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Therapeutic Management of Hidradenocarcinoma: Case Report and Literature Review

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Abstract **Case Report**

Hidradenocarcinoma is a rare and aggressive carcinoma arising from sweat glands that manifests as an asymptomatic, slow-growing skin tumor. It metastasizes to regional lymph nodes and distant viscera. We report a case of 77 years old man who presented with an invasive hidradenocarcinoma of the scalp without lymph node involvement, treated by wide local excision and Post-operative radiotherapy. The management of this rare entity is not yet well defined. The results are still poor despite the use of multiple treatment modalities.

Keywords: Hidradenocarcinoma, sweat glands, wide local excision, adjuvant treatment.

INTRODUCTION

Hidradenocarcinoma is a rare and potentially aggressive malignant tumor arising from the sweat glands that manifests as a solitary, asymptomatic, slow-growing skin lesion. It is known for its tendency to invade locally and metastasize to distant sites[1].

Usually, it is a so-called de novo carcinoma without any sign of previous hidradenoma, more classically observed in the head and neck [2]; from the age of 40 [3].

The management of this rare entity is not well codified, with poor results despite the use of multiple therapeutic modalities [4]. We report the case of a man with hidradenocarcinoma of the scalp revealed by a progressive lumpy lesion.

CASE

A 77-year-old man, followed for hypertension, presented to the dermatological consultation for an itchy nodular lesion of the scalp which gradually increased in size over a period of 10 years without associated pain, fever or bleeding. Then the patient was referred to us for further therapeutic care.

At the initial examination, it was a median mass at the level of the scalp, with firm consistency, fixed and painless without inflammatory signs which measured 5 cm in diameter.

A cerebral CT was made, objectified the presence of a vertex lytic lesion measuring 47x42 mm, and arrives in intimate contact with the superior sagittal venous sinus whose permeability is preserved, infiltrates the scalp and under arachnoid, with a negative remote extension assessment.

The complement by cerebral MRI was in favor of a tumor process of the scalp in front of the vertex that measure 45x 43 mm, infiltrates outside the soft tissues with budding aspect of the cutaneous surface. This process bombs in endocranial and infiltrates the meninges, compresses the upper sagittal sinus, the superficial veins beside and bulges in the subarachnoid space, without anomaly in the cerebral parenchyma.

The patient underwent a large tumor resection. The anatomopathological study morphological immunohistochemical aspect were in favor of a malignant adnexal cutaneous tumor, hidradenocarcinoma type, which measured 4 cm, with vascular emboli and positive deep tumor limit, without signs of residue or locoregional aggressiveness at the post op MRI.

Surgical revision was judged; by the neurosurgeons; not possible at the level of the positive limit. The patient is scheduled for adjuvant radiotherapy.

DISCUSSION

Hidradenocarcinoma is a rare aggressive intradermal malignancy known for its tendency to invade locally and metastasize remotely, reported for the first time in 1948 in Brazil [5]. It comes from the sweat glands and accounts for less than 0.001% of all tumors [6].

The most common age of presentation is between the fifth and seventh decade of life, the average age of onset is being 50 years with a slight female predominance. Classically it occurs de novo and rarely on pre-existing hidradenoma [7, 8].

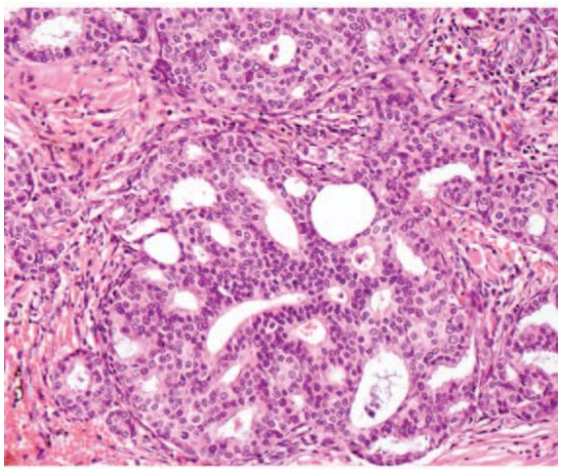
This rare tumor most commonly occurs in the head and neck area, especially on the face, rarely in the extremities [9].

