

Case Report (III)

Microcystic adnexal Carcinoma in a 10 year old child: A Case Report and Review of Literature

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ABSTRACT:

Microcystic adnexal carcinoma (MAC) is a locally aggressive tumor of skin appendages that presents a confusing problem to surgeon and pathologist due to its unusual behaviour. Diagnosis is usually difficult and late. Most often these neoplasms are mistaken for other benign disorders. Metastatic disease from MAC is exceedingly rare and has been reported in the last 5- 6 years. We report a child with recurrent microcystic adnexal carcinoma of chest-wall with axillary node metastasis. This case is being reported due to its rarity in a child.

INTRODUCTION

Adnexal neoplasm constitutes a wide spectrum with lesions of hair, sebaceous glands, and eccrine glands. These tumors may be benign or malignant. The origin of these tumors is not clear. Hence the descriptive term microcystic adnexal carcinoma (MAC) is accepted. These tumors are very rare and represent less than 1% of all skin tumors. Synonyms for microcystic adnexal carcinoma include sweat gland tumor, sclerosing sweat gland carcinoma, sweat duct carcinoma with syringomatous features, and combined adnexal tumors of the skin.

MAC primarily affects middle-aged individuals with equal male to female distribution. Occasionally these tumors have been reported in pediatric population²⁻⁴.

MAC cause minimal symptoms. They usually occur as a solitary lesion about 1 x 1cm in size,

which are slowly progressive. They may have cystic, solid or nodular consistency⁵. The tumor is most commonly located in head & neck area⁶. They lack distinct clinical and histologic features. Pathogenesis of these tumors is not completely understood. Exposure to ionizing radiation may be an etiologic factor. Radiation may precede MAC by as long as 40 years⁴. Cases with metastatic disease are even rarer. There are no definite guidelines for treatment.

CASE:

A 10-year-old boy presented in 1996 with a painless, slowly progressive nodular swelling over the right chest wall. There were no other constitutional symptoms. Examination revealed a firm 1 x 2 cms mass, which was mobile, non-tender and there was no regional nodal enlargement. Systemic examination did not reveal any abnormality. Blood counts and biochemistry were normal. Chest X-ray and abdomen ultrasound were normal. Excision biopsy was suggestive of a microcystic adnexal carcinoma (sweat gland tumor) of intermediate grade. The patient had local recurrence after one year with pain and enlarged hard axillary node mass (2cms x 2cms). Wide excision of chest wall tumor along with axillary dissection was done. Histopathology confirmed recurrence of MAC, cut margins were free of tumor and one out of six lymph node was positive for metastasis. The patient was followed up and 5 years later in July 2002 had recurrence of 1 x 1 cms nodule over scar site over the chest wall. There was no axillary node enlargement and the systemic examination was normal. Excision of

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the nodule confirmed recurrence of microcystic adnexal carcinoma. Patient was subsequently lost to follow up.

DISCUSSION:

Daniel et al⁷ have reported a case of MAC who had metastatic disease 10 years after initial presentation. They found estrogen and progesterone receptor positivity in the tumor and antiestrogen therapy was useful in this patient. Adjuvant radiotherapy with or without chemotherapy is advocated in recurrent disease even after aggressive surgery. The microcystic adnexal carcinoma (MAC) in this patient originated from the chest-wall. It recurred after one year with axillary metastatic disease and second local recurrence after five years.

MAC is uncommon locally aggressive neoplasm with usually a very low metastatic potential. Though this entity has been recognized for more than three decades, cases are often reported benign on initial biopsy. These tumors may be confused with benign adnexal neoplasms like desmoplastic trichoepithelioma, trichoadenoma, and syringoma. Goldstein et al⁸ described specific characteristics of MAC, such as islands and strands of small basaloid adnexal keratinocytes within the dermis. In addition, horn cyst formation surrounded by keratinocytes with very little cytologic atypia is found. Cells near the periphery are arranged in a palisade fashion. "Perineural invasion is common⁵ and involvement of subcuticular tissue, muscle and bone⁹ may also occur."

The cell of origin appears to be pluripotent adnexal keratinocytes, which is capable of both, follicular and sweat gland differentiation¹⁰. MAC expresses various glycoproteins of CEA (carcino embryonic antigen) family. These can help in diagnosis but cannot differentiate it from metastatic carcinoma⁵.

MAC has some characteristic clinical features. In most published cases MAC arose from the skin of face usually the lip¹⁰. In a Japanese study of 51 patients, 43 had lesions on face while only two patients had lesion over chest-wall¹². Typical symptoms are sensation of fullness, pain, burning, anesthesia, and paresthesia. These symptoms could be related to perineural invasion. MAC is rare in children. Chow et-al² has reported a series of 3 children, youngest of seven months age; one child had local recurrence while two had rapid extensive metastasis. MAC has high propensity for recurrence, sometimes years after initial surgical excision.

The appropriate management of MAC is not yet clearly known, as there are few case reports. A variety of treatment approaches have been used including primary excision, Moh's excision⁹ and radiotherapy. Moh's excision has been reported to be preferred method of surgical excision [11]. The goal of achieving tumor free margins by using frozen section should be attempted. Despite these approaches risk of relapse is high.

It suggests biologic behavior of the tumor rather than treatment failure¹⁰. Recurrent tumors have been observed many years after excision with negative margins. Conversely, cases in which negative margins could not be achieved have been well for years. Non-Moh's surgical excision is associated with recurrence rate of 47 to 55%. Complete surgical excision, even in presence of extensive metastasis is recommended. Initially it was believed that these tumors do not metastasize but now it has been well reported^{4,5,8,12,13}.

In summary, MAC is an uncommon cutaneous neoplasm, which has an indolent course with multiple recurrences. Diagnosis is usually delayed and surgical excision is the treatment of choice even in case of metastatic disease.

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