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Case Report

Recurrent cutaneous myoepithelioma of the scalp case report: management and histological analysis.

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Introduction

Myoepithelial cell neoplasms consists of an uncommon group of tumors. These may be both malignant and benign. Even if rare there is a quite proper characterization. Actually, the best known is the salivary gland myoepithelioma, but recently extra salivary examples have been reported (1).

In this clinical case, a cutaneous location of myoepithelioma (CM) is described. This is an even more under-recognized tumor that generally behaves in a benign fashion. These neoplasms are often misdiagnosed. Furthermore, their features mimic several dermatological diseases. To date, only forty cases of CM have been reported in the literature.

They usually occur in adults, but there are few pediatric case reports which show that this tumor seems to act more aggressively in younger age. The characterization of cutaneous myoepithelial tumors is varied, ranging from benign mixed tumors to myoepitheliomas to myoepithelial carcinomas (2).

Clinically, it presents as a slow-growing, painless, nodular mass typically localized to the extremities. Surgical excision and histological evaluation are required to confirm this uncommon diagnosis. Microscopically, there can be a wide range of cytologic and architectural features. Recently, EWSR1 rearrangement has been described in a subset of soft tissue myoepithelial tumors, whereas the cutaneous counterparts showed this aberration in a minority of cases. (3,13) Cytologic atypia is the solely known predictor of an aggressive behavior in soft tissue tumors. When their histology shows benign characteristics, surgical resection is the most appropriate and definitive modality of treatment for CM tumors. Recurrence after radical surgery is unusual, but may happen in some cases.

Case report

A 26-year-old Caucasian male presented in December 2020 with a whitish and low-growing mass of the scalp. The lesion was completely resected, with clinically free margins. The biopsy was sent for histological evaluation.

It was a multinodular and well-circumscribed mass without any invasive growth. Tumor cells were arranged in tubules and partially dilated, rare trabeculae with low intercellular cohesion in an abundant myxoid stroma (Figure. 1,2).

The single tumoral cell was medium-sized with mild eosinophilic cytoplasm, round nuclei, and small nucleoli (Figure. 3). Mitotic activity was present but low (1/10 HPF). No cytologic atypia was found.

Immunohistochemistry showed a diffuse positivity for cytokeratin AE1-AE3 and S-100, a focal intense positivity for EMA, and Ki67 positivity lower than 5%.

The sample was negative for CAM 5.2 and CK7, CK20, CK14, GFAP, p63, SMA ,CD117,Melan A, SOX10, CD31, and podoplanin. For the detection of a translocation involving the EWSR1 gene Fluorescent In

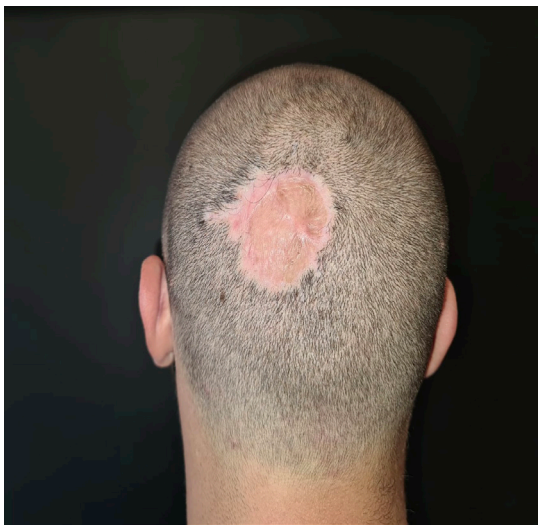

Figure 1: *well-circumscribed tumor with reticular architecture with myxoid stroma*

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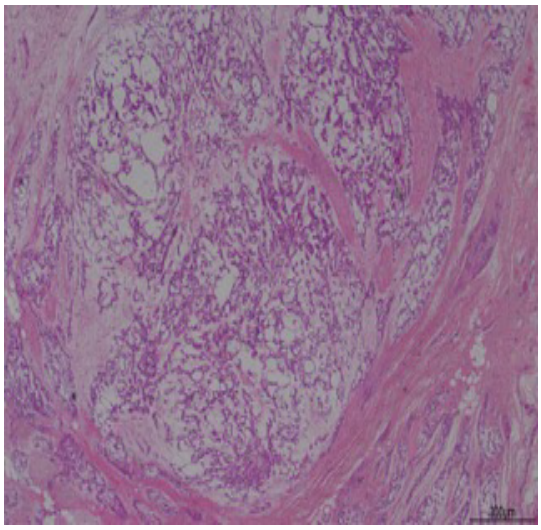

Figure 2: *The tumor is composed of plump epithelioid cells in an abundant myxoid stroma*

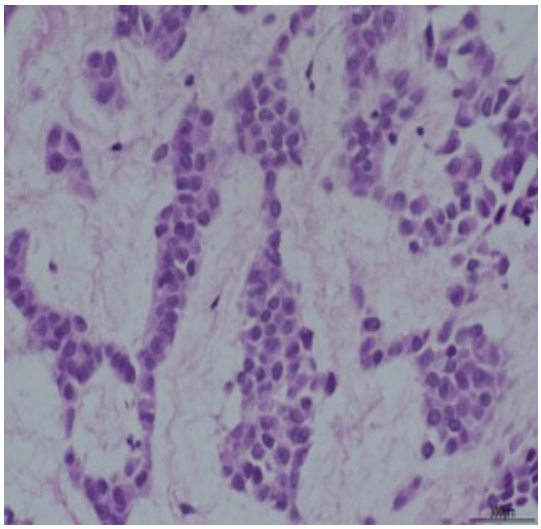

Figure 4: *Three months after second enlargement surgery and acellular dermal matrix covarage*

Situ Hybridization analysis (FISH) was performed on 3 mM sections of formalin-fixed, paraffin-embedded tissue. EWSR1 rearrangement was not found.

