

## Malignant Adnexal Carcinoma of the Face: A Case Report

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### Abstract

### Case Report

Malignant adnexal tumors (MAT) form a group of rare, typically low-grade- malignancy carcinomas with follicular, sebaceous, apocrine, or eccrine differentiation or a combination of the first 3 subtypes, the biopsy is required to establish the differentiation subtype and the definitive diagnosis. our case is a 67 years old female presented to our department of maxillofacial surgery of Ibn Sina hospital in Rabat with a left frontal skin lesion that had begun evolving 6 months ago. An excisional biopsy was performed afterwards using an elliptical excision with 1 cm of margins laterally and including the periost deeply, and then the skin defect was sutured in 3 layers. Malignant pilosebaceous tumors are more rare, especially those of the hair follicle. Sebaceous gland carcinoma commonly arises in the periorbital area. Surgical resection is the most commonly implemented treatment modality in MATs.

**Keywords:** Malignant adnexal tumors (MAT), biopsy, Adnexal carcinomas, Malignant pilosebaceous tumors.

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## I- INTRODUCTION

Adnexal carcinomas of the skin are rare and they derive from structures such as sweat glands, sebaceous glands, and hair follicles. Adnexal tumors represent 1–2% of skin cancers. Adnexal carcinomas of the skin derive from structures that have a common origin such as the apocrine and eccrine sweat glands, sebaceous glands, and hair follicles. Malignant adnexal tumors are frequently located in the head and neck region but may appear on the fingers and toes, the trunk as well as the extremities [1, 2]. Their clinical presentation is usually unremarkable, and biopsy is required to establish the differentiation subtype and the definitive diagnosis. Due to their rarity, no clear consensus has been reached on which treatment is most effective. Mohs micrographic surgery is considered to be the best option to prevent recurrence in the majority of patients. Radiotherapy and chemotherapy have been studied in very few cases and have rarely been shown to be effective.

## II- CASE REPORT

A 67-year-old female presented to our department of maxillofacial surgery of Ibn Sina hospital in Rabat with a left frontal skin lesion that had begun evolving 6 months ago. There was a long history of

recreational sun exposure. Her brother had a history of melanoma of his leg in his 50s.

The physical exam found a lesion on her left frontal region of the face measuring 3, 5 × 3, 5 cm in diameter. It was pigmented and was covered with black crust and well-differentiated, mobile relative to the deep plane (Fig. 1, 2). Dermatoscopically the lesion exhibited a dense black with fine linear branched blood vessels centrally.

Firstly a diagnostic biopsy was done showing a malignant adnexal carcinoma. An excisional biopsy was performed afterwards using an elliptical excision with 1 cm of margins laterally and including the periost deeply, then the skin defect was sutured in 3 layers. The specimen was submitted for assessment by a specialist anatomopathologist. Histological exam of the excisional biopsy confirmed the diagnosis of MAT, showing islands of neoplastic cells invading the full thickness of the dermis, several neoplastic aggregates showing a tendency to converge with sebaceous differentiation in the form of sebocytes at different stages of maturation. The excision margins were satisfying as well as the functional and esthetic outcome (Fig. 3, 4).



**Fig.1:** Image showing the macroscopic aspect of the left frontal skin lesion



**Fig.2:** Image showing the macroscopic aspect of the lesion



**Fig.3:** Image showing the surgical outcome after 15 days



**Fig.4: Image showing the surgical outcome after 15 days**

### III- DISCUSSION

Malignant adnexal tumors (MAT) form a group of rare, typically low-grade- malignancy carcinomas with follicular, sebaceous, apocrine, or eccrine differentiation or a combination of the first 3 subtypes. Most tumors of cutaneous appendageal structures are uncommon, and as the clinical appearances are subtle, the majority are generally diagnosed only after excision and pathological analysis [3]. Diagnostic biopsy is therefore not recommended, as it would rarely achieve precise preoperative diagnosis. Clinical diagnosis of most adnexal skin carcinomas is difficult not only between them but also with adnexal benign tumors.