Clinically, the patient may present; for a relatively variable period; a firm subcutaneous nodule or erythematous lesion, discomfort, pain, ulceration, bleeding on contact with the lesion or remaining asymptomatic in most cases [6]. The evolution starts with an aggressive regional extension then metastatic lymph node then at distant sites [10].

The diagnosis of hidradenocarcinoma is very difficult, because it has a wide histological spectrum, it must be differentiated from skin tumors such as malignant melanoma, basal cell carcinomas and squamous cell carcinomas, from other adnexal tumors and also from several benign tumors, hence the need for an immunohistochemical analysis [11].

Based on histological and immunohistochemical analysis, hidradenocarcinoma has two different cell types: dark spindle cells with eosinophilic cytoplasm and larger clear cells with atypical mitotic figures and nuclear pleomorphism. Other histological criteria can be observed such as vascular and lymphatic invasion, perineural invasion, deep extension or necrosis [6].

As in other eccrine tumors, hidradenocarcinoma cells express cytokeratins, EMA (epithelial membrane antigen), CEA (carcinoembryonic antigen) and S100 protein and a strong positivity for Ki-67 and p53 [10]. The hidradenocarcinomas can be; variably; positive for androgen receptors (AR), estrogen receptors (ER), progesterone receptors (PR), EGFR (epidermal growth factor receptor) and HER-2 (epidermal growth factor receptor 2 human) in 36%, 27%, 16%, 85%, 12% of cases respectively. Mutations in PIK3CA, AKT-1 and TP53 were detected in 23% of the cases reported in the literature [6].



Cribriform architecture, polygonal neoplastic cells and eosinophilic cytoplasm are characteristic features of apocrine carcinoma [12].

As being an aggressive and extremely rare tumor, there is no consensus regarding its management. The treatment of choice is surgery by wide local excision with negative margins and selective lymph node dissection, that its role is still under debate according to the very limited data in the literature; hence the interest of early diagnosis [6].

The aggressiveness of this tumor requires the use of adjuvant treatment for better local control [5]**.** Radiotherapy is used selectively in patients who present certain histopathological criteria of high risk of recurrence such as positive margins, lymph node involvement, perinervous invasion, vascular invasion, deep infiltration or histology poorly differentiated [13, 14]. The dose used is between 66 to 70 and 50 Gy to the tumor bed and to regional lymphatic chains respectively by combining photons and electrons. The possible radiotherapy complications and sequelae are less frequent and better tolerated with the advent of new radiotherapy techniques [15].

Adjuvant chemotherapy is not widely used as its effectiveness has not yet been proven either alone or in combination with radiotherapy due to very few case reports. In a metastatic situation, various chemotherapy regimens have been reported: first-line capecitabine, second-line doxorubicin and platinum, cyclophosphamide, vincristine and bleomycin, but its role in sweat gland carcinoma remains unclear [6]**.**

Targeted therapy has proven useful in some reported cases in metastatic conditions; trastuzumab plays a role in disease stabilization according to HER-2 expression levels, and tamoxifen in metastatic hidradenocarcinomas with positive estrogen receptors. The role of EGFR inhibitors, Akt / PI3K / mTOR pathway inhibitors and anti-androgen therapy remains unclear; these therapies are currently in clinical trials [6, 12]**.**

Another recent, well-tolerated therapeutic option suitable in particular for skin and subcutaneous areas is electrochemotherapy (ECT) which involves a combination of locoregional or intravenous administration of very low doses of chemotherapy (bleomycin or cisplatin) with cell membranes electroporation, which improves the penetration of these agents into the cytoplasm of the cancer cell with fewer side effects [16].

The 5-year disease-free survival rate reported in the literature is less than 30% [5], and the local recurrence rate after surgery can reach 50% [6], therefrom the tumor poor prognosis. Early diagnosis is therefore essential for a better therapeutic result and a better patient's quality of life.

CONCLUSION

Hidradenocarcinoma is a rare aggressive tumor with high rates of local recurrence and distant metastasis. The management of this rare entity is not yet well defined. The results are still poor despite the use of multiple treatment modalities.

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