CASE REPORT

Cutaneous myoepithelioma: A rare challenging lesion–A Case report and Literature review

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ABSTRACT

A case of cutaneous myoepithelioma arising in the plantar area in a 56-year-old man is presented. The morphology and immunohistochemical profile are described. The literature on the subject is analyzed.

Key Words: Myoepithelioma skin, Cutaneous myxed tumor

1. INTRODUCTION

The common ectodermal origin makes it possible, albeit rarely, for characteristic neoplasms of the salivary glands type to appear in the skin. Therefore the observation of a case of Cutaneous Myoepithelioma (ME)is considered worthy of being reported.

2. CASE PRESENTATION

A 56-year-old man, has for some time presented a small subcutaneous swelling in the left plantar region, slightly painful. In the clinical suspicion of a fibromatous lesion, on an outpatient basis the nodule is removed with excisional biopsy. The nodule measured 1.5 cm × 1 cm.

2.1 Materials and methods

The surgical specimen, 1.5 cm × 1 cm in size, of hard-elastic consistency, with a translucent section surface, of a pearly white color, is fixed in toto in buffered formalin and embedded in paraffin. Microscopic sections are stained with Hematoxyl-Eosin. A panel of antibodies for immunohistochemistry is also used comprising: CK AE1-AE3, S100, AGFAP, SMACT.

2.2 Histology

Located in the mid-deep dermis. It is an ovoid, nodular, growth bound by a thick fibrous capsule (see Figure 1a). On the surface, a strongly cornifying epidermal festoon can be recognized. The bulk of the nodule consists of a proliferation of epithelioid elements arranged in trabeculae (see Figure 1b), separated from a myxoid matrix (see Figure 1c). The epithelioid elements have a globose-polyhedral shape, sharp edges and acidophilic cytoplasm. The nucleus is rounded, rather voluminous, sometimes central, often peripheral giving the element a plasmacytoid appearance. There is no appreciable atypia or abnormal mitotic activity (see Figure 1d).

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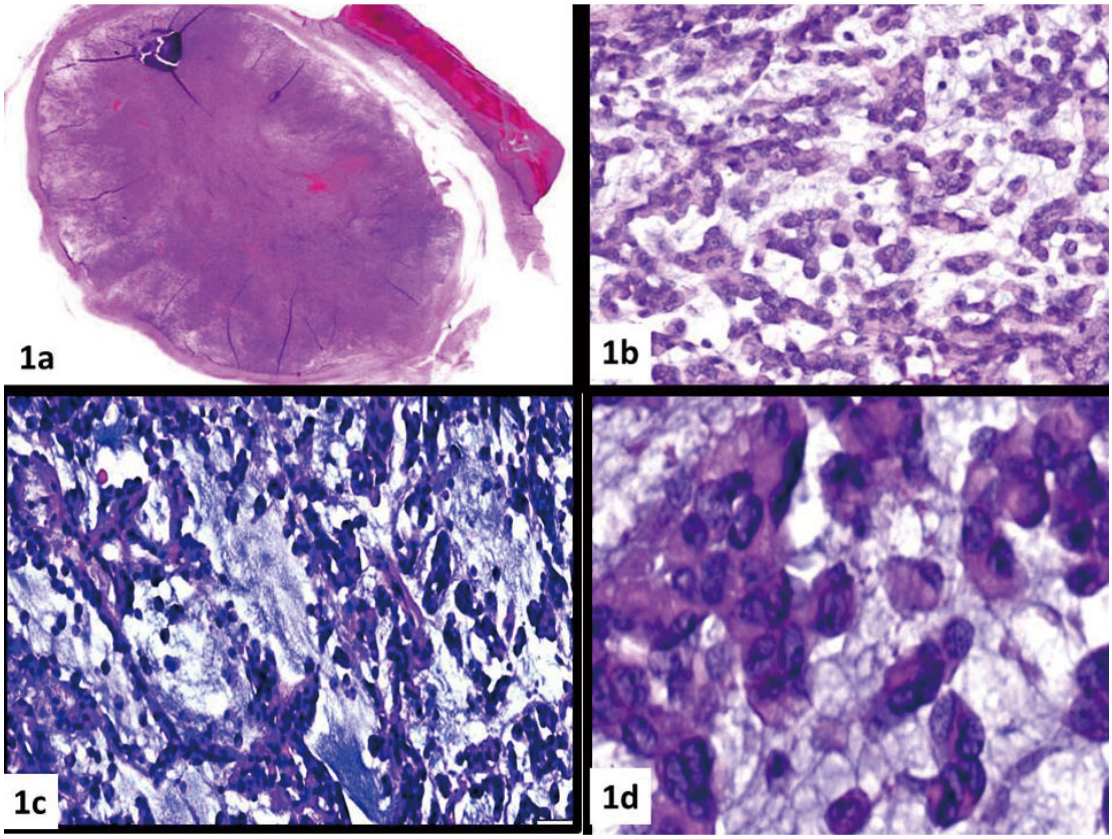

