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Hidradenocarcinoma on hidradenoma: rare diagnosis and even rarer degeneration

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ABSTRACT

Background: Hidradenocarcinoma is an extremely rare malignant adnexal tumor and rarely occurs on pre-existing hidradenoma.

Method: It is a rare case for 54 years female.

Results: The confirmation is histopathological and is done on specific architectural and cytological criteria.

Conclusion: Adjuvant radiotherapy is mandatory in case of local recurrence factors.

Keywords: hidradenocarcinoma, hidradenoma

INTRODUCTION

Cutaneous hidradenocarcinoma is a rare malignant tumor developed on eccrine sweat glands. It represents 6% of the eccrine malignant tumors [1]. It classically occurs de novo and rarely on preexisting benign hidradenoma [2]. We report the case of a patient with a hidradenocarcinoma of the scalp developed on a preexisting hidradenoma to highlight the existence of this malignant transformation.

CASE REPORT

A female-aged 54 years, with no medical history, had a small cystic nodule on her scalp for 17 years. An excisional biopsy was performed in favor of a macrophagic folliculitis. Reccurence occured two years later. In view of the growing size of the mass, the patient consulted our department for treatment. The dermatological examination found a nodular, polylobed, firm, erythematopoietic lesion, and 5 cm in size (Figure 1). A biopsy revealed a benign cutaneous adnexal tumor: hidradenoma. A CT scan showed noendocranial extension. Two months later, an excision of the mass was performed. The histological study showed an intradermal tumor proliferation, not connected to the epidermis, largely necrotic, with a lobular architecture in the surface and forming solid masses and infiltrating trabeculae in depth (Figure 2a). The tumor cells were moderately pleomorphic, with mitotic nuclei, mostly vesicular and nucleolated. The cytoplasm was either clear or eosinophilic. (Figure 2b) The periosteum in depth was infiltrated, in contrast to the bone, which was not infiltrated. This histological aspect was compatible with a hidradenocarcinoma resulting from a pre-existing hidradenoma.

The extension work-up was normal. The defect has been grafted secondarly, (Figure 3a and 3b) and additional radiotherapy has been performed after healing. The current follow-up is eight months (Figure 4) without local recurrence or distant extension on CT.

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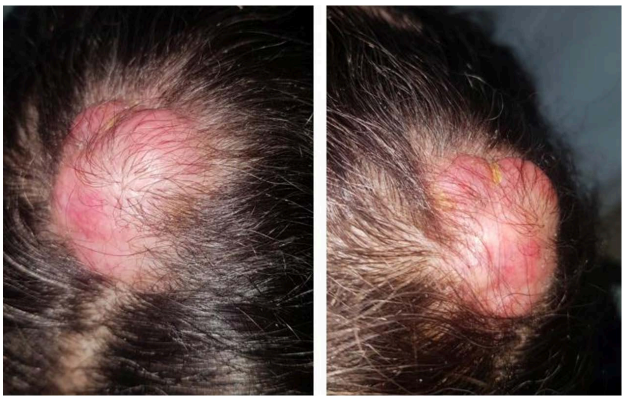

Fig. 1. Erythematous tumor of approximately 5 cm located in scalp

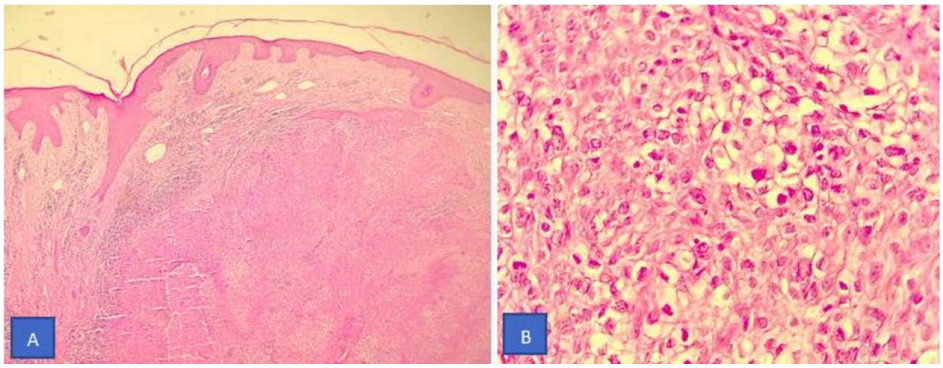

Fig. 2(a). Histological section showing an intradermal proliferation of skin tissue, centered by tumor necrosis (HE x 40). **2 (b).** Made of moderately pleomorphic tumor cells (HE x 400)

