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Cutaneous Mucinous Cystadenocarcinoma

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Introduction

Primary cutaneous mucinous carcinoma (PCMC) is a rare low-grade malignant neoplasm originally believed to be of eccrine origin(1) These tumors are slow growing, arising on the face (particularly the eyelids), axilla, scalp, and trunk of middle-aged and older individuals(2). The lesions typically present as erythematous, asymptomatic nodules measuring 0.5 to 7 cm in diameter; however, larger variants have been reported(3-5) The contemporary, but still somewhat controversial, view is that these tumors actually demonstrate apocrine-type differentiation(6-8)

Morbidity related to PCMC is primarily associated with incomplete resection(9,10) While PCMC tends to grow slowly and have a good prognosis, late recurrences and rare metastases have been reported. Recurrence portends substantial morbidity, as recurrent tumors do not tend to respond to radiation treatment or chemotherapy(11)

As is the case with many rare nonmelanoma tumors, there is no standard of care for the surgical treatment of PCMC. Currently employed modes of treatment for PCMC vary from standard excision to wide local excision, including dissection of regional lymph nodes(11). Mohs surgery was first introduced as a means for PCMC treatment by Weber et al(11,12) in 1988. Due to the rarity of PCMS, the studies included were observational, primarily case reports and case series

Case Report

A 64 year old male presented to our clinics with chief complains of left axillary swelling for 4 month which was histologically proven to be cutaneous deposits of mucinous cystadenocarcinoma

Patient was examined and found to have no abnormality in oral cavity and no palpable cervical and supraclavicular lymph nodes. In right axilla approximately 3cm*2.5cm firm non tender mobile lymph node was palpable.L axilla was NAD.

Contrast CT scan Face and Neck showed B/L small space deposits at level II & III. Small in durative lesion in R axilla with focal stranding of fat plane no abnormality detected in thoracic cage.

Ultrasound of right upper arm showed round well defined lesion with internal collection and solid component noted in right axilla with slight internal vascularity on colour doppler

Biopsy from axilla showed cutaneous deposits of mucinous cystadenocarcinoma.

Patient was planned for chemotherapy (paclitaxel+cisplatin).First cycle of chemotherapy was given and patient was discharged in stable condition.

Discussion

Primary cutaneous mucinous carcinoma is a rare sweat gland neoplasm of low malignant potential[14]. The first case was described by Mendoza *et al*. in 1971[17] Most of the cases involve eyelids (41%), scalp (17%) face (14%), and trunk[13,14,17] Axilla is relatively an uncommon site of this neoplasm[17]. The tumor occurs at elderly age (average age: 62 years) with a male preponderance[17]. Clinical presentation of primary mucinous carcinoma is a slow growing, painless solitary mass[13,15,18] Sometimes, the surface may show superficial ulceration/crusting[13,15,18]

Metastatic mucinous carcinomas to axillary lymph nodes are more common than primary mucinous eccrine carcinoma at axillary region. Correct diagnosis needs other ancillary investigations to rule out the metastatic mucinous carcinoma. On immunohistochemistry, primary mucinous carcinoma is positive for CK-5, 7, 14, 17, epithelial membrane antigen, carcinoembryonic antigen, ER, and PR but negative for P63, CDX2, and CK20[17,19,20,21] In the cases of metastatic mucinous carcinoma of breast, the tumor cells may express ER and PR but negative for p63[18,19] Metastatic mucinous carcinoma of gastrointestinal tract expresses CK20 and CDX 2, which helps to differentiate from primary cutaneous mucinous carcinoma[20,21]

Primary mucinous eccrine carcinoma is a slow growing low-grade malignancy with a risk of local recurrence in 30%–40% of cases[13,15,22] Distant metastasis is very rare and found only in 3% of the cases[13,15,22] Wide local excision and follow-up is the treatment of choice for this malignancy. Primary cutaneous mucinous carcinoma is chemo- and radio-resistant[19,22]. Prognosis is very good in comparison to secondary mucinous carcinomas. Follow-up of the patients at a regular interval is necessary to detect any recurrence or metastasis[19].

PCMC is an uncommon subtype of sweat gland tumor. Only about 100 cases of PCMC have been reported[23,27] since the first report by Lennox *et al*.[32] in 1952. PCMC shows a slight male predominance and typically affects people aged 50–70 years. The eyelid was most commonly affected, followed by the scalp, face, axilla, chest/abdominal wall, vulva, neck, extremity, canthus, groin, and ear[27]. The primary lesion of PCMC is often solitary, and the size of the neoplasm varies in diameter from about 0.7–8.0 cm.

It is still controversial whether PCMC has eccrine or apocrine differentiation. Determining the eccrine or apocrine differentiation continues to be problematic because there is no clear distinction in histologic and immunohistochemical features. Robson *et al*.[33] suggested that apocrine differentiation was determined when encountering the tumor in its usual anatomic location with numerous apocrine glands and with its propensity for coexistence with follicular tumors, which means it is derived from the follicular-apocrine-sebaceous rather than eccrine axis. The presence of decapitation secretion is also known to be a hallmark of apocrine differentiation. However, some authors[33] described apocrine carcinoma without decapitation secretion, and they relied on the identification of tumor cells with abundant eosinophilic cytoplasm in tumors that arises from a typical location including the axilla or groin

For PCMC, some authors have favored eccrine differentiation based on immunohistochemical studies and ultrastructural analysis[24-26]. On the other hand, other studies have suggested that PCMC has an apocrine origin[28-31]. Riquena *et al*.[28] suggested an apocrine origin for PCMC because it shows identical histologic features with mucinous carcinoma of the mammary glands, known as modified apocrine glands. Another study reported a case of PCMC coexisting with trichofolliculoma, also providing evidence of apocrine lineage[30].

In summary, PCMC is an uncommon variant of sweat gland tumor. Our case is a rare PCMC. Minor poorly differentiated components were also observed, which suggested apocrine differentiation. However, further studies are required to clarify the histogenesis of PCMC. Clinical differentiation from metastatic mucinous carcinoma is most important, and a careful search for an *in situ* component can be a helpful morphologic finding that identifies the primary nature of the carcinoma in addition to the immunohistochemical panel.

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