Figure 1. a. Overall view of the nodule at low magnification Note the distance from the overlying epidermis (HE 7X); b. Trabecular pattern of proliferation (HE125X);c.Intertrabecular myxoid matrix (HE 12x); d. Epitheiod and Plasmocytoid cells(HE 175X).

2.3 Immunohistochemistry

The immunohistochemical investigation demonstrated intense and widespread expression for CK AE1-AE3 (see Figure 2a), S100 (see Figure 2b) and GFAP antibodies (see Figure 2c). SMACT presented weak and focal positivity (see Figure 2d). Based on the morphological pattern and the immunophenotypic profile, the diagnosis of Cutaneous Myoepithelioma (ME) was formulated.

3. Discussion

Before starting the discussion, it should be noted that the neoplasm in question does not refer to the tumor described as Myoepithelioma by Lever and Castelman, subsequently named Clear Cell Hidradenoma or Eccrine Acrospiroma.[1] Skin adnexal tumors in which a myoepithelial component is present have long been known as chondroid syringomas or mixed skin tumors, analogous to mixed salivary tumors commonly referred to as pleomorphic adenomas.

The cutaneous ME to which we intend to refer are those neoplasms made up exclusively of myoepithelial cells that faithfully repeat the morphology of Salivary Gland Myoepithelioma which is defined as follows: "benign epithelial tumor composed of sheets and islands of various proportion of spindle, plasmacytoid, epithelioid and clear cells that exhibit myoepithelial, but not ductal differentiation. These tumors sometimes have abundant, acellular mucoid stroma, but lack chondroid or myxochondroid foci".[2]

Between 1998 and 2004, a series of publications showing lesions with morphological characteristics identical to those of the aforementioned salivary neoplasm appeared in the literature. The first report refers to a single case.[3] The second report 12 cases in which 4 tumors involved the dermis and superficial parts of the subcutis and 8 extended deep into the subcutaneous soft tissue.[4] The third concerns 5 cases retrieved from files of 4 different institutions, which produces in-depth analysis of the morphological and immunophenotypic characters of the lesion. According to the authors, these tumors “may be considered the monophasic variant of cutaneous mixed tumors, i.e., purely the myoepitheliomatous variant of cutaneous mixed tumors. Probably there exists a morphologic continuum of cutaneous myoepithelial neoplasms ranging from predominantly ductal neoplasms or cutaneous mixed tumors to pure myoepitheliomas of the skin, which are mostly solid neoplasms with little or no evidence of ductal differentiation.”[5]

