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Aggressive Microcystic adnexal carcinoma (MAC)

Jagannath K.P[1], Ajay G.V[1], Thejaswini B[1], Lokesh .V[1], Asha latha.D[1]

[1]Department of Radiation Oncology, Kidwai Memorial Institute of Oncology, Bangalore

Abstract

Microcystic adnexal carcinoma (MAC) is a malignant appendageal tumor first described in1982. It can be clinically and histologically confused with other malignant and benign cutaneous neoplasms,leading to inadequate initial treatment. This neoplasm is locally aggressive and deeply infiltrating,characterized by high morbidity and frequent recurrence.

Key Words: Microcystic adnexal carcinoma, Bone metastasis, Electron beam therapy

INTRODUCTION

Microcystic adnexal carcinoma (MAC) is a rare, malignant appendage tumor commonly classified as a low-grade sweat gland carcinoma that typically occurs on the head and neck, particularly the central face it can be clinically asymptomatic with a benign appearance. This tumor was first described as a specific entity by Goldstein et al1 in 1982.[1] MAC can appear clinically benign and has been associated with much histological confusion.

Microcystic adnexal carcinoma is a rare tumor, with only slightly more than 300 cases reported worldwide.[2] Thomas et al[3]reported a mean number of 1.63 cases per year in a rural northeastern area of the United States. Most cases (90%) of microcystic adnexal carcinoma occur in whites, but it may occur in persons of other races such as Latinos and Asians.[4,5] Fifty-two cases have been reported in Japan.[6,7] In African Americans, 9 cases in total have been reported in 8 patients.[2,8-13] The overall clinical behavior of microcystic adnexal carcinoma appears to be similar in all races. No significant sexual predilection is reported, although some studies suggest a slight female predominance.[14-17]

This neoplasm is locally aggressive and deeply infiltrating, characterized by high morbidity and frequent recurrence despite aggressive treatment with surgery, radiation therapy, or both. Despite subsequent widespread recognition of MAC as a discrete clinicopathologic entity, its precise relationship to and histologic discrimination from other putative locally aggressive sweat gland carcinomas (reported under a variety of names, including sclerosing sweat duct carcinoma, syringoid eccrine carcinoma, syringomatous carcinoma, and eccrine epithelioma) remains unresolved and has provoked considerable nosologic and diagnostic confusion. Cooper et al described MAC as synonymous with group 1 sclerosing sweat duct carcinomas.[18]

CASE PRESENTATION

A50 year old female patient presented with complaints of swelling in scalp since 1year which was associated with recent increase in size for which a simple excision was done. 3months later there was recurrence of the swelling for which she was referred to radiotherapy. Her medical history was remarkable but recently she has developed pain all over the body. Local examination of the scalp revealed two hyper pigmented nodules that was firm on palpation and measuring around 1cms in its greatest diameter.

A repeat biopsy was performed.Biopsy shows sections shows tumor cells arranged in a microcystic pattern in subcutaneous plane with microcysts are line by single cells with bland nuclei, few showing hyperchromasia , without pleomorphism against a sclerotic back ground. underlying dermis is free (Picture 1).Resected margin in the section shows tumor. Bone scan revealed multiple skeletal metastases.

Patient was treated with palliative Radiation therapy with Electron beam of 30Gy electron in 10 fractions to the scalp and P-32 therapy and zoledronic acid for bone metastasis

This female patient had MAC of the of the scalp. The neoplasm in this case was located in the prototypical, centrofacial region, and patient has multiple bone metastases. Even though MAC appears to be benign and show a benign course, some patients MAC can have an aggressive behavior. Hence patients with MAC need to be treated aggressively with wide excision / resection and local Radiation therapy treatment planned in close margin status upfront to ensure good locoregional control. Elective chemotherapy may be considered

DISCUSSIONS

MAC is an aggressive carcinoma of the centrofacial region, it occurs most frequently on the upper lip. Other regions of the face including the lower lip, chin, nose, nasolabial folds, cheek, and eyelids are less common locations.(1,19) MAC has been reported to occur in areas other than the face, specifically the scalp and axilla (14,19).

