Localized primary cutaneous nodular amyloidosis - Case report *

Amiloidose localizada cutânea primária nodular - Relato de caso

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Abstract: Amyloidosis results from deposition of fibrous and insoluble amyloid protein in extracellular spaces of organs and tissues. Amyloid deposition can be localized or systemic and either primary or secondary. We report a case of localized primary cutaneous nodular amyloidosis manifested by papular-nodular, reddish-brown lesions affecting the nasal area, without evidence of systemic involvement. Immunohistochemistry showed the presence of immunoglobulin kappa light chain.
Keywords: Amyloidosis; Plasma cells; Skin

Resumo: A amiloidose resulta da deposição de proteína amiloide fibrosa e insolúvel em espaços extracelulares de órgãos e tecidos. O depósito da substância amiloide pode ser localizado ou sistêmico e pode ser de natureza primária ou secundária. Relataremos um caso de amiloidose localizada cutânea primária nodular, manifesta por lesões pápulo-nodulares, eritêmato-acastanhadas, acometendo a região nasal, sem evidência de acometimento sistêmico. O estudo imunoistoquímico demonstrou presença de imunoglobulinas de cadeia leve *kappa*.
Palavras-chave: Amiloidose; Pele; Plasmócitos

INTRODUCTION

Amyloidosis is a generic term used for a group of diseases characterized by deposition of a substance chiefly composed of fibrous protein, called amyloid, which may produce compression and/or dysfunction of several organs, among them the skin. These diseases may be divided into systemic or localized forms. The localized forms may be primary or secondary. The primary cutaneous forms include macular amyloidosis, lichen amyloidosis and nodular or tumefactive amyloidosis. Systemic amyloidosis can be primary, hereditary or not, secondary to chronic inflammatory diseases or neoplasias and be associated with hemodialysis. The distinction between primary localized cutaneous amyloidosis and the systemic forms should be made through a careful physical examination and supplementary tests to rule out the presence of extracutaneous amyloid deposits and/or plasmacyte dyscrasias.[1,2,3]

We present a case of primary cutaneous nodular amyloidosis localized on the nose. Immunohistochemical screening demonstrated mainly the presence of immunoglobulin *kappa* chains close to groups of plasmacytes. The patient did not demonstrate any systemic involvement signs in a one-year follow-up. This disease should always be considered in differential diagnosis of papulous or nodular face lesions.[4]

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CASE REPORT

A male, 39-year-old patient referred the onset of nose lesions for a period of 2 years. He denied relevant pathological antecedents. Upon examination, erythematous papulonodular lesions were observed in groups on the top of the nose and right nasal wing (Figure 1).

A cutaneous biopsy was performed and the histopathological exam revealed rectified epidermis and masses of eosinophilic amorphous material occupying the entire extension of the dermis, with compression of cutaneous adnexa and surrounding vessels (Figures 2 and 3). Congo-red staining revealed a mass of orange amorphous material occupying the dermis and hypodermis diffusely; the diagnosis of cutaneous amyloidosis was made. The immunohistochemical screening performed at the Dermatology Departament, University of Graz (Austria) with the immunoperoxidase technique, using monoclonal antibodies from Dako laboratory (Glostrup-Denmark) showed strongly positive immune marking for immunoglobulin *kappa* chain and with less intensity for *lambda* chain. It also demonstrated plasmacyte clones infiltrate around the vessels (Figures 4 and 5). Immune marking for cytokeratins was negative (Figure 6). The supplementary tests: hemogram, lipid profile, liver and kidney function, 24-hour proteinuria, protein electrophoresis, urinalysis type I, antinuclear factor, rheumatoid factor, HIV and B and C serologies did not show abnormalities. The patient is being followed at the outpatient clinic (clinical and laboratory evaluation twice a year), without any evidence of systemic involvement. The lesions will be surgically removed.

