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RESEARCH ARTICLE

MICROCYSTIC ADNEXAL CARCINOMA OF THE SCALP WITH NODAL METASTASIS TREATED WITH SURGERY AND RADIOTHERAPY: CASE REPORT

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Abstract

Microcystic adnexal carcinoma is an uncommon type of cutaneous neoplasm known by its slow growth but local aggressivity and no potential of systemic metastasis. The standard diagnostic approach is histopathological study of a deep incisional or excisional biopsy. The literature has detailed a variety of treatment modalities, including the Mohs micrographic surgery, standard excision, radiation therapy, chemotherapy, or observation. Mohs is nonetheless the recommended approach nowadays. We describe a unique case of a 69-year-old woman presenting recurrent Microcystic adnexal carcinoma of the scalp with histologically proven spinal lymph node involvement, treated with surgery, lymph node dissection, and adjuvant radiotherapy. the ongoing assessment shows no evidence of clinical or radiological recurrence at 28 months post-treatment.

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Introduction:

Microcystic adnexal carcinoma (MAC) is an uncommon, slow-growing, malignant cutaneous tumor that develops from sweat glands of the head and neck region. it displays an aggressive local behavior extending beyond clinical margins into the dermis and perineural tissues. To date, there are only few reports of histologically proven lymph node involvement with MAC.

Surgical management is the standard care of MAC including Mohs micrographic surgery or complete excision with deep margin assesment (1, 4)

Adjuvant radiation therapy can be an option for high-risk MAC with deep infiltration, unclear margins or recurrent microcystic adnexal carcinoma. (3, 21).

Case presentation:

A 69-year-old woman had a 12-year history of a histologically proven Microcystic Adnexal Carcinoma of the right parietal scalp for which a wide excision was performed. Subsequently the patient underwent a skin graft reconstruction with no adjuvant therapy.

7 years later, an asymptomatic, indurated scalp plaque appeared of the left parietal region gradually growing larger that was surgically managed with large excision and a deep tissues assesment.

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Histopathology study revealed a microcystic adnexal tumor infiltrating massively deep hypodermis with no involvement of the epidermis. However, the quality of excision was not assessed given the fragmented and non-oriented nature of the surgical sample. No adjuvant therapy was indicated.

A plastic surgeon was consulted 2 years later for guidance and treatment of a new lesion on the last excision scar. The initial examination revealed a 3cm exophytic firm scalp plaque of the left parietal excision scar with indurated base and no surface ulceration. An ultrasound of the cervical region revealed a 2.5 cm right spinal adenopathy that was quite suspicious during the examination of the cervical lymph nodes.

To aid in preoperative planning, computed tomography (CT) scan was performed showing a left parietal, well-defined, heterogeneous subcutaneous lesion measuring 27/17mm with foci of bone lysis and erosion of the underlying bone, without any endocranial extension. On the post-contrast study, the lesion displayed heterogeneous enhancement. The right spinal Adenopathy was detected and had criteria of malignancy with a necrotic center and a thickened cortex ; There was no imaging evidence of distant metastasis. **(Figure 1)**