Factors suggesting malignancy include poor demarcation, tumor necrosis, ulcers, border, tumor infiltration, etc. Most benign skin adnexal tumors differentiate along a single adnexal line, resulting in distinct tumor subtypes whose structure, cytochemistry, and immunohistochemistry resemble those of the corresponding appendage [4, 5], Given the common origin of all cutaneous appendages, however, tumor subtypes may share many features not only among themselves but also with other epidermal tumor types. It is not unusual to find evidence of differentiation along 2 or more adnexal lines in synchronous neoplasms or even within the same neoplasm, especially in the malignant appendageal tumors. In such cases, the tumor is described as cutaneous adnexal carcinoma with divergent differentiation [6].

Malignant eccrine tumors usually occur in the fifth to ninth decade of life [7]. They mostly occur as long-standing lesions of the head and neck and, in particular, of the scalp [7]. Some arise in the extremities and infrequently in the trunk. Apocrine lesions are much less common and are found mainly in body folds (including the axillary groin and anogenital regions), as

well as the umbilicus and eyelid. Mixed eccrine/apocrine malignant tumors are also reported [5]. Of the malignant eccrine tumors, microcystic adnexal carcinoma (MAC) has been the focus of the literature because of its aggressive nature despite its bland and asymptomatic presentation and specific difficulties in histopathologic diagnosis. Microcystic adnexal carcinoma occurs predominantly in the midface (nasolabial area) and in middle-aged to older individuals of white origin [8]. They do, however, infrequently develop in other areas, and there have been reports of MAC occurring in Afro-Caribbean [10, 11] and Japanese patients [11]. Malignant pilosebaceous tumors are more rare, especially those of the hair follicle. Sebaceous gland carcinoma commonly arises in the periorbital area [12], they are typically found in women, more often in the seventh decade, and usually are in the upper lid margin. Pilosebaceous adnexal skin carcinomas have the potential for local recurrence and distal metastasis, with mortality from the sebaceous gland carcinoma rising to 9% [13].

Surgical resection is the most commonly implemented treatment modality in MATs [14]. In particular, wide resection is associated with decreased recurrence rates [15]. In a review, Nazemi *et al.*, [16], reported that wide resection was the most preferred method in the treatment of eccrine porocarcinomas; however, the Mohs micrographic surgery could yield more favorable results. In the present case, we performed surgical resection with tumor continuation in the surgical margins or proximity to the adjacent tissues. It is well known that surgical margin is of utmost importance in all skin cancers, as in malignant melanomas. However, there is no consensus regarding the exact surgical margins for each tumor size. In our case, the surgical margin was 1 cm. A wide surgical resection may be helpful to prevent local recurrence.

Frequently, the first excision corrects the diagnosis of the biopsy, without being sufficient to treat the tumor. In other words, the first excision is more often diagnostic than carcinological. These tumors being often insidious, the margins macroscopic ones are not reliable. The excision margins are often incomplete requiring iterative excisions. In the Cuminet series, 57% of excisions were incomplete [17]. Seventy-five percent of excisions were incomplete laterally with a margin of 0.5 cm, 22% when the margin was 1 cm and 25% with a margin of 1.5 cm. Thus, 1 cm of margin with the deepening at the first healthy barrier seems to be an appreciable compromise between the risk of revision and unnecessary mutilation at the facial level. In our case the margins were sufficient with satisfying 1 month and 3 months follow-up.

In all, the treatment of adnexal carcinomas is above all surgical with appropriate resection margins. Radiotherapy would only have its place in adjuvant treatment of certain forms or would be reserved for inoperable patient. As for chemotherapy, it could best slow down the progression in cases of sweat gland carcinomas or sebaceous metastases [18, 19]. Therefore, the treatment strategy should be adapted to each histological type of MAT.

#### IV- CONCLUSION

Adnexal skin carcinomas are both very rare and infrequent compared to other non-melanoma tumors (basal cells and squamous carcinomas). The head and the neck region is the favourite site of presentation. The treatment of election is a complete surgical resection of the tumor. When regional lymphatic metastatic nodes are present, radical neck dissection is the treatment of election. It may be suggested that radiotherapy could significantly influence patients outcomes, and should be considered. When the treatment is oncologically sufficient, the result is a safe and long survival.

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