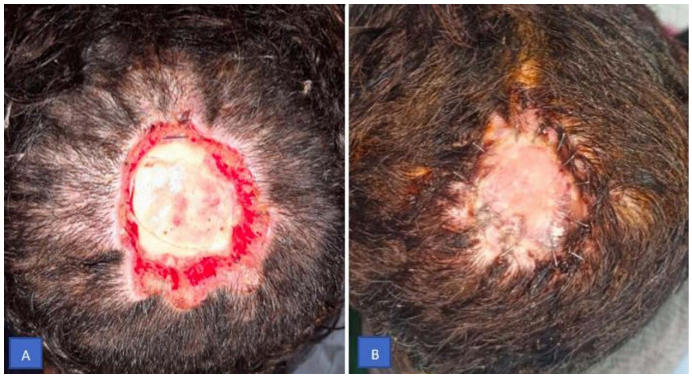

Fig. 3 (a). Excision of the tumor in a sub-periosteal plane with bone cutting **3 (b).** Skin graft on the defect

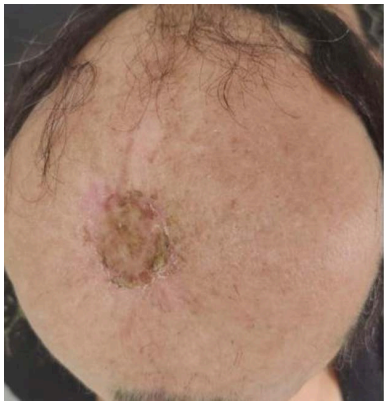

Fig. 4. One month after the end of radiotherapy showing the absence of local recurrence

DISCUSSION

Hidradenocarcinoma was first described by Keasby and Hadley in 1954 [3]. It represents less than 0.001% of all tumors. To date, only about 50 cases have been described in the literature [4]. It most often occurs de novo, rarely on degeneration of a pre-existing hidradenoma (only about 7% of cases) [4]. The epidemiological characteristics are not specific; They are the same as a non-degenerated hidradenocarcinoma. It affects essentially patients between 50 years and 70 years old with female predominance [5]. Classically, the head and neck are the most affected sites; and presents as a single, painless, firm or cystic nodule that can rapidly grow to about ten centimeters in size within a few weeks after incomplete excision or trauma [6].

The histopathology is essentiel. It confirms the eccrine and malignant nature of the lesion, and rules out other differential diagnoses, mainlyrcinomas, melanomas and skin metastases [6]. It shows a sheet-like intradermal tumor proliferation, with sweat differentiation, intersected by a dense and hyalinized stroma with nuclear pleomorphism and mitosis figures [7]. The form developed on pre-existing hidradenoma is characterized by a histological continuum between the clearly benign hidradenoma and the hidradenocarcinoma [7].

Treatment is based on wide carcinological surgery with safe margins. The French ENT Society recommends a lateral margin of 10 to 20 mm with deep margins to the hypodermis unless it is invaded [8]. Some authors recommend a 3 cm margin if it is possible [9]. For Lymphadenectomy, the french ENT society do not recommend it in the absence of palpable lymph nodes [6]. However, based upon the high incidence of distant metastases, other authors recommend regional lymphadenectomy [10, 11]. As there is no proven clear evidence of usefulness of the selective neck dissection, its role is still under debate [12, 13]. Because of the high risk of local recurrence, postoperative radiotherapy seems to be the appropriate therapy [1]. The efficiency of adjuvant chemotherapy has not been demonstrated either alone or in combination with radiotherapy [14]. Targeted therapy with Trastuzumab is used as an effective targeted therapy for treating various solid cancers. In the management of metastatic hidradenocarcinoma, he also plays a role by stabilising the disease [15].

The prognosis is poor with a high risk of locoregional recurrence and distant metastases, essentially lymph nodes [4].

CONCLUSION

Hidradenocarcinoma on hidradenoma is an extremely rare and very aggressive tumor characterized by the frequency of locoregional recurrence and distant metastases. Wide excision surgery is themainstay of treatment. All hidradenomas should be resected with histopathologic study to prevent this rare degeneration, in view of the poor prognosis.

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