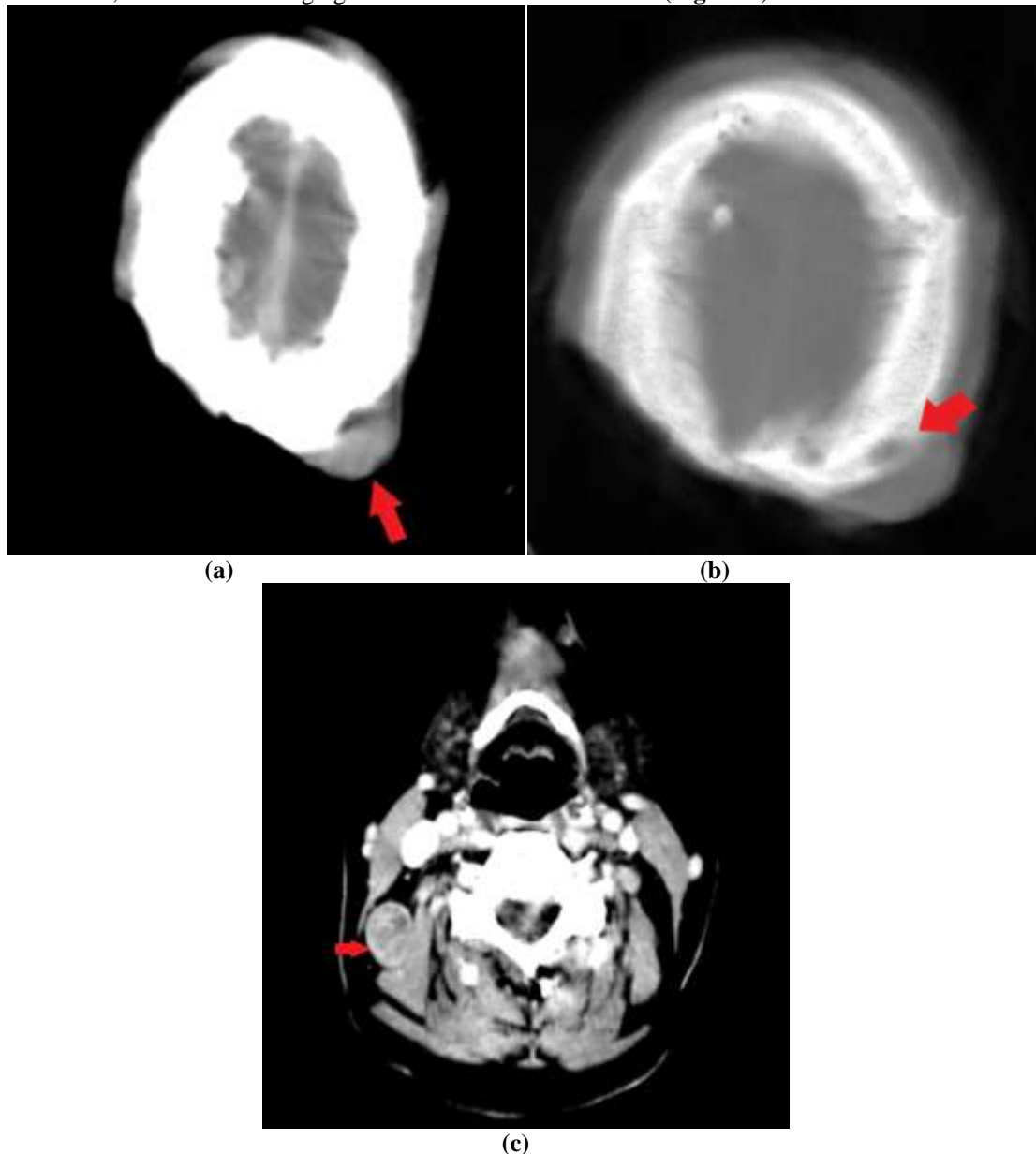


Figure 1:- Computed Tomography (CT) scan showing microcystic adnexal carcinoma lesion of the left parietal scalp (a) with underlying bone lysis (b) and right spinal adenopathy (c)

Histopathology analysis of the biopsy revealed carcinoma cells organised in dermal ductal structures with cords, squamous nests with horn cysts, and basaloid cells with surrounding sclerotic stroma and perineural involvement.

Immunohistochemical study showed focal expression of Bcl2, AML in tumor cells, with negative BERRP4 and anti-PS100 which confirmed the MAC diagnosis. The Ki67 was focally high.

The case was discussed with the tumor board who recommended surgical management and adjuvant radiotherapy.

