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Case report

Poikiloderma-like cutaneous amyloidosis – a rare presentation of primary localized cutaneous amyloidosis

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Abstract

Poikiloderma-like cutaneous amyloidosis (PCA) is a rare variant of primary cutaneous amyloidosis. It was first described in 1929 and there are two clinical forms of PCA, the ordinary type and PCA syndrome. The characteristics of PCA include poikiloderma-like skin changes, lichenoid papules, blister formation, and cutaneous amyloid deposits on histological examination. These skin lesions usually occur at the extremities, consistent with the few cases that have been reported. We present a case of a 62-year-old man who presented with the features of poikiloderma-like cutaneous amyloidosis. Diagnosis of this unique condition is a challenge and a skin biopsy is necessary in such instances. A discussion of the differential diagnosis of this condition is also included.

Keywords: Cutaneous amyloidosis, Poikiloderma, Dyschromia

Introduction

Primary cutaneous amyloidosis is the deposition of amyloid in the papillary dermis without systemic involvement. It commonly presents as a papular (lichen amyloidosis) or macular form. We report a case of poikiloderma-like cutaneous amyloidosis (PCA) which is a rare distinct type of primary cutaneous amyloidosis.

Case synopsis

A 62-year-old man presented with a 3-year history of pruritic poikilodermatous skin lesions. The lesions started as erythematous papules and gradually hyperpigmented plaques formed. Some lesions subsided into small hypopigmented macules. There was sporadic recurrence of blisters over the papules, patches, and plaques. These mainly affected the waist, groin, hip, and intergluteal region symmetrically, sparing the extremities. He did not have any photosensitivity. There was no history of contactants, no paraesthesia preceding the blisters, and no new oral medications. He had a history of hypertension, hyperlipidemia, and Type 2 diabetes mellitus. Oral medications included metformin, lovastatin, and valsartan. There was no family history of skin disorders. Systemic review was unremarkable.

Physical examination revealed a tense blister with reddish brown papules on a background of hyperpigmented patches and plaques, with guttate leukoderma symmetrically distributed over the waist, groin, hip, and intergluteal region (Figures 1-4). This produced a striking mottled appearance, especially over the trunk. He was of normal stature with a height of 1.74m. Palms were not affected and examination of the hair, nails, teeth, and mucosa was normal. Systematic examination was also unremarkable.

Our initial differential diagnosis included cutaneous dyschromias, bullous lichen planus, recurrent herpes simplex virus infection, immunobullous diseases, and poikilodermatous mycosis fungoides.

