

Corymbiform nodular amyloidosis *

Amiloidose nodular com lesão corimbiforme

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Abstract: Amyloidosis is part of a group of deposition diseases. Nodular amyloidosis is a rare form of primary cutaneous amyloidosis. It affects men and women, usually over the age of 60 years. Presenting manifestation of the disease are yellowish-erythematous or brownish nodules or plaques in single or multiple infiltrates. Systemic evaluation should be performed to rule out involvement of other organs. Follow-up of the patient is important because the condition may progress to systemic amyloidosis. We report a case of nodular amyloidosis in which the lesion had a corymbiform aspect without systemic involvement and no recurrence after two years of follow-up.

Keywords: Amyloid; Amyloidosis; Congo red

Resumo: As amiloidoses constituem um grupo de doenças de depósito. A amiloidose nodular é uma forma rara de amiloidose cutânea primária. Acomete homens e mulheres, geralmente acima de 60 anos. Apresenta-se com nódulos ou placas eritemato-amareladas ou acastanhadas infiltradas isoladas ou múltiplas. A avaliação sistêmica deve ser feita para descartar comprometimento de outros órgãos. É importante o seguimento devido a possibilidade de evolução para amiloidose sistêmica. Relatamos um caso de amiloidose nodular com lesão de aspecto corimbiforme sem sistematização e sem recidiva após dois anos de seguimento.

Palavras-chave: Amiloidose; Amilóide; Vermelho congo

INTRODUCTION

Amyloidosis is a term that describes a group of disorders characterized by extracellular deposition of β -fibril protein substance in tissues. This substance may be deposited in any organ of the body or may be restricted to a certain tissue.¹ In Dermatology, the term “corymb” or corymbiform means a main central lesion and other satellite lesions.² We present a case of nodular cutaneous amyloidosis that is a rare form of primary cutaneous amyloidosis with corymbiform lesion.

CASE REPORT

A black, male, 62-year old patient presented with a complaint of asymptomatic lesions of gradual growth on the dorsum for two years. Dermatologic examination revealed a corymbiform lesion formed by a 2.5 cm brownish nodule, surrounded by papules, some skin colored, while others were light brown, with fibroelastic consistency and localized in the left scapular region (Figure 1). On physical examination, painless, isolated, elastic, firm, enlarged inguinal

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FIGURE 1: Corymbiform nodular amyloidosis. Corymbiform lesion formed by brownish nodule surrounded by skin-colored papules

lymph nodes, 0.8 cm in size, were found bilaterally. The patient had a personal history of controlled arterial hypertension and was medicated with hydrochlorothiazide. Family history was negative for any similar disorder.

An incisional biopsy of the lesion showed deposition of eosinophilic, diffuse amorphous material in the dermal layer of the skin and subcutaneous tissue, stained in red by Congo red dye and apple-green birefringence under a polarizing microscope (Figures 2, 3 and 4). Immunohistochemistry revealed that about 75% of the plasma cells present in the dermis expressed kappa chain, while the remaining plasma cells expressed lambda chain (Figure 5 and 6).

Hematologic, kidney and liver function tests, serum glucose, lactic dehydrogenase, electrophoresis

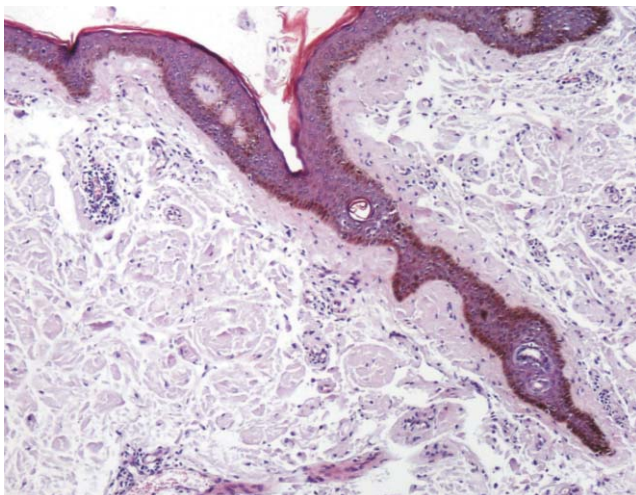


FIGURE 2: Deposition of amorphous eosinophilic material in the dermis (HE 100x)

