

First report of hemosiderotic adenodermatofibroma in Brazil

Primeiro relato de adenodermatofibroma hemossiderótico no Brasil

Miguel Augusto M. Pereira; Luciana Pantaleão; Mayra C. Rochael

Universidade Federal Fluminense (UFF), Niterói, Rio de Janeiro, Brazil.

ABSTRACT

Hemosiderotic adenodermatofibroma is a recently recognized lesion, characterized by a dermal nodule with cystic structures of an apocrine gland, surrounded by a dermatofibroma-like stroma with hemosiderotic macrophages. We present the first case report of this entity in Brazil together with representative images, in addition to a review on the subject and discussion about the apocrine origin of this lesion.

Key words: benign fibrous histiocytoma; hemosiderin; dermis.

RESUMO

Adenodermatofibroma hemossiderótico é uma lesão recentemente conhecida, caracterizada por um nódulo dérmico com estruturas císticas de uma glândula apócrina, circundado por um estroma do tipo dermatofibroma com macrófagos hemossideróticos. Apresentamos o primeiro relato de caso dessa entidade no Brasil junto a imagens representativas, além de uma revisão do assunto e discussões sobre a origem apócrina dessa lesão.

Unitermos: histiocitoma fibroso benigno; hemossiderina; derme.

RESUMEN

El dermatoadenofibroma hemossiderótico es una lesión recientemente descrita, caracterizada por un nódulo dérmico con estructuras quísticas de una glándula apócrina, rodeado por un estroma del tipo dermatofibroma con macrófagos hemossideróticos. Presentamos el primer reporte de caso de esa entidad en Brasil, junto a imágenes representativas, además de una revisión del tema y discusiones acerca del origen apócrina de esa lesión.

Palabras clave: histiocitoma fibroso benigno; hemossiderina; dermis.

INTRODUCTION

Adenodermatofibroma is a lesion recently recognized by the medical community, characterized by dermatofibroma-like stroma, with histiocytes, fibroblasts as well as glandular structures with apocrine characteristics. The entity, considered a mixed tumor, was presented for the first time in 2005, by the Chilean

Sergio González, to the American Journal of Dermatopathology, in the report form of two cases with a descriptive diagnosis. It was a dermal nodule with cystic structures of an apocrine gland, surrounded by a dermatofibroma-like stroma with hemosiderotic macrophages⁽¹⁾. In 2013, the adenodermatofibroma nomenclature was proposed by Santos-Briz *et al.* (2013)⁽²⁾ to The American Journal of Dermatopathology.

We describe here the first case of adenodermatofibroma in Brazil and one of the few cases from the world medical literature, with hemosiderotic characteristics, according to a review of the US National Library of Medicine National Institutes of Health (PubMed) and the Scientific Electronic Library Online (SciELO) databases.

CASE REPORT

A 46-year-old man was presented to the clinic with an asymptomatic nodular lesion of approximately 10 cm in the leg, with no signs of inflammation. There was no surgical history. Histopathological examination of the skin revealed a poorly delimited, unencapsulated nodule with cystic spaces (epithelial lining and apocrine differentiation) with proteinaceous content and solid areas. The surrounding stroma is composed of a mixture of fibrocytes and macrophages with large hemosiderin granules, hemorrhage foci, and dense fibrous stroma with typical storiform areas, constituting an apocrine cyst with a similar hemosiderotic dermatofibroma stroma (**Figures 1 and 2**). Vessel wall thickening was identified. There were no mitoses, atypia, or necrosis.

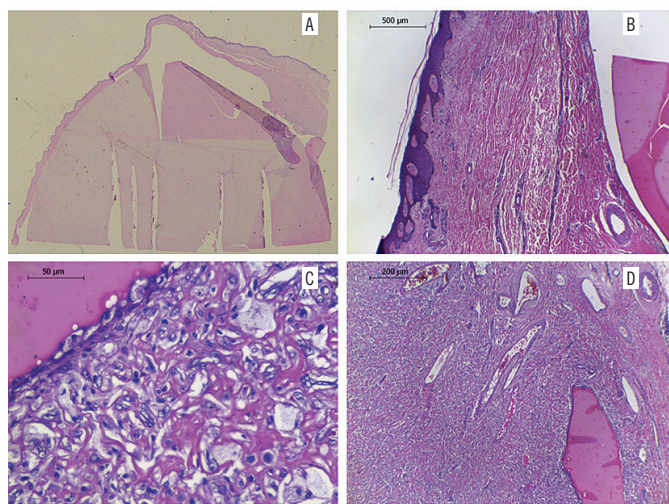


FIGURE 1 – A) panoramic view of the lesion; B) lower magnification showing the involvement of the epidermis (acanthosis) and dermis (HE, 40×). Note the stroma with dense appearance and deeply a cystic space with proteinaceous content (not fully represented); C) higher magnification showing in detail the apocrine epithelium and the stroma represented by macrophages, fibrocytes, and collagen fibers. Note that the wall of the glandular structure is composed of two layers, and the vacuolated inner cells protruded into the lumen (HE, 100×); D) cystic space with proteinaceous content surrounded by the fibrohistiocytic component of the tumor. Detail of the presence of congested venules with a thickened muscular layer (HE, 40×); the lesion had many blood vessels
HE: hematoxylin and eosin.