DISCUSSION

Localized primary cutaneous nodular amyloidosis is the rarest cutaneous presentation of amyloidosis. It is characterized by the diffuse deposition of amyloids in the dermis, subcutaneous tissue and small vessels in the dermis. The amyloid substance deposited is of the AL type, which is also found in primary systemic amyloidosis and in the form associated with multiple myeloma, composed of immunoglobulin light, *kappa* and *lambda* chains. It is considered a form of extramedullary plasmacytoma.[5]

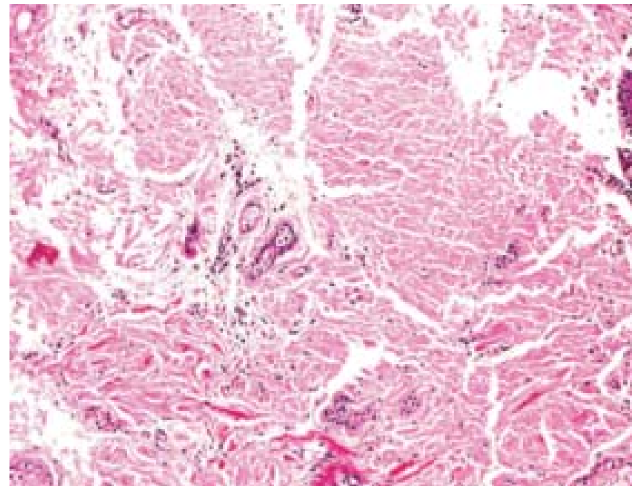

FIGURE 2: Deposit of amorphous material occupying the entire dermis (HE 10x0.25)

The patients present asymptomatic nodules or plaques, single or multiple, rose-brown in color with a tendency to involve the face, mainly the nose and periauricular areas, genitals, trunk and limbs. These lesions are similar to those found in primary systemic amyloidosis associated with lymphoproliferative plasmacytary disease.[6]

It occurs equally in both genders, with mean age at diagnosis of 60.8 years. The diagnosis of the dis-

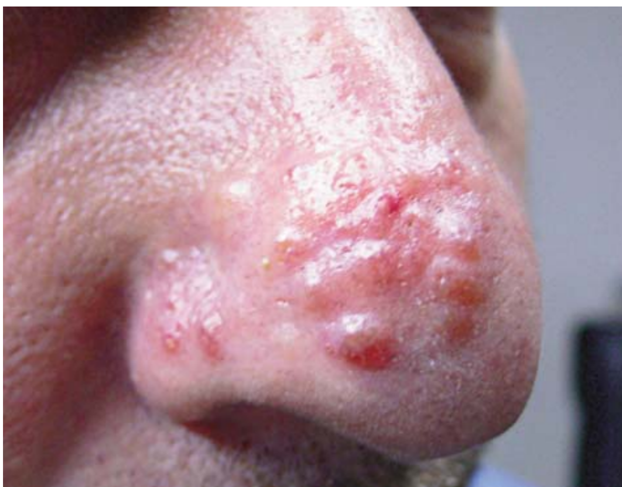

FIGURE 1: Papulous lesions grouped on the tip of nose

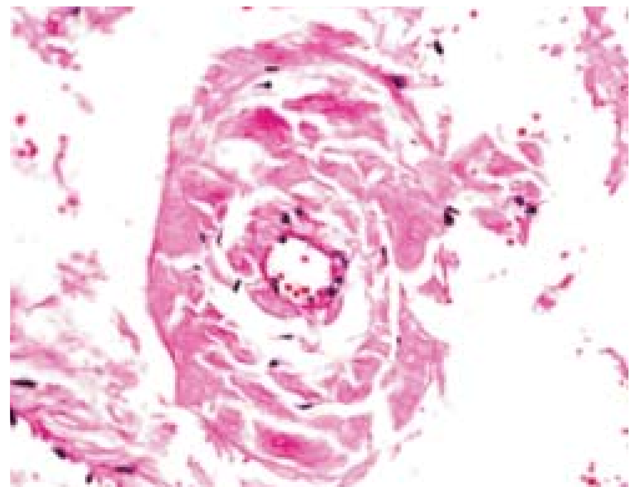

FIGURE 3: Deposit of amyloid material surrounding vessels (HE 10x0.3)

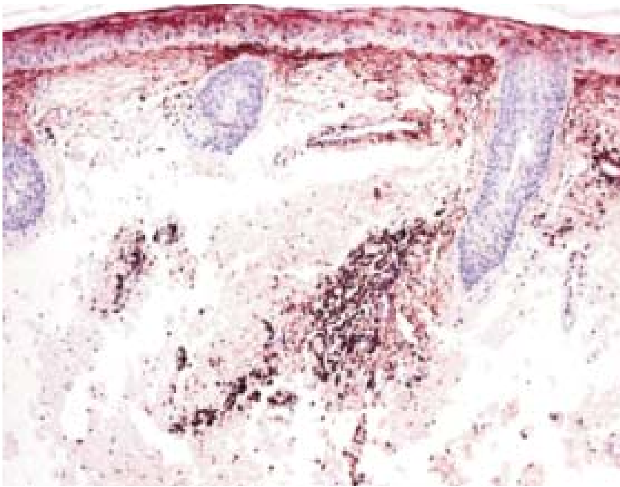

Figura 4: Plasmacytes and highly positive immune marking of the *Kappa* chain demonstrate monoclonality (immune marking utilizing monoclonal antibodies 10x0.4)

