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MACULAR AMYLOIDOSIS: ATYPICAL LOCATION IN MEN

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Abstract: Amyloidosis is characterized by the deposit of a modified protein. It can affect various organs, including the skin, when deposited on the papillary dermis. Macular amyloidosis is the main form of primary cutaneous amyloidosis, characterized by hyperchromic macles predominantly in the interscapular region. This report is of a 53-year-old man, for 4 years with a hyperchromic, lichenified and pruriginal spot on the posterior region of the left upper limb, knee and back of the left hand. No associated systemic changes. Skin biopsy, in histopathological study, showed small focal deposits of amorphous eosinophil material in dermal papilla and positive for congo red color. The present work aims to report cutaneous amyloidosis in an atypical location.
Keywords: cutaneous amyloidosis, location and atypical location

INTRODUCTION

Macular amyloidosis is the most common form of localized primary cutaneous amyloidosis. Its physiopathogenesis is not fully known, but many factors are associated with its development, such as epstein-barr, uvb, synthetic tissue, race, genetic factors and atopias. It starts around the age of 30, with equivalent incidences in both sexes, with a predomination in the white race. The lesions are typically described as reticulated hyperchromic macules, whose most common location is the interscapular region, being rare in limbs and chest. Histopathology presents with deposit of amorphous crystals in the dermal papillas stained by congo red and if seen in polarized light they show clear birrefringence.

CASE REPORT

Male patient, 53 years old, white, hypertensive, dyslipidemic and diabetic, reporting hyperpigmented and prurigious lesions in the posterior region of the left upper limb, left knee and left hand for 3 years, without other associated symptoms and without results advertisement to the use of moisturizers and dexamethasone ointment. During dermatological examination, a brown hyperchromic macule was evidenced, lichenified, descamative, with poorly defined edges, about 5 cm in diameter on the back, hand and knee region. The patient had no systemic complaints and the remainder of the physical examination showed no other changes. Complementary serum exams, including hemogram, kidney and liver injury tests and serology, without changes. In the biopsy, with histopathological study, the dermis was observed with small focused deposits of amorphous eosinophil material in the dermal papilla, associated with some melanophages and positive in alkaline congo red color (material suggesting amyloid substance), firming so the diagnosis. In accordance with the result and in the absence of systemic changes, topical therapy with clobetasol 0.05%, Phototherapy and clinical follow-up were instituted.

DISCUSSION

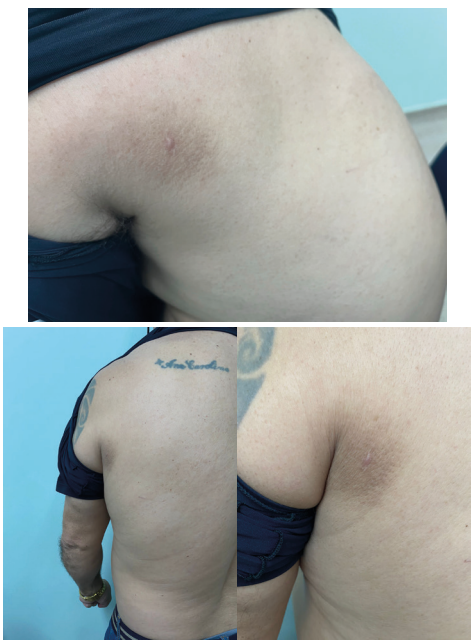
Amyloidosis, regardless of the tissue affected, are deposits of abnormal proteins, resulting from the union of polysaccharides with globulin. Primary cutaneous amyloidosis is divided into 4 subtypes: lichen amyloidoticus, nodular amyloidosis, biphasic amyloidosis and macular amyloidosis, the last of them being the most common. Macular amyloidosis is characterized by brown reticulated or serpentine spots, mainly in the interscapular region. This report is an example of a typical shape but with atypical location,

in the posterior region of the left upper limb (figures 1,2 and 3), left knee and back of hand. In biopsy with histopathological study, the presence of amorphous material in dermal papilla, stained by congo red, confirmed the diagnosis. It is most commonly reported in asian and latin american countries, especially in caucasian patients. Its incidence increases with age. The patient described is latin american and in the 50s.

There is a strong relationship between this disorders and paresthetic notalgia. Studies suggest that many cases of chronic notalgia parestetica may result in the formation of macular amyloidosis due to secondary chronic attrition. This is a hypothesis for the patient in question, who presented intense itching in long-standing injuries.

Cutaneous amyloidosis resembles many other clinical conditions, including lichen simple chronicus, lichen plans, atopic dermatitis, pythriasis versicolor, melanoderma toxia, lichen sclerosus, and hemochromatosis. Which can lead to difficulty and delay in defining the final diagnosis. In addition to histopathological confirmatory diagnosis, immunohistochemistry with c4d has shown good results for tracking cutaneous amyloidosis, being an easier and quicker test than congo red staining and with good sensitivity and specificity (100% and 75.5%).

Treatment is usually disappointing, with a wide therapeutic option - retinoids, glucocorticoids, cyclophophamide, cyclosporines, amitriptyline, colchicine, vitamin d3, tacrolimus, laser, phototherapy - although without much success. The use of laser in treatment has promised but still inconclusive results in the long term, because they are recent studies. The most promising practical results are achieved by the combination of phototherapy with grape and high potency topic corticoids. Exactly the same treatment prescribed for the patient in this case, associated with skin hydration and clinical follow-up. This study shows an atypical form of presentation due to the location of the lesions, in addition to demonstrating the difficulty of the treatment. Hereby, it can be observed the need for more studies in the area that contribute to pathophysiological elucidation and expand horizons in front of constantly effective therapies.



(Figures 1,2 and 3: cutaneous amyloidosis in the posterior region of the left upper limb)

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