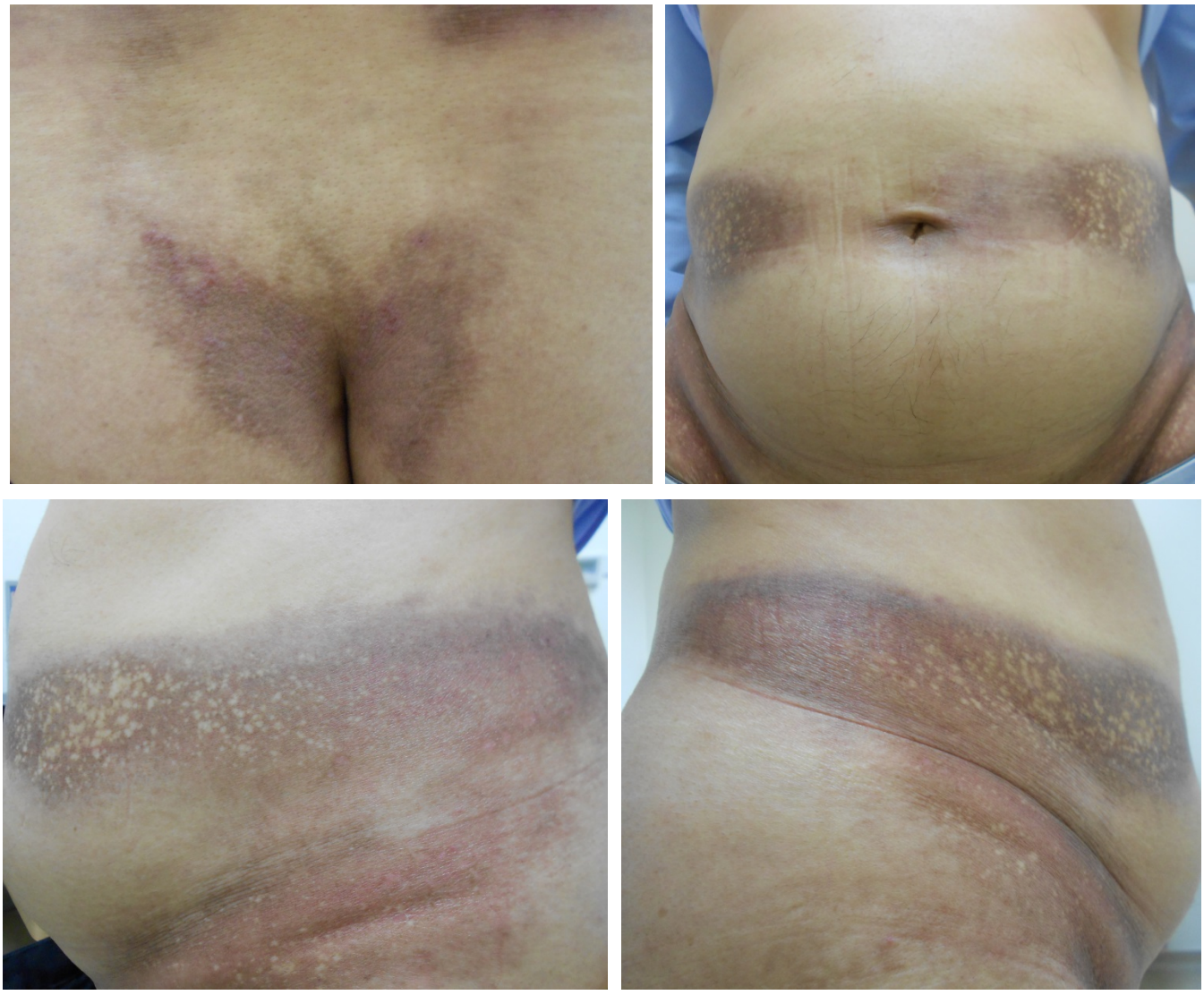
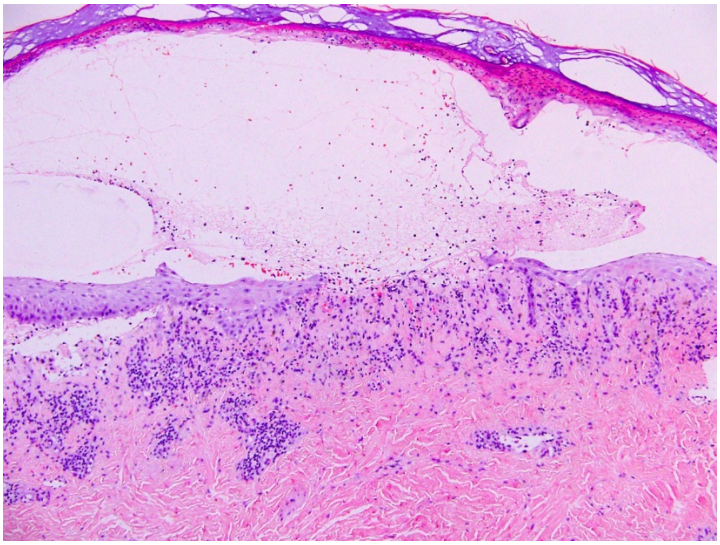


Figure 1-4. Symmetrical poikilodermatous lesions over waist, groin and hips. Lichenoid pigmented papules in the intergluteal region.