The diagnosis was cutaneous myoepithelioma of the scalp, with positive surgical margins. In April 2021, the patient underwent a second surgery during which was performed an enlargement of the previous scar (distance of 0,5 cm and debulking until the galea plane). This second procedure confirmed the identification of recurrent myoepithelioma of the scalp. At this time, surgical margins were clear and showed no evidence of residual tumor cells.

In September 2021, the patient came back for a follow-up and a new clinically evident nodule was discovered just below the previous scar. Therefore, an ultrasound of the area was performed which could not exclude the recurrence of the previous cutaneous myoepithelioma. This suspicion led to a third surgical procedure to remove the new mass with 1 cm clinical margin. The histological examination confirmed the presence of cutaneous myoepithelioma recurrence. At 16 months follow-up from the last surgery no recurrences were found (Figure. 4).

Discussion

Myoepithelial cells are specialized basal cells that derive from the ectoderm. They are found in different tissues (5). In healthy skin, myoepithelial cells are arranged around the terminal portion of the secretory ducts of eccrine and apocrine sweat glands between the basement membrane and the secretory cells. These cells contract to help the extrusion of glandular content during the secretory phase (7–8). Their morphology is variable. It has been shown that myoepithelial cells produce fibronectin, laminin, and collagen. Their immunohistochemical reactivity is related to the lineage of differentiation (myoid or epithelial). Moreover, there cells show variable expression of vimentin, cytokeratins, epithelial membrane antigen (EMA), S-100 protein, muscle actins (SMA), and glial fibrillary acid protein (GFAP).

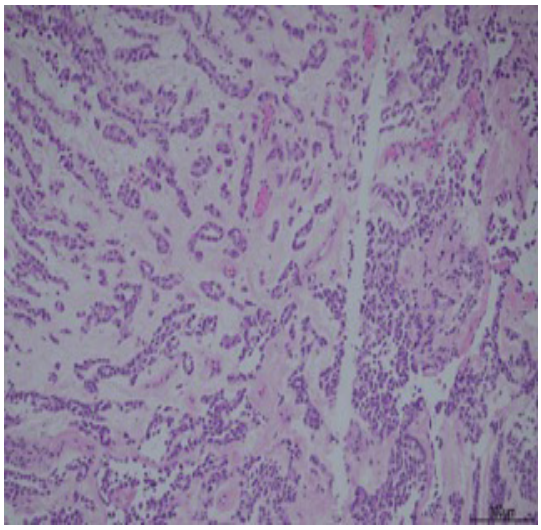

Figure 3: *Single tumoral cell was medium-sized with mild eosinophilic cytoplasm, round nuclei, and small nucleoli; mitotic activity is present but low. No cytologic atypia was found.*

The clinical course of myoepithelioma does not always correlate with their histological characteristics. Most metastatic skin and soft tissue myoepitheliomas showed cellular atypia, high mitotic rate, and infiltrating margins, but a small number of cases without these characteristics showed unusual malignant behavior (9-10). Cell atypia is the only recognized criterion of malignancy and is also the best predictor of malignant behavior. In the absence of atypia, a high mitotic rate, dense chromatin, prominent nucleoli, nuclear pleomorphism, and necrosis denote a greater risk of malignancy (9). The reported CM recurrence rate is 20% and, in general, even after a recurrence, the clinical course is usually favorable. In view of the above, the treatment of choice is complete resection of the lesion with negative margins.

This clinical report is particularly interesting for three reasons. Firstly, it is an extremely rare tumor subtype. Only about 40 cases are reported in literature, the majority of them are benign. The unusual location of this growth is the second notable issue. Myoepithelial tumors are well-defined in the salivary glands, but their occurrence is less represented in the skin. The microscopic finding of tubular organization (reminiscent of salivary gland myoepithelioma) in this patient is also noteworthy. The major aspect to highlight is the recurrence. Cutaneous myoepitheliomas are reportedly not likely to recur once removed with free margins (6;18% if the margins are not free, 1% if the margins are negative) (4). Yet, despite a radical surgery and absence of criterion of malignancy a clinically evident recurrence of disease can be appreciated within just a few months in our patient. It seems to be even more interesting because, from a histopathological point of view, it could be

considered benign (total absence of cellular atypia and mitosis). Therefore, it is probably necessary to search for other criteria which properly predict the behavior of this unusual tumor (5).

Conflicts of interest : No

Funding : None

All patients gave consent for their photographs and medical information to be published in print and online and with the understanding that this information may be publicly available

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