink to yellowish-brown, waxy tumefactive nodules ranging from several millimeters to several centimeters in size.

PLCNA lesions may progress to large bullae and the epidermis may become atrophic. The nodules may be friable, contain superficial telangiectasias, or hemorrhage as a consequence of perivascular amyloid deposition. Traumatic injury is proposed as a triggering factor for the development of PLCNA in some cases[4]. The clinical differential diagnosis of PLCNA includes lymphoma cutis, pseudolymphoma, pretibial myxedema, cutaneous sarcoidosis, granuloma annulare, reticulohistiocytoma and multicentric reticulohistiocytosis and granuloma faciale[5].

In our patient, the diagnosis is primary localized cutaneous nodular amyloidosis due to the compatibility of histopathology by H & E and

Congo red staining. Investigations for systemic amyloidosis were all negative. Interestingly she has the lesion on the scalp which is an unusual location. Although frequent cutaneous sites of involvement are the extremities, trunk, and genitalia, it can occur anywhere on the skin. The scalp lesion has firstly been reported from Korea which occurs after local minor trauma such as hitting a ball with the head[4]. However, in our case, the patient denies any previous trauma on the scalp.

The diagnosis of nodular amyloidosis is primarily made of histopathology using (H&E stain) which shows amorphous nodular eosinophilic hyaline material in the entire dermis. The confirmed diagnosis is made by Congo red staining[6]. The Congo red staining with polarizing microscopic examination revealed apple-green birefringence throughout the whole dermis.

The PLCNA has been reported 7 to 50% risk of progression from localized nodular cutaneous amyloidosis to systemic forms on long term follow-up, while macular and lichen variants do not[4,5,7]. On the other hand, some reports found 40% of patients with primary systemic amyloidosis can present with PLCNA[5]. Therefore, evaluation for systemic involvement at the time of diagnosis is recommended. In our patient, we have investigated for possible systemic manifestations which include renal, cardiac, hepatic, peripheral nervous system evaluations. There was no any evidence suggestive of systemic amyloidosis in this patient. The PLCNA linkage to Sjögren's syndrome as a result of a benign clonal proliferation of plasma cells in the skin that is part of the lymphoproliferative diseases has been reported[2,8]. Therefore It is essential to investigate for any evidence of Sjögren's syndrome in the patients diagnose with PLCNA.

In this patient, the results were all negative. In the absence of investigation findings for secondary systemic amyloidosis, the condition was mostly compatible with primary localized cutaneous nodular amyloidosis

Treatment of nodular amyloidosis is challenging due to lack of effective treatment and local recurrences are common. Management options consist of surgical excision with or without split-thickness skin graft, cryotherapy, electrodesiccation and CO_2 laser. Other satisfactory treatment options are dermabrasion, pulsed dye laser, intralesional methotrexate, intralesional steroid, localized radiation and combination use of cyclophosphamide and prednisolone[5,9,10,11]. There are some new reports about the treatment of amyloidosis including the depletion of the ubiquitous amyloid P component via complexes with the experimental agent and immunotherapy to promote clearance of

amyloid using specific antibodies[12,13]. In our patient, the lesion has not been excised yet due to the large size of the lesion and her cosmetic concern. But we do stress the importance that she must regularly follow-up with us for examination and surveillance of systemic amyloidosis that may occur in the future. Systemic evaluation should be performed at least yearly and when clinically indicated.

In conclusion, we report a rare variant with unusual location of primary localized cutaneous nodular amyloidosis presented with a cluster of multiple scalp nodules without previous trauma. Due to the potential progression to systemic disease of PLCNA, the patient should be advised to undergo long term follow-up. This case emphasizes a practical point that subcutaneous scalp nodules which may appear clinically benign at first should not be overlooked.

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