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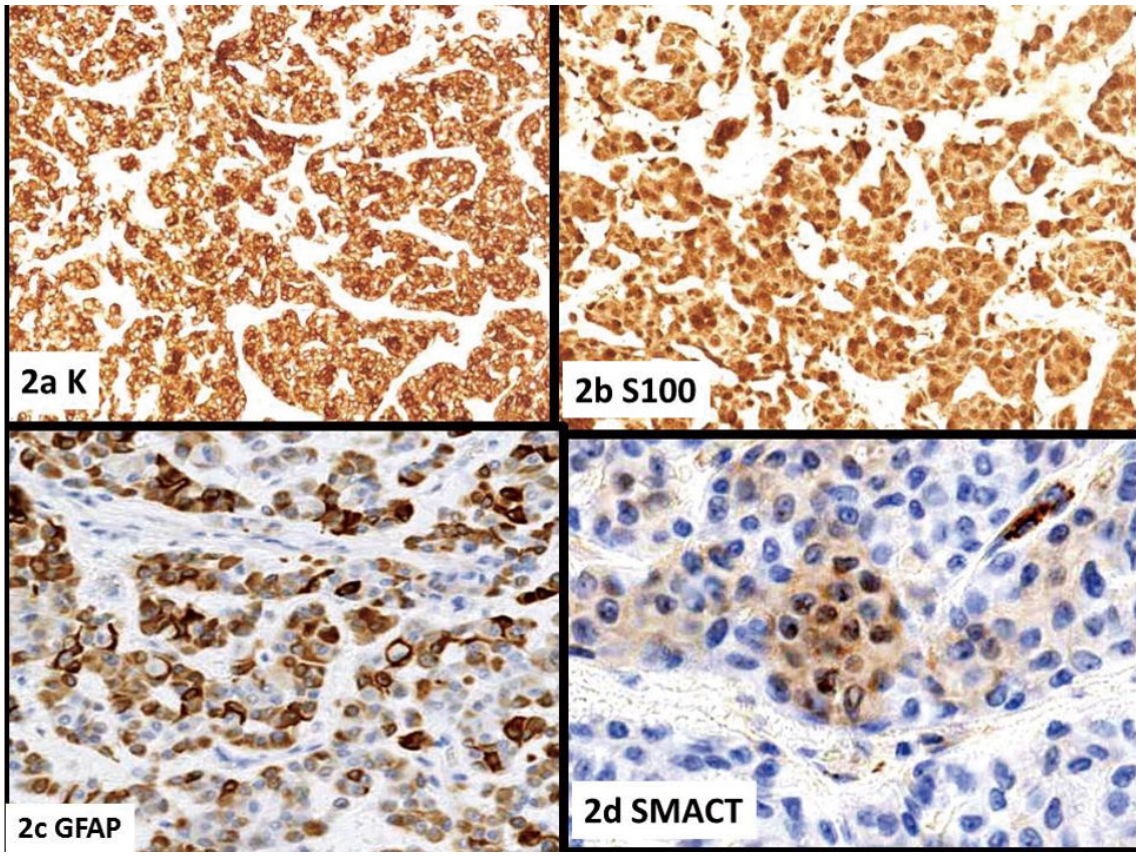

Figure 2. a. Cytokeratin (135X); b. S100 (125X); c. GFAP 125X); d. SMACT (125X).

A further study on 20 cases (9 chondroid syringomas, 9 myoepitheliomas and 2 myoepithelial carcinoma), as well as prove that the chondroid syringoma is a dermal epithelial/myoepithelial proliferation, also demonstrates that, albeit rarely, the proliferation can also be exclusively constituted by myoepithelial cells, without tubular component (ME). The detection of myoepithelial proliferation nodules in the context of typical chondroid syringomas would indicate that the chondroid syringoma and ME represent two different aspects of the morphological spectrum of myoepithelial proliferations.[6] In 2004 a further series of 14 cases is published (some of which have already been published in the previous report) with similar conclusions.[7] On the immunohistochemical level, the authors agree that these tumors have a net positivity for EMA, S100, Cytokeratin, variable for myogenic markers and GFAP.

All these publications show that these tumors are uncommon dermal lesions that typically involve the extremities of children and adults. They would be part of a spectrum of lesions with myoepithelial differentiation ranging from mixed tumors rich in myoepithelial cells to myoepitheliomas to myoepithelial carcinomas.

The cytoarchitectural aspects of these lesions are those of solid proliferation of ovoid, spindle or plasmacytoid cells in an often-abundant myxoid or hyaline stroma. In the aforementioned publications,[5–8] 40 cases of myoepithelioma have been reported. From the data, it is clear that 70% of patients were male and the average age was 41 years with a range between 10 to 93 years. There were five cases with recurrence and one with metastasis and death. There were nine cases (22%) under the age of 20 and 22% of them (2 cases) had relapse representing 33% of all adverse outcome".