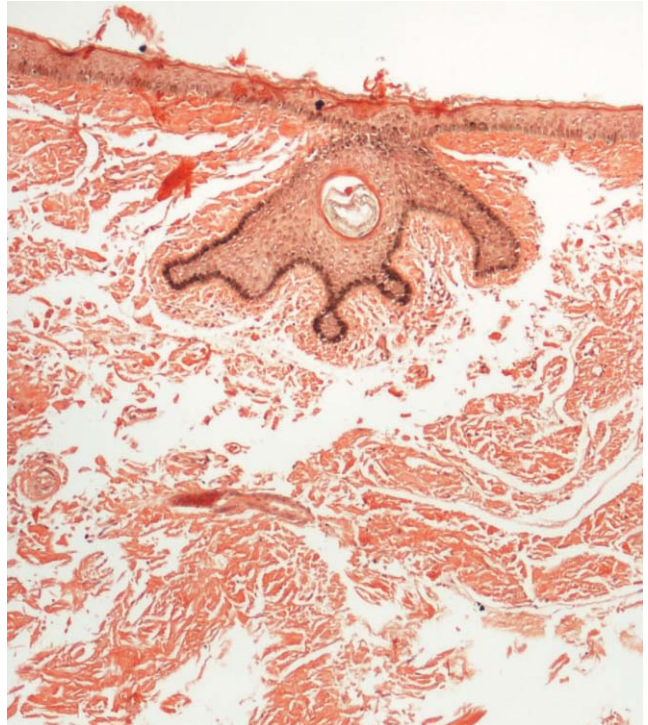


FIGURE 3: Congo red 100x. Staining with Congo red dye showing amyloid material in red

and immunoelectrophoresis of serum and urinary proteins were normal. Twenty-four hour urine protein was slightly altered (170 mg/24 hours – reference value: 140 mg/24 hours). A chest x-ray and electrocardiogram showed no alterations.

The patient was treated with surgical resection of the lesion. After a 2-year follow-up, recurrences have not occurred and the patient exhibits no signs of systemic amyloidosis.

DISCUSSION

Nodular amyloidosis is a rare form of primary cutaneous amyloidosis. It occurs in men and women of any race, who are usually older than 60 years of age.^{3,4} The disease is manifested as infiltrated, isolated or multiple yellowish-erythematous or brownish nodules or plaques in the limbs, face, scalp, trunk and genitals. Occasionally there may be bullae or atrophic lesions in this form and it may be clinically similar to primary systemic amyloidosis. However, the absence of monoclonal plasma cell proliferation in the bone marrow establishes the diagnosis of nodular amyloidosis.⁵

The word corymb in Botany refers to racemose inflorescence in which flowers are situated at the same level due to a gradually shorter length of pedicels along the axis. In Dermatology, corymbiform means a larger central lesion with other satellite lesions, resembling an explosion.² In 1876

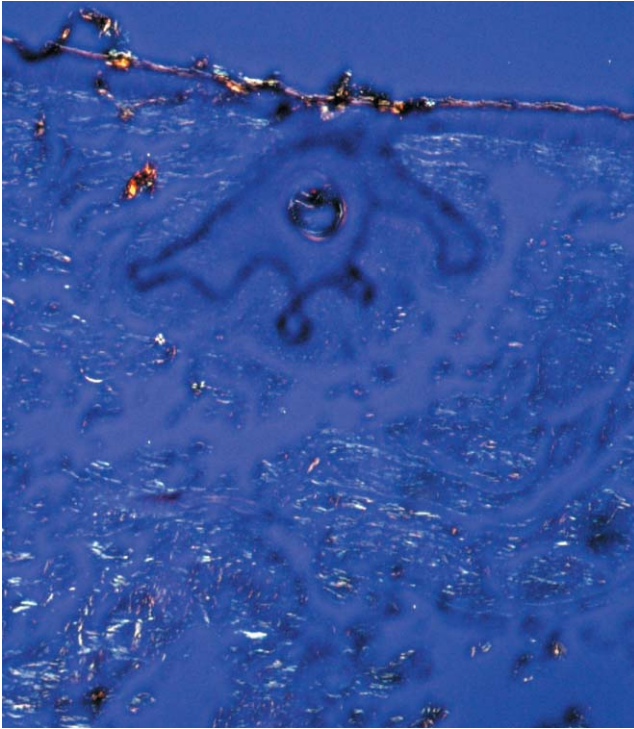


FIGURE 4: Polarizing microscope 100x. Under polarizing microscope, the amyloid material shows apple-green birefringence

Hutchinson stated that eruptions of corymbiform aspect indicated nerve involvement, citing as an example herpetic lesions.⁶ It is known that nerve involvement does not occur in cutaneous amyloidosis. Syphilis is a disease in which corymbiform lesions are most characteristic.⁷

Amyloidoses may be primary or secondary, systemic or localized, hereditary or acquired, making

