An Unusual Presentation of Primary Skin Amyloidosis

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Authors’ contributions

This work was carried out in collaboration among all authors. Authors R. Ayari and MAS designed the study, performed the statistical analysis, wrote the protocol and wrote the first draft of the manuscript. Authors R. Amri, KA and FBD managed the analyses of the study. Author RT managed the literature searches. All authors read and approved the final manuscript.

Article Information

Editor(s):
(1) Dr. Kaushik Bhattacharya, CAPFs Composite Hospital, India.

Reviewers:
(1) Nwagu Marcellinus Uchechukwu, Edo University Iyamho, Nigeria.
(2) Shigeki Matsubara, Jichi Medical University, Japan.

Complete Peer review History: http://www.sdiarticle4.com/review-history/56403

Received 10 February 2020
Accepted 17 April 2020
Published 25 April 2020

ABSTRACT

Amyloidosis is a group of diseases in which abnormal proteins, known as amyloid fibrils, build up in tissue. Primary localized cutaneous amyloidosis is a form limited to the skin without involving any other localization. An amorphous material composed of amyloid is produced and deposited in the dermis, with a varied clinical presentation. Nodular amyloidosis is a rare presentation of primary localized skin amyloidosis. We report an unusual clinical presentation made of bilateral nodular lesions, mimicking a skin tumor. Anatomopathological study showed primary localized nodular skin amyloidosis. Surgical excision with reconstruction was performed in view of the unsightly appearance of the lesions. Regular monitoring is necessary to detect any potential systemic progression of the disease. The patient uneventfully recovered without recurrence at the one-year follow-up.

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Keywords: Amyloidosis; primary localized cutaneous amyloidosis; nodular amyloidosis; bilateral nodular amyloidosis; cutaneous amyloidosis.

1. INTRODUCTION

Amyloidosis is a large group of diseases defined histologically by the presence of fibrillar proteins, arranged in a pleated sheet with a beta spatial configuration. The extracellular deposition of insoluble proteins, called amyloid deposition, tends to invade and destroy various tissues. The anatomopathological study allows the diagnosis of amyloidosis and helps characterize its biochemical nature with immunohistochemistry.

Amyloid deposition can be built up in a single organ, such as the skin without any systemic involvement. In case of cutaneous amyloidosis, there are three categories: primary localized cutaneous amyloidosis, secondary localized cutaneous amyloidosis of skin tumors and systemic amyloidosis with cutaneous involvement. Primary localized cutaneous amyloidosis can have several forms. Macular amyloidosis, Lichen amyloidosis and biphase amyloidosis are the most common variants [1,2]. Nodular amyloidosis is a rare presentation accounting for approximately 1.5% of the cases [3,4,5].

2. CASE PRESENTATIONS

We report the case of a Tunisian 76-year-old patient with no medical history presented on February 2011 with a 03 month-history of two painless bilateral jugal lesions. Physical examination showed nodular firm lesions, retracting the surrounding skin with no underlying ulceration or infiltration of the subcutaneous plane. They had the same skin color with a smooth surface measuring approximately 1 x 1.4 cm. The edges of the lesion were clearly delimited (Fig. 1).

There was no other associated lesion.

A skin biopsy was performed extending to the surrounding normal skin.

The histopathologic examination demonstrated scattered plasma cells and amorphous pink material in the dermis and subcutis that was apple-green birefringent with Congo-red staining and polarized light, which is consistent with nodular amyloidosis (Fig. 2).

The immunohistochemical study concluded with non-AA amyloidosis.

The following investigations were performed: a hemogram showing a hemoglobin level of 12.6 g/dl, white blood cells count of 5700 and a platelet count of 21000. A renal and hepatic laboratory assessment, as well as an immunoelectrophoresis of the blood and the urine, a chest radiography, a biopsy of the abdominal fat, cardiac and abdominal ultrasound were also performed. All of them were negative. Therefore, the diagnosis of nodular primary localized cutaneous amyloidosis was retained. A total bilateral excision with reconstruction was made within the same day at the request of the patient who was bothered by the unsightly aspect of the lesions with a good result (Fig. 3).

Follow ups were made regularly. There was no sign of disease progression in the past year.

Fig. 1. Morphological structure of primary skin amyloidosis
3. DISCUSSION

The etiopathogenesis of primary localized cutaneous amyloidosis is not yet fully known. Numerous hypotheses were made such as: a genetic predisposition, prolonged friction and environmental factors mainly a viral infection.

The precursor protein is not fully identified. During macular and Lichen amyloidosis, the amyloid is probably derived from keratinocytes [6]. On the other hand, the origin of the nodular amyloid is very different, it is formed from light chains of immunoglobulin. This suggests a plasma cell origin, like the ones of the nodular skin lesions of primary systemic amyloidosis [7,8].

The main clinical presentation of the nodular primary localized cutaneous amyloidosis is a
single or more frequently multiple nodules on the face, trunk or limbs. Their size is very variable, ranging from a few millimeters to several centimeters.

Sometimes, it can be presented in the form of infiltrated plaques, or a solitary localized deposit mimicking a tumor, called tumoral amyloidosis. Amyloidoma is the least common presentation of tissue amyloid deposits [9] which is presented by our patient. It is very poorly described in the literature [10-13].

The diagnosis is based on the histological study, associated with a positive staining with Congo red.

Nodular primary localized cutaneous amyloidosis is often successfully treated surgically. Various therapeutic means were described in the literature such as surgical excision, cryotherapy, electro-desiccation, curettage and cauterization [14], dermabrasion [15] and laser ablation.

The evolution of nodular primary localized cutaneous amyloidosis is frequently prolonged but benign (8). Nevertheless, patients should be regularly screened to detect the evolution to a systemic disease (paraproteinemia and overt systemic amyloidosis), which can occur in 5 to 50% of the cases [16]. However local recurrences are frequent [5].

4. CONCLUSION

We reported this case of nodular primary localized cutaneous amyloidosis, in order to highlight this rare entity. We recommend skin biopsy for any suspicious nodular lesion to improve the understanding of this rare entity.

Close monitoring of patients is useful since this primary localized form may be the precursor of a systemic amyloidosis.

CONSENT

We obtained informed consent of the patient to share his case and photos in this study.

ETHICAL APPROVAL

As per international standard or university standard written ethical approval has been collected and preserved by the authors.

ACKNOWLEDGEMENT

We thank Dr Mohamed Ali Sbai head of the department of Plastic, Hand Surgery and Burns at the Maamouri Hospital, Nabeul, Tunisia for his great help and supervision.

COMPETING INTERESTS

Authors have declared that no competing interests exist.

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