The patient had a wide tumor excision with 1cm macroscopic safety margins, exposing the infiltrated bone, which was scraped off. Skin closure did not require skin reconstruction. Additionally, bilateral lymphadenectomy of level I, II, III, IV, and V was performed.

The histopathological study of surgical specimen revealed the presence of microcystic adnexal carcinoma with undifferentiated foci displaying follicular and ductal structures in a sclerotic stroma, measuring 2 cm as greater dimension. Resection margins were clear except for the deep bone border which was still infiltrated, with perineural involvement.

The right spinal adenopathy had been proven to be invaded by MAC cells, without capsular effraction or vascular emboli.

After surgery, a CT body scan revealed no signs of gross disease or distant metastases.

Adjuvant radiotherapy was initiated 8 weeks after surgery.

The patient received intensity modulated external beam radiotherapy with 6MV photons and a 0,5 cm bolus on the scalp. A total dose of 60Gy in 30 fractions was delivered to the high risk volume including the tumor bed and right lymph nodes levels II, III, IVa and the spinal level (Figure 2). A prophylactic dose of 54Gy was delivered to the low risk volume with a 1,5cm margin to the tumor bed and contralateral lymph nodes levels.

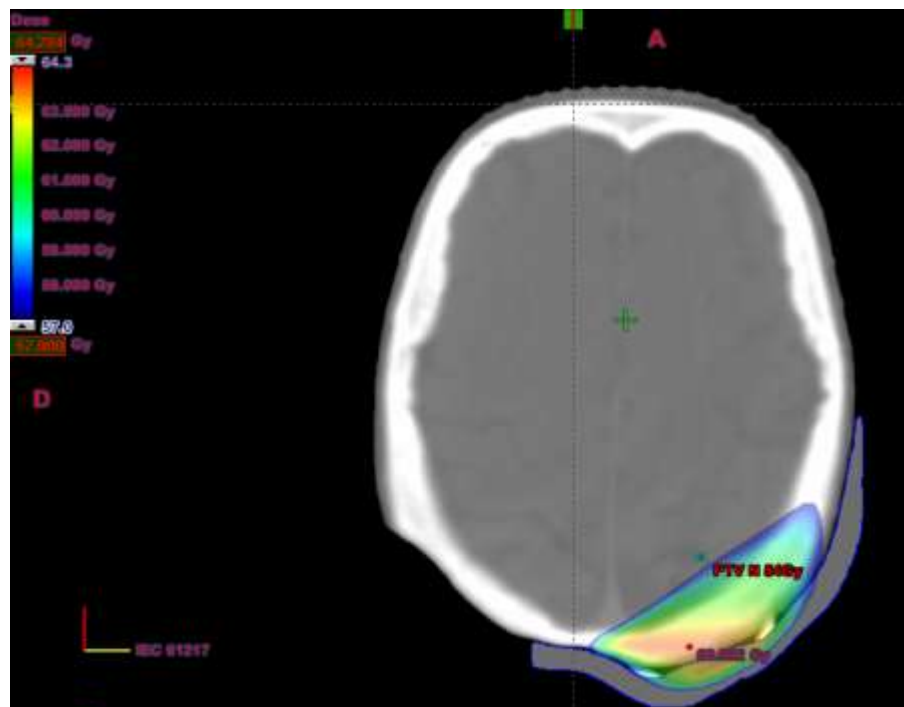


Figure 2:- Radiation therapy dose distribution of 60Gy delivered to the tumor bed with a 0,5 cm bolus

The patient experienced grade 2 dysphagia throughout therapy, which was treated symptomatically, and grade 2 radiodermatitis on the tumor bed with moderate desquamation. (Figure 3)



Figure 3:- Radiation dermatitis during the course of radiation treatment.

After 28 months of follow-up, the patient developed radiation-induced alopecia with skin atrophy in the irradiated area (figure 4) but there was no evident clinical and radiological sign of recurrence up until the time of preparing this report.



