

Diagnostic imaging

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Clinical and dermoscopic diagnosis of a case of exuberant macular amyloidosis

Diagnóstico clínico e dermatoscópico de um caso de amiloidose maculosa exuberante

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ABSTRACT

Amyloidosis is characterized by the deposition of a modified protein. It can affect the skin by accumulating in the papillary dermis. In general, it develops with brownish maculae predominant in the interscapular region, and rarely coursing with generalized forms. This report describes a 59 year-old female patient who had had a spot in the lower back for 43 years, which in the last 8 years had progressively spread for the entire body. Dermoscopy has proven extremely effective in locating the various foci of pigmentary incontinence, which together with apoptotic keratinocytes constitute the pathophysiological basis for the formation of the amyloid protein.

Keywords: amyloidosis; congo red; dermoscopy

RESUMO

A amiloidose é entidade caracterizada pela deposição de uma proteína modificada. Pode acometer a pele depositando-se na derme papilar. Em geral cursa com máculas acastanhadas predominantes na região interescapular e raramente com formas generalizadas. O presente relato refere-se a paciente do sexo feminino com queixa de manchas hipercrômicas de aumento progressivo para todo o corpo. A dermatoscopia se mostrou extremamente eficaz para a localização dos diversos focos de incontinência pigmentar, que junto com os queratinócitos apoptóticos constituem a base fisiopatológica para a formação da proteína amiloide.

Palavras-chave: amiloidose; vermelho-congo; dermatoscopia

Amyloidosis is a buildup, in any affected organ, of an abnormal protein which is the result of the combination of polysaccharides and globulin. Macular amyloidosis is the most common subtype of cutaneous amyloidosis. It typically presents with pigmentation in a reticular or wavy pattern on the interscapular region, affecting the extensor aspect of arms, forearms and legs¹ to a lesser extent, rarely becoming generalized. Histopathology shows deposits of amyloid substance in the papillary dermis and, when stained with congo red under polarized light microscopy, shows greenish birefringence.² Areas of pigmentary incontinence are also seen on the same location.³ We followed a 59-year-old female patient, born and living in São José dos Campos (SP), who presented with a complaint of a brownish patch in the sacrum for 43 years and progressive enlargement over the past 8 years.

On physical examination, the patient had brownish patches, some of them reticulated, interspersed with normal skin all over the body, sparing only fingers, feet and scalp (Figure 1). On dermoscopy, multiple brown waves on the inferior aspect of the

right leg, formed by brownish squared structures with fine streaks in the center of the lesion were of interest (Figures 2 and 3). She denied previous health issues or use of regular medications. Histopathology showed with apoptotic keratinocytes in the epidermis and enlargement of the papillary dermis, with deposition of hyaline eosinophilic globules, besides melanophages and mild superficial perivascular lymphomononuclear infiltrate (Figure 4).

Cutaneous amyloidosis can be classified in primary and secondary. Of the primary forms, macular amyloidosis, like the case described, is the most common type. Still, the generalized presentation, such as our patient's, is rarely seen in clinical practice.¹ Diffuse forms can simulate nevroid pigmentation, such as the poikilodermatous form.² Clinically, brown-gray patches, with varying diameters between 2-3mm are seen. The reticular or serpiginous pattern is typical. The diagnosis, initially clinical and



FIGURE 1: Profuse lesions on admission



FIGURE 2: Macroscopic image of the right thigh

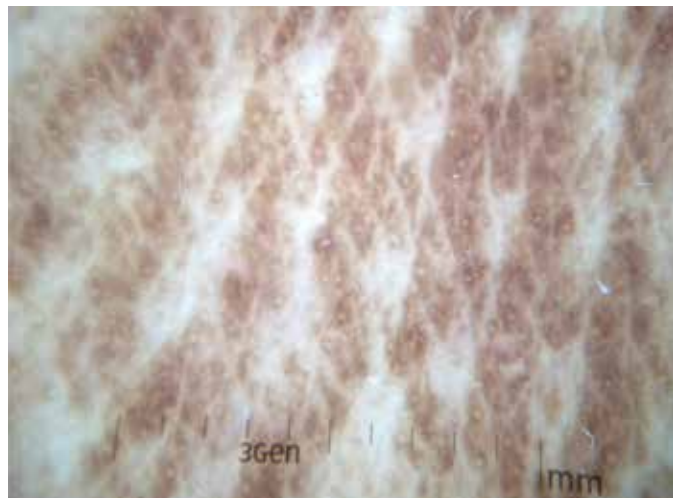


FIGURE 3: Dermoscopic image of the patient's right thigh

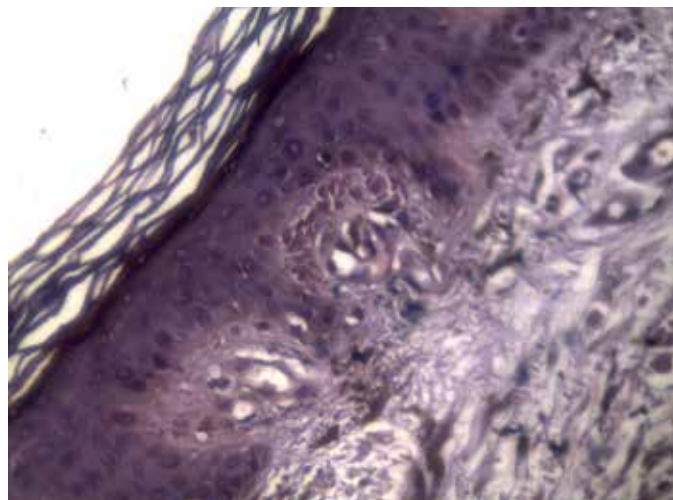


FIGURE 4: Congo red staining showing amyloid protein buildup in the patient's skin

easily mistaken for many other conditions including lichen simplex chronicus, atrophic lichen planus, lichen sclerosus, atopic dermatitis, hemochromatosis, xanthoma, pityriasis versicolor, toxic melanoderma, among others, can be differentiated on dermoscopy by the findings of multiple small brown central cubes with fine streaks radiating from the center (Figures 5 and 6). The distinctive histopathological feature is amyloid substance deposition in previously healthy skin without deposition in other organs. It is more commonly

seen in Central and South Americas, Middle East and Asia, perhaps because of cultural habits. There are associated genetic factors as well, as we can see familial cases described in the literature. Other associated factors include ultraviolet B radiation (UVB), Epstein-Barr virus and race.³ During puberty, this type of amyloidosis affects both sexes equally, but there is female predominance between 20 to 50 years of age.^{3,4}

The treatment for this condition is generally disappointing. Topical superpotent steroids are usually used for a short



FIGURE 5:
Macroscopic
image of the
left flank



FIGURA 6: Dermoscopic image of the patient's left flank

time. Calcipotriol and phototherapy are similarly of limited use.⁵ Dimethyl sulfoxide can improve pigmentation, but it completely recurs upon discontinuation.³

We demonstrate a florid case of disseminated macular amyloidosis, with characteristic dermoscopic pattern that helps differentiating from other conditions that are clinically similar. There is no question about the importance of the histopathological examination, but we highlight the importance of dermoscopy for the diagnosis of yet another dermatological condition. ●

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DECLARATION OF PARTICIPATION:

Rafael Abdala Beicher:

Data analysis and interpretation
Study design and planning
Critical review of the literature

Elizabeth Leocádia Fernandes:

Approval of the manuscript's final version
Study design and planning

Denise Steiner:

Intellectual participation in the therapeutic approach
Critical review of the literature Intellectual participation in the therapeutic approach
Critical review of the manuscript
Critical review of the manuscript