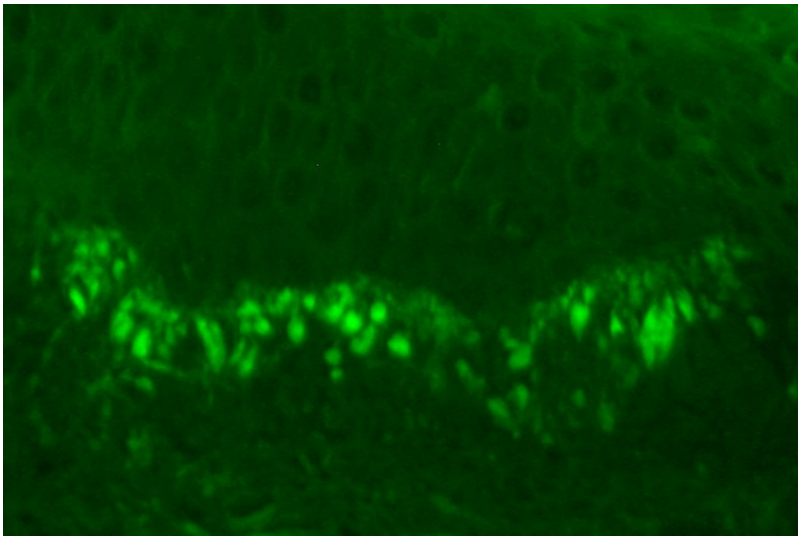


Figure 5. Histology showing subepidermal blister with basal vacuolar changes and superficial perivascular infiltrate consisting mainly of lymphocytes (magnification x 40) **Figure 6.** Positive staining with Thioflavin-T in papillary dermis corresponding to hyaline deposits on light microscopy

A skin biopsy was taken from a blister edge over the distinct poikilodermatous patch on the right hip. Histology showed a subepidermal blister. The blister contained some neutrophils and cell debris. There was mild basal vacuolar change at the blister edge (Figure 5). The underlying dermis showed superficial perivascular infiltrate of mainly lymphocytes. Melanophages and cytoid bodies were prominent. There were mild hyaline deposits at the papillary dermis beneath the basement membrane zone (Figure 6). A Thioflavine-T stain was positive for amyloid in the papillary dermis corresponding to the mild hyaline deposit and cytoid bodies on light microscopy (Figure 7). Amyloid AP was positive whereas AA was negative. Immunofluorescence showed granular deposits of C3 and C1q. There were also deposits of IgG and fibrin. The immune deposits were present in the papillary dermis beneath the basement membrane zone. Periodic acid Schiff and gram stain were negative. The skin biopsy was also negative for viral cytopathic effect, herpes simplex virus, and cytomegalovirus immunoreaction.

After the skin biopsy, our focus shifted to amyloidosis and other investigations were done to exclude the presence of systemic amyloidosis. Investigations such as full blood count, renal function, liver function, urine analysis, chest X-ray, and abdominal ultrasound scan were within normal limits. Serum protein electrophoresis was normal and he had a negative urine Bence-Jones protein test.

In view of clinical and histopathological findings, we diagnosed the patient with poikiloderma-like cutaneous amyloidosis. He was given topical mometasone cream to apply to the skin lesions and subsequently declined further follow up.

Discussion

Poikiloderma-like cutaneous amyloidosis is a rare variant of primary cutaneous amyloidosis. There are two clinical forms of PCA described: the ordinary type and PCA syndrome [1]. The ordinary type is characterized by the presence of poikilodermatous lesions, lichenoid papules, and blisters usually occurring on the limbs with an adult onset. PCA syndrome has other features which include an early onset, photosensitivity, short stature, and occasionally palmoplantar keratosis. Our patient is classified as the ordinary type of PCA. He did not have palmoplantar keratosis, photosensitivity, or short stature. The lesions also appeared in his adulthood. The additional unique feature of his condition was the distribution of his skin lesions. The extremities were spared and the skin lesions were mostly concentrated at the lower trunk. The few cases that have been reported had lesions affecting the extremities [1-5] and therefore, this is the first case report of PCA with an uncommon truncal distribution.