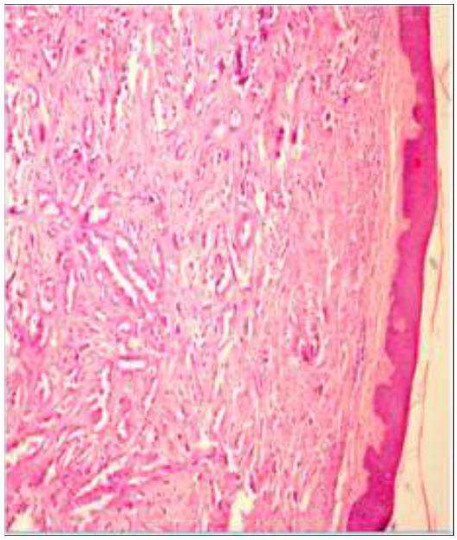
The differentiation pathway of microcystic adnexal carcinoma (MAC) has provoked considerable debate. Goldstein et al[1]initially suggested that it showed dual pilar (the superficially located keratocysts resembling follicular infundibula) and eccrine differentiation. Others have supported this notion, noting, in particular, that the keratocystsoften expressed pilar-type keratins and that occasionally, trichohyalin granules are present. However, some authors have suggested that microcystic adnexal carcinoma shows only eccrine

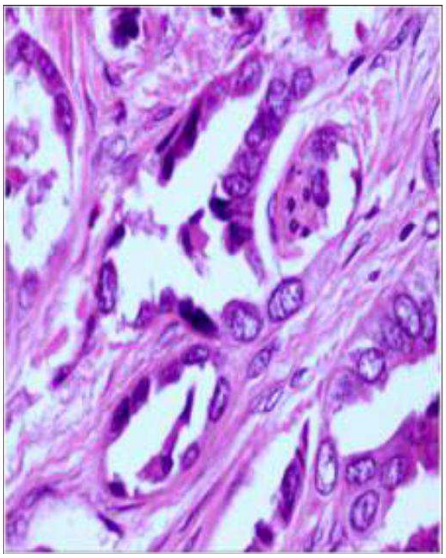
Address for correspondence*

Dr Jagannath K.P

Department of Radiation Oncology, Kidwai Memorial Institute of Oncology, Bangalore

E mail : dr.jagannath8@gmail.com





Picture 1 (a & b): Microscopic feature showing tumor cells arranged in a microcystic pattern in subcutaneous plane with microcysts are line by single cells with bland nuclei, few showing hyperchromasia , without pleomorphism against a sclerotic back ground. underlying dermis is free.

differentiation.

More recently, based on the ontogenetic relationship between hair follicles and apocrine glands, microcystic adnexal carcinomas have been theorized to display folliculo-apocrine or sometimes folliculo-sebaceous-apocrine differentiation rather than folliculo-eccrine differentiation. Support for this premise is provided by occasional cases showing focal apocrine decapitation secretion and the demonstration of sebaceous foci in some tumors. Little information is available on the molecular pathophysiology of microcystic adnexal carcinomas, although one report documented a deletion on arm 6q in one case. A case of microcystic adnexal carcinoma occurring in siblings also suggests a genetic role.[20]

Morbidity is high because of the deeply infiltrating nature of the tumor, which can invade into bone, muscle, blood vessels, cartilage, and nerves. In one study, the mean clinical lesion size was 3 cm2, but the final defect size was 18 cm2, highlighting its occult extension.[3]Exquisitely rare orbital extension can occur, and a primary orbital presentation has also been reported.[21]

Mortality from metastatic disease is rare, with only 6 local,[22-27] and 3 distant metastases reported.[25-27] Local metastatic disease may, in most cases, be occult contiguous extension along neurovasculature Bundles.[23-25] Considering that more than 300 cases have been reported in the medical literature worldwideonly 2 deaths have been attributed to microcystic adnexal carcinoma,[8,23] both due to metastatic disease.

Fernandez-Figueras et al reported a case of a high-grade carcinosarcoma with an architectural pattern similar to microcystic adnexal carcinoma but also exhibiting nuclear pleomorphism, hyperchromasia, and large nucleoli histologically. The patient developed metastatic lung disease from the tumor and died within 6 months.[28] The authors further noted histologic high-grade features in their case that were similar to other cases reported with metastatic disease,[24,27,29] and they suggested this may be an indicator for more aggressive disease.

CONCLUSION

As MAC is a rare case and seen mainly in the head and neck region clinically unrecognizable tumor spread and perineural invasion often encountered with MAC. Aggressive initial treatment by microscopically controlled excision appears to offer the greatest likelihood of cure for this neoplasm, while providing conservation of normal tissue. Radiation is used as adjuvant treatment.

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