After 2004, another 14 case reports concerning cutaneous mucoepithelioma appeared. In this series, the average age was 41 years and male/female ratio was 8/7. 3 patients (21%) were less than 20 years old. One of them had a malignant course.[8–22]

From the examination of these two series, it is clear to note that one out of 5 patients is under the age of 20 and of these 25% presented an aggressive course. This more aggressive attitude of myoepithelial neoplasms in childhood had already been reported previously. Specifying that the superficial lesions, although morphologically innocent, it tend to end up of having the same unfavorable course of the morphologically more typical deep lesions.[23]

The subject of ME of the skin cannot be dealt with without mentioning the Syncytial Variant identified and described by Fletcher as follows, "it is a distinct variant showing solid growth of spindled, ovoid, or histiocytoid cells with palely eosinophilic cytoplasm that more frequently affects men, occurs over a wide age range, and usually presents on the extremities. and incontrovertible to most myoepithelial neoplasms, keratin staining is infrequent. On the basis of preliminary follow-up data, the syncytial variant appears to behave in a benign manner and only rarely recurs locally".[24]

In the nosography of salivary gland neoplasms, the "pure" myoeptelioma is considered a separate entity, distinct from the mixed tumor (pleomorphic adenoma). At the cutaneous level, we tend to consider the Chondroid Syringoma (Cutaneous Myxed Tumor) and ME as different expressions of the same continuum.

In the descriptions of this neoplasm, we tend to consider together those cutaneous and those arising at the level of soft tissues and bone.[25] Research on the genotype of these neoplasms has shown that the genotypic expressiveness of skin and soft tissue ME differs substantially from that of Mixed Salivary Tumors. In the same research, genotypic heterogeneity was found between the various cases of cutaneous and soft tissue ME, although having the same morphological configuration and immunophenotypic expressivity. This latest data would indicate a different histogenetic origin between tumors with the cutaneous onset and those originating at deeper levels.[26]

The case we observed should be placed among the "pure" ME due to the total absence of tubular differentiation. Its morphological pattern is of the trabecular type and the elements are epithelioid, with a modest share of the plasmacytoid type. No atypia or abnormal mitotic activity. An abundant myxoid matrix completes the morphological picture. The tumor is clearly delimited from the surrounding structures by a thick fibrous capsule. The immunohistochemical profile with a clear and widespread positivity for CK, S100, GFAP and a focal and weak for SMACT represent the classic immunophenotypic pattern of the ME.

The differential diagnosis should be made with all epithelioid tumors that arise in the dermo-hypodermic and soft tissues. There is a broad spectrum of lesions, with the closest morphologic lesions being extraskeletal myxoid chondrosarcoma, ossifying fibromyxoid tumor and proximal epithelioid sarcoma. The differentiation between all these lesions must be made on a histochemical and cytogenetic basis, where necessary.

The morphology and delimitation of our case suggest a favorable prognosis, without forgetting that about 18% of histologically benign lesions had a recurrence. It is believed to be due to inadequate excision.

4. Conclusion

The study of this case and related literature indicates that ME it is a non-frequent, but not particularly rare, neoplasm, it presents a very polymorphous morphological picture that makes the differential diagnosis very difficult with other epithelioid neoplasms; Its attribution to the skin or soft tissues is not always easy and, therefore, often arbitrary it is localized in preference to the limbs, while not lacking localizations to the trunk or the head and neck. Is no predominant sex or age; The prognosis is usually good, but in about 1/5 of lesions, histologically benign, they tend to recur when not completely removed. In the pediatric age the neoplasm presents more aggressive behavior.

The case reported by us is morphologically and immunophenotypically in line with the morphological and immunophenotypic parameters indicated for the diagnosis of ME. it is not hazardous to predict a benign behavior, given that the lesion is histologically innocent and completely removed.

The cutaneous ME can be histogenetically inserted between adnexal skin tumors. While for similar tumors arising in the subfascial, intramuscular or even intraosseous site, the histogenetic origin is not yet well defined.

Conflicts of Interest Disclosure

There is no conflict of interest.

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