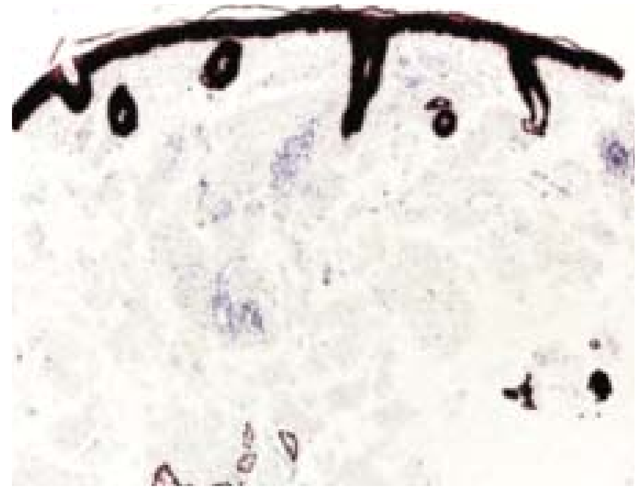

Figure 6: Immune marking of cytokeratins with negative result

ease is often late and the mean time from onset of lesions to diagnosis is 13.5 years.[5]

The diagnosis is established through skin biopsy, when a diffuse deposit of amyloid substance is observed subcutaneously in the dermis, on the walls of small vessels. A plasmacyte infiltrate is also found in perivascular disposition. The amyloid substance is better visualized in routine Congo-red staining and immunohistochemistry reveals monoclonal plasmacytes and immunoglobulin *kappa* and *lambda* chains.[4,7] Cytokeratins are not present.[8] The process may have a chronic, localized and benign course, but the patients must be monitored regarding progression to systemic amyloidosis and plasmacyte dyscrasias, which occur in 7 to 50% of patients.[7,9] There are reports of association with Sjögren syndrome and diabetes mellitus. The clinical lesions, the demonstration of amyloid substance by Congo-red staining and the presence of monoclonal chains of immunoglobulin light chains, mainly *κappa,* found in the studied case, favors the hypothesis of type AL, that includes the nodular variant of the localized primary cutaneous form, primary systemic amyloidosis and systemic secondary amyloidosis associated with multiple myeloma, monoclonal gammopathy or plasmacytary discrasia.[7] As no systemic alterations were detected, the final diagnosis was localized primary cutaneous nodular amyloidosis. The fact that there was no evidence of deposition of cytokeratin filaments rules out other clinical forms of cutaneous amyloidosis. The patient is being monitored regarding progression to systemic amyloidosis and the presence of plasmacytary discrasias. Although all forms of treatment present a high rate of local recurrence, a joint decision was made with the patient for surgical excision of lesions to improve his physical appearance. [10, 11] ❑

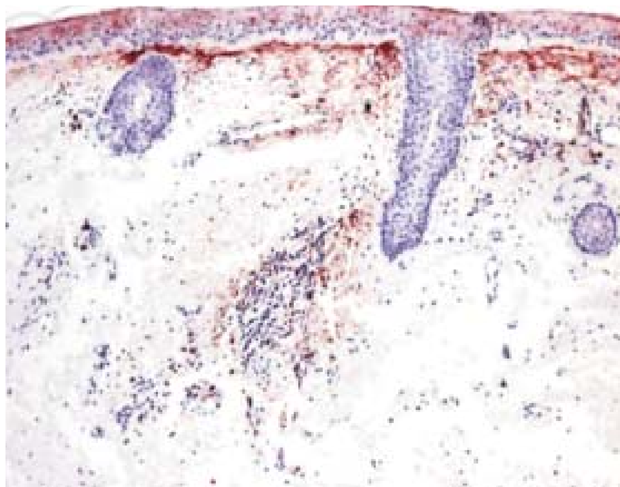

Figure 5: Plasmacytes and weak expression of the *lambda* chain (immune marking utilizing monoclonal antibodies – 10x0.4)

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