Other more common primary cutaneous amyloidosis include the macular and lichen variants. Macular amyloidosis manifests as brown, rippled macules distributed symmetrically usually over the upper back. Lichen amyloidosis is characterized by itchy hyperkeratotic papules over the extensor surface of the lower limbs. When macular amyloidosis and lichen amyloidosis co-exist, this is termed as biphasic amyloidosis as proposed by Brownstein [6]. Nodular amyloidosis presents as brown-pink, waxy nodules, often with telangiectasia. These are usually found on the face, extremities, trunk, and genitalia [7]. Apart from these 3 main types, rare variants include PCA, amyloidosis cutis dyschromia (ACD), and bullous, vitiliginous, and anosacral forms [2].

The differential diagnoses in this patient include ACD, poikilodermatous mycosis fungoides, confluent and reticulated papillomatosis, bullous lichen planus, dyschromatosis symmetrica hereditaria, and recurrent herpes infection with post inflammatory dyspigmentation (Table 1). The presence of amyloid material in histological examination excluded all the other conditions mentioned except for ACD. ACD is characterized by a generalized distribution of dotted, reticular hyperpigmentation with hypopigmented macules, no or little itch, onset before puberty, and focal subepidermal amyloid deposition [8]. It may be difficult to differentiate between ACD and PCA in some patients due to overlapping clinical features. However, our patient's presentation was clearly of PCA with the pruritic poikilodermatous lesions and adult onset.

Table 1. Differential Diagnoses of PCA

Disorder	**Clinical features**	**Distribution**	**Histology**
Poikiloderma-like Cutaneous Amyloidosis	i) Poikilodermic lesions, lichenoid papules, and blisters ii) Adult onset	Predilection for the limbs	Amyloid deposits in the papillary dermis
Amyloidosis Cutis Dyschromia	i) Dotted, reticular hyperpigmentation with hypopigmented macules	Nearly all of the body	Focal amyloid deposition under the epidermis

	ii) No or little itch iii) Onset before puberty		
Poikilodermatous Mycosis Fungoides	i) Atrophy, telangiectasia, and reticulate or mottled hyper- and hypopigmented plaques ii) No or little itch	May be localized or diffuse. Often noted on the breasts, hips, buttocks, and flexural areas.	Atypical T-cell infiltrate in the papillary dermis with epidermotropism, epidermal atrophy, dilated blood vessels in the dermis, melanophages, and melanin incontinence
Confluent and Reticulated Papillomatosis	Grayish blue hyperkeratotic, papillomatous papules which coalesce to form confluent plaques centrally and a reticular pattern peripherally	Usually on the trunk. First lesions usually appear between the breasts and in the midline of the back.	Hyperkeratosis, papillomatosis, and mild acanthosis, with a mild superficial perivascular dermal infiltrate
Lichen Planus	Can present with a variety of lesions, the most common presentation is "6 P's" of lichen planus: planar (flat-topped), purple, polygonal, pruritic, papules, and plaques	May affect any area, but is most often seen on the front of the wrists, lower back, and ankles	Band-like infiltrate of lymphocytes at the epidermal-dermal junction and damage to the basal cell layer. Epidermal changes include wedge-shaped hypergranulosis and irregular acanthosis leading to a saw-toothed appearance.
Dyschromatosis Symmetrica Hereditaria	i) Hyperpigmented and hypopigmented macules of various sizes ii) Appear in infancy or early childhood	Dorsal aspects of extremities and face, occasionally on trunk	Hyper- and hypomelanization in hyper- and hypopigmented lesions, respectively

There is no single effective treatment for cutaneous amyloidosis. Patients seek treatment to relieve symptoms of itch and improve the appearance of the lesions. As pruritus is commonly reported, first line treatments include topical and intralesional steroids and antihistamines. Topical dimethyl sulfoxide, hydrocolloid dressing, calcipotriol, calcineurin inhibitors, acitretin, cyclophosphomide, phototherapy, dermabrasion, and cryotherapy have also been used with varying effects. Laser treatment used include pulsed-dye, carbon dioxide and neodymium:yttrium aluminium garnet (Nd:Yag) [9-11]. Most cases of cutaneous amyloidosis remain chronic.

In conclusion, we present this rare case to alert dermatologist to consider PCA in patients with cutaneous dyschromia. In patients presenting with poikilodermatous skin lesions, it may be prudent to perform a skin biopsy as diagnosis can pose as a challenge.

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