

A Case of Lichen Amyloidosis with Complete Response to Mesalamine

Fakhreddin Sabooniha, MD

Sabzevar university of medical sciences

Razavi Khorasan province, Sabzevar , Farmandari St, Amin building ,3rd floor ,

Abstract:- Primary cutaneous amyloidosis (PCA) is the deposition of the amyloid in the skin without systemic involvement. It has three subtypes: macular, lichen and nodular. Lichen amyloidosis is the most common subtype and usually presents as severely pruritic plaques on the shins or other extensor surfaces,. lichen amyloidosis has also been reported in association with autoimmune diseases such as systemic sclerosis, systemic lupus erythematosus and primary biliary cirrhosis.

I report the case of a 41-year-old female patient who developed lichen amyloidosis as an early presentation of underlying inflammatory bowel disease (IBD) with complete response to IBD treatment. IBD has been reported with various skin manifestations but this association has not been reported in the literature.

Keywords:- Lichen Amyloidosis , Inflammatory Bowel Disease, Mesalamine ,Primary Cutaneous Amyloidosis.

Abbreviations :	PCA	Primary cutaneous amyloidosis
	ESR	erythrocyte sedimentation rate
	ANA	anti- nuclear antibodies
	IBD	inflammatory bowel disease
	ASCA	anti-saccharomyces cerevisiae antibody
	IgG	immunoglobulin G
	EIM	extraintestinal manifestations

I. INTRODUCTION

Amyloidosis is classified into systemic forms, with involvement of several organ systems, and localized forms, in which deposits are limited to a single organ such as the skin.¹Primary systemic amyloidosis is associated with hematologic disorders such as multiple myeloma and affects mostly muscles, heart, gastrointestinal tract and peripheral nerves .Secondary systemic amyloidosis has greater association with chronic infections and inflammatory diseases e.g. tuberculosis and rheumatoid arthritis with a predilection for the kidneys and gastrointestinal system involvement ².

Primary cutaneous amyloidosis (PCA) is the deposition of amyloid in the skin without systemic involvement. The disease has three subtypes: macular, lichen and nodular ². About half of IBD patients may be complicated by

extraintestinal manifestations (EIMs) such as erythema nodosum and lichen planus and the prevalence of EIMs is higher in Crohns' disease compared to ulcerative colitis ³.

Here, I report a case of a 41-year-old patient with lichen amyloidosis as an early presentation of underlying IBD with complete response to mesalamine. Previous report in the literature about this type of association was not found.

Primary cutaneous amyloidosis can have an unfavorable effect on quality of life of the patients due to the pruritus and cosmetic issues ⁴.

Lichen amyloidosis is the most common form of primary cutaneous amyloidosis with predilection for the shins or other extensor surfaces ^{5,6}.

Initial lesions are discrete, firm skin-colored or hyperpigmented papules, which later coalesce into plaques with a rippled pattern. At first, lesions are usually unilateral, but over time a bilateral distribution can develop ⁶.

Macular and/or lichen amyloidosis has also been described in association with various autoimmune disorders ⁷. But their association with inflammatory bowel disease has not yet been reported in the literature.

II. CASE REPORT

A 41-year old female patient presented to our clinic with a pleuritic chest pain. Vital signs were normal. Physical examinations were normal except for a well-defined skin lesion on the anterior side of right shin with multiple discrete and firm brown papules and an area of central scar and atrophy(Fig1).

The lesion lasted for 8 years and was strongly pruritic .These findings were consistent with a diagnosis of **Lichen Amyloidosis** ,a disorder classified as primary (localized) cutaneous amyloidosis(PCA) .she also complained of chronic low back pain with morning stiffness lasting about 1hour, constipation and bloating. Electrocardiography was normal .On echocardiography, only a mild pericardial effusion was detected. Laboratory tests revealed an elevated ESR and negative ANA .Radiography showed bilateral sacroiliac joints sclerosis .(Fig 2).Her mother has a history of crohns' disease. She was not smoker and used no alcohol or illicit drugs. Her drug history was negative.

Based on her symptoms and signs, family history and laboratory and radiologic findings, a diagnosis of inflammatory bowel disease (IBD) was suspected and due to patient's unwillingness to undergo any procedure, an IBD serology panel was requested that showed a positive anti-saccharomyces cerevisiae IgG antibody (ASCA IgG) which has a high specificity for Crohn's disease.

Therefore, a trial of mesalamine 800 mg four times a day started with a near complete resolution of skin lesion in addition to improvement of other symptoms (Fig3).

III. CONCLUSION

Lichen amyloidosis is a chronic skin disease with suboptimal response to current therapies. It has numerous medical and social morbidities for the patients. Generally, Treatment is directed to alleviating the symptoms. Side effects of systemic therapies and the high recurrence rate of surgical interventions make it essential to use more targeted therapies against underlying conditions if they exist. In addition, the skin manifestations of IBD might precede its gastrointestinal sign and symptoms as in my case and therefore awareness of this skin finding as an early presentation of underlying IBD may help clinician to start more appropriate and targeted therapies with better outcome.

REFERENCES

- [1]. Richard W. Groves .Amyloidosis .In Jean L. Bologna, Julie V. Schaffer, Lorenzo Cerroni .Dermatology 4th ed Elsevier 2018: 754-760
- [2]. Jean R. Tafarela, Lara B. Lemos, Patricia M. Oliveiraa, Cutaneous amyloidosis associated with primary biliary cirrhosis, European Journal of Gastroenterology & Hepatology 2007, 19:603–605
- [3]. BrianL. Huang, Stephanie Chandra and David QuanShih , skin manifestations of inflammatory bowel disease ,Frontiers in physiology doi: 10.3389/fphys.2012.00013
- [4]. Fang S, Shen X, Chen AJ, et al. Health-related quality of life in patients with primary cutaneous amyloidosis. PLoS ONE 2015;10:e0120623.
- [5]. Sipe JD, Benson MD, Buxbaum JN, et al. Amyloid fibril protein nomenclature: 2010 recommendations from the nomenclature committee of the International Society of Amyloidosis. Amyloid 2010;17:101–4.
- [6]. Wang WJ. Clinical features of cutaneous amyloidoses. Clin Dermatol 1990;8:13–19
- [7]. Breathnach SM. Amyloid and amyloidosis. J Am Acad Dermatol 1988;18:1–16.

IMAGE LEGENDS



Fig1:- multiple uniform discrete brown papules with rippled appearance on anterior side of the right shin consistent with *lichen amyloidosis*.



Fig 2:- Near complete resolution of the papules after six weeks of treatment with Mesalamine with residual postinflammatory hyperpigmentation



Fig 3:- sclerosis of both sacroiliac joints with left predominancy .