Figure 4;- clinical examination of the tumor bed 1-year one year after radiotherapy's completion.

Discussion:-

Microcystic adnexal carcinoma (MAC) is a rare form of cutaneous tumors. It is clinically presented as a slow growing, smooth-surfaced, flesh-colored asymptomatic lesion with a predilection for the head and neck, including the scalp (10-17). It can also appear in the axilla, chest, back, peri-anal region or lumbs (6). As a sign of perineural dissemination, some patients report paresthesia or burning sensation at the tumor site.

MAC is characterized by its local aggressivity and perineural involvement that increases the risk of local recurrence which has been reported in several cases, reported to occur 6 months–15 years after standard surgical removal (12). lymph nodes metastases are not common (5,6) but are described in the literature, including a case of Bier-Laning et al (1) reporting MAC on the right side of the face with histological evidence of contralateral cervical lymph node metastasis. The case we described provides significant evidence that MAC can metastasize to lymph nodes.

A deep biopsy specimen including subcutaneous fat helps achieving an accurate diagnosis (3). Superficial biopsies often lead to a misdiagnosis since MAC does not show any superficial cells atypia and frequently invades the dermis, fat, muscle, and underlying bone. (3,7,8). The tumor's histological features include epithelial nests, strands, and cords; small cysts surrounded by 3 to 5 concentric cell layers or located in tumor clumps and centered by laminated keratin (13); ductal structures and a sclerotic stroma (8). Perineural involvement is commonly observed (9).

Although no antibody is sensitive and specific enough to distinguish microcystic adnexal carcinoma from other cutaneous carcinomas, the combination of immunohistochemistry with histological, clinical and radiological criteria can lead to a definitive diagnosis (23). MAC does not normally express BEREPA, an epithelial marker; it binds PHLDA1, with variable expression of CK15, and CK19, P63. (14, 23)

Contrary to other skin malignancies, there has been no consensus on the diagnostic and care management of MAC, until 2019 when a Consensus clinical practice guidelines was reached by American Medical Association through collaborative review of the literature and evidence tables (3). Preoperative imaging is recommended to evaluate local tumor extension, and includes computed tomography with bone window if bone involvement is suspected. Gadolinium enhanced magnetic resonance imaging is indicated when extensive perineural invasion (PNI) is suspected (2, 3). Given the rarity of distant metastases, it is not necessary to perform a whole-body scan, but any clinically detected lymphadenopathy should also be explored. (3,8)

The main objective of treatment for microcystic adnexal carcinoma is complete excision with clear surgical margins, while maintaining function and cosmesis. (3,6) The best treatment for MAC at the moment is Mohs micrographic surgery with a great tumor local control rate documented through many case reports (4, 5, 10, 11, 15, 16-19). A simple excision has a much higher failure rate recurring in 6 months–30 years (19), which may explain the local failure experienced by our patient.

Sentinel lymph node biopsy is not recommended for the staging of MAC since nodal metastasis rate is approximately 2% based on currently available data (17-18-19-22). If node involvement is histologically proven, the adenopathy must be surgically removed, without completion lymph node dissection. (3, 19)

Radiotherapy is not usually a first line treatment but may be considered for patients for whom surgery may be disfiguring with poor functional and cosmetic results (3, 20, 21, 22). Adjuvant radiotherapy (60-66 Gy, 2 Gy per fraction) is considered for MAC to improve local control, when there is perineural involvement, when margins cannot be cleared surgically or in case of recurrent disease. A margin of 3 to 5 cm, if possible, around the surgical bed is suggested (20,22). There is no evidence to support use of prophylactic nodal radiotherapy (21, 22)

In this case report, we describe an uncommon case of recurrent MAC of the scalp with node involvement and bone lysis who underwent surgery, lymph node dissection and adjuvant radiation therapy as an effective therapeutic procedure an excellent clinical local control after a 2-year follow-up, and acceptable side effects.

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