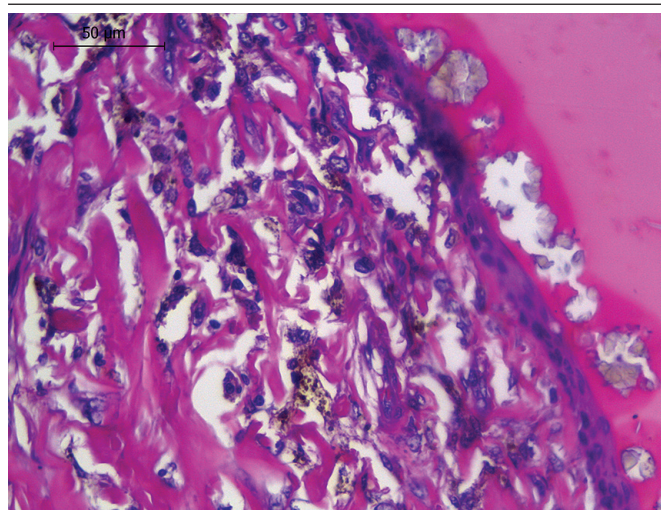


FIGURE 2 – Higher magnification showing the limit between the fibrohistiocytic stroma and the wall of a glandular structure composed by layers of a non-keratinizing squamous epithelium. Note the presence of hemosiderin inside the cells (HE, 400×)
HE: hematoxylin and eosin.

DISCUSSION

The first reports – Gonzalez (2005)⁽¹⁾ – were on a lesion on the back of a 52-year-old woman with a clinical diagnosis of lipoma, and a 41-year-old man with a nodule on the forehead, considered as an epidermal inclusion cyst. Both were histologically very similar: ovoid dermal nodules, smaller than 20 mm in diameter, with hemorrhagic foci as well as macrophages with cytoplasmic inclusions of hemosiderin, not encapsulated, poorly delimited, composed of cystic spaces filled with proteinaceous content, solid areas, and thickened muscle layer of vessel walls.

However, there are still possible variations of the same cutaneous lesion, but without hemosiderosis⁽²⁾, and even with no epithelial cyst component with apocrine features^(3, 4).

Several authors believe that adenodermatofibroma is a true mixed tumor rather than an induction phenomenon^(5, 6). A tumor with an epithelial component characterized by an apocrine differentiation cyst and another mesenchymal component, due to the hemosiderotic dermatofibroma⁽⁵⁾. Thus, the creation of a new entity, rather than a variant of dermatofibroma or an apocrine cystadenoma with a reactive stroma, was advocated⁽⁵⁾.

This case favors the argument of a mixed tumor, since we observe apocrine glandular tissue far from its usual topography, making it unlikely that its origin is by collision or by incarceration of the apocrine gland^(1, 5). Besides, the intimate mixing, in different proportions, of the epithelial and mesenchymal components, also

favors a more neoplastic origin than a reaction to the stroma, besides contributing to discard the possibility of a collision of tumors by chance, which would present transition points between the tissues⁽⁵⁾.

The patient has a leg lesion at the same anatomical region as the case presented by Allen in the “Arkadi M. Rywlin International Pathology Slide Seminar” in 2008⁽⁷⁾. Gonzalez’s reports the lesions were on the forehead and back. Similar lesions were already reported on the pubic area and on the left scapular region⁽²⁾. All considered as sites devoid of apocrine glandular tissue.

The most recent case was published in 2018 in the Journal of Cutaneous Pathology by Muto *et al.* (2018)⁽⁸⁾ and brings a 67-year-old woman with a 4 cm symptomatic nodule (pruritus and pain) on the back. For the first time the hypothesis that glandular structures of the lesion originate from incarcerated eccrine glands was presented, which is apocrine metaplasia. According to the authors when considering the eccrine or apocrine nature of the adenomatofibroma, it is necessary to consider the location of the lesion as well as the characteristics of the glandular structures contained in the area of the lesion⁽⁸⁾.

CONCLUSION

To conclude, we describe the first case of an adenomatofibroma in Brazil, one of the few recorded in the medical literature. We believe that the lesion in this report deals with a true mixed neoplasm, based on the topography of the lesions, which do not present apocrine glands. Thus we discard the hypotheses of dermatofibroma induction phenomenon, random collision or apocrine metaplasia. We noted from the literature the confusion with the clinical diagnosis of lipoma. Also, the lack of recurrence, the absence of mitoses, atypia or necrosis reinforces the benign nature of the lesion in the cases.

The presence of these several interrelated components and the discussions about the origin of this lesion make this variant of a simple diagnosis such as dermatofibroma, rare and beautiful. However, it is not yet possible to postulate risk factors or other epidemiological characteristics of the disease due to the reduced number of cases, therefore, further studies are needed to consolidate this diagnosis.

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CORRESPONDING AUTHOR

Miguel Augusto Martins Pereira  0000-0001-5646-2253
e-mail: mappereira@icloud.com



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