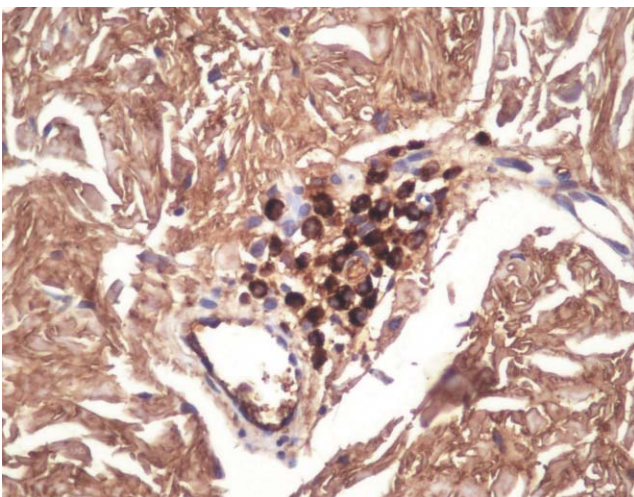


FIGURE 5: Immunohistochemistry with anti-kappa antibody indicating 75% of plasma cells with positive expression in the cytoplasm (IH 400x)

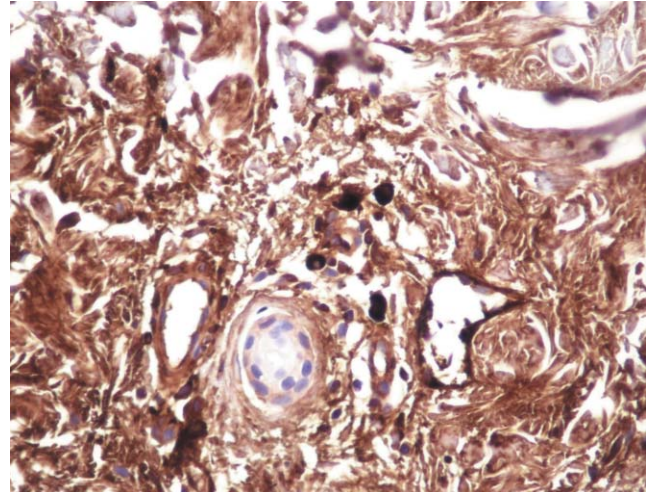


FIGURE 6: IH 100x Immunohistochemistry with anti-lambda antibody showing 25% of plasma cells with positive expression in the cytoplasm (IH 100x)

classification of the disease difficult. Primary systemic amyloidosis is generally associated with lymphoproliferative plasmacytic disease, such as multiple myeloma. In contrast, secondary systemic amyloidosis is correlated with infectious chronic inflammatory processes or malignancies, while hereditary amyloidosis includes various forms of systemic amyloidosis of familial nature. Macular amyloidosis, lichen amyloidosis and nodular amyloidosis are described as forms of primary cutaneous amyloidosis. Secondary cutaneous amyloidosis has no clinical expressions and occurs in melanocytic nevi, tumors, keratoses, etc.^{8,9}

Regardless of the type of amyloidosis, tissue dysfunction occurs due to infiltration of insoluble protein deposits resistant to proteolysis. These deposits are composed of fibrils that suffer a degenerative process, transforming their natural α -helical structure into β -pleated structure, characteristic of amyloid deposition. In nodular amyloidosis, precursor proteins are light chain immunoglobulins.¹⁰ Some authors have defined nodular amyloidosis as an extramedullary plasmocytoma in which amyloid fibrils are locally produced by plasma cells. Although it has been suggested that patients with nodular amyloidosis have plasma cell dyscrasia, there is no evidence of paraproteinemia.¹

Clinical differential diagnosis of nodular amyloidosis was made with lymphomas, pseudolymphomas, sarcoidosis, facial granuloma, reticulohistiocytoma and multicentric reticulohistiocytosis.⁸ Due to the corymbiform aspect of the lesion, secondary syphilis was also a differential diagnosis.⁷

Histopathologic exam of nodular amyloidosis demonstrated deposition of amyloid material in the dermis, subcutaneous tissue and also around blood

vessels, explaining the capillary fragility that may occur in these lesions. Tissue sections were stained with Congo red dye and observed under a polarizing microscope for the characteristic apple-green birefringence.^{8,10}

Systemic evaluation of these patients should always be performed to rule out involvement of other organs by amyloidosis. Hematologic, kidney and liver function, electrophoresis and immunoelectrophoresis of serum and urinary proteins, electrocardiogram

and chest x-ray are fundamental tests.⁸

At the beginning of the clinical picture, around 40% of patients with primary systemic amyloidosis may have cutaneous lesions similar to those found in nodular amyloidosis. In addition, 7% of patients with nodular amyloidosis may progress to systemic amyloidosis.¹⁰

Therapeutic modalities include surgical resection, electrosurgery, cryosurgery and CO₂ laser, although recurrence is common.^{1,4} □

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