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Primary cutaneous mucinous carcinoma in a periorbital lesion: two case reports and literature review

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Case Report and Literature Review

Primary cutaneous mucinous carcinoma (PCMC) is a rare malignancy of the sweat glands that most commonly affects the periorbital area. It is characterized by slow growth over a prolonged period, and its morphology can be easily confused with a benign tumor, such as an epidermal cyst. Consequently, many patients experience recurrence after undergoing multiple resections. However, there are few reports concerning the surgical management of PCMC. We present two cases of PCMC originating in the periorbital area. The first case involved a 76-year-old man with a mass measuring 3.0×1.5 cm that had been increasing in size. The second case was a 61-year-old man with two masses, each measuring 1.0×1.0 cm, that were also growing. Both patients underwent wide excision with a 5-mm safety margin, which was determined based on the widest view of the cross-section of the mass on the magnetic resonance imaging. Subsequently, based on the intraoperative frozen biopsy results, both patients underwent additional excision with a 5-mm safety margin in only one direction. This report shows that, when determining the surgical margin of PCMC in periorbital area, employing imaging modalities and intraoperative frozen biopsies can be helpful for narrowing the surgical margin.

Abbreviations: CK, cytokeratin; CT, computed tomography; PCMC, primary cutaneous mucinous carcinoma

Keywords: Case reports / Eyelid neoplasms / Margins of excision / Skin neoplasms

INTRODUCTION

Primary cutaneous mucinous carcinoma (PCMC) is a rare malignancy of the sweat glands that typically arises on the face—particularly in the periorbital area, which accounts for 40% of cases—as well as the axilla, scalp, and trunk of middle-aged and older individuals [1]. It grows slowly over a long period of time, and its morphology can lead physicians to mistake it for a benign soft tissue mass, such as an epidermal cyst [2]. Therefore, many patients experience recurrence despite multiple resections [3,4]. Given the rarity of PCMC and other nonmelanoma tumors, there are few reports on their management. There is still no consensus in the literature regarding treatment protocols, including the appropriate surgical margin. In this context, we present two cases of PCMC with no evidence of recurrence during a prolonged follow-up period.

CASE REPORT

The first patient, a 76-year-old man, presented to the outpatient clinic with a growing mass measuring 3.0×1.5 cm on the right temple area (Fig. 1A). This mass had remained relatively stable in size for over 20 years. Prior to his first visit to our center, he had sought treatment at a local clinic several months earlier to

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have the mass removed. The mass recurred and was larger after the excision. An excisional biopsy was subsequently performed, leading to a histopathological diagnosis of mucinous carcinoma. To evaluate for potential occult primary malignancy and metastasis, a comprehensive whole-body work-up was conducted. This included chest, abdominal, and pelvic computed tomography (CT) scans, all of which were negative for occult malignancy. Additionally, esophagogastroduodenoscopy and colonoscopy results were normal. The biopsy sample exhibited a distinctive histological feature of mucin-producing sweat gland carcinoma, characterized by mucinous pools described as "floating islands of tumor cells" (Fig. 2A). Immunohistochemistry further confirmed this finding with cytokeratin (CK)-7 positive results (Fig. 2B). Through this systematic work-up and biopsy analysis, PCMC was identified. Treatment involved a wide excision with a 5-mm safety margin (Fig. 3) and the creation of a cheek flap. However, due to the involvement of the margin at the 12-o'clock position, an additional excision with a 5-mm safety margin was necessary. The patient did not receive adjuvant chemotherapy or radiation therapy.

The second patient was a 61-year-old man who presented to the outpatient clinic with two enlarging masses, each measuring 1.0×1.0 cm, located on the left upper eyelid and the left lateral canthal area (Fig. 4A). He had previously sought treatment at a local clinic where the mass was initially presumed to be an epidermal cyst. He had undergone mass excision twice at the local clinic, once 5 years ago and again 2 years prior, however, the tumor had recurred each time. Following the same diagnostic process as the first patient, he was diagnosed with PCMC. Histological examination revealed neoplastic cells arranged in nests, strands, or as individual units, suspended in lakes of extracellular mucin, and immunohistochemistry showed CK-7 positivity. Due to frozen biopsy results indicating margin involvement at the 6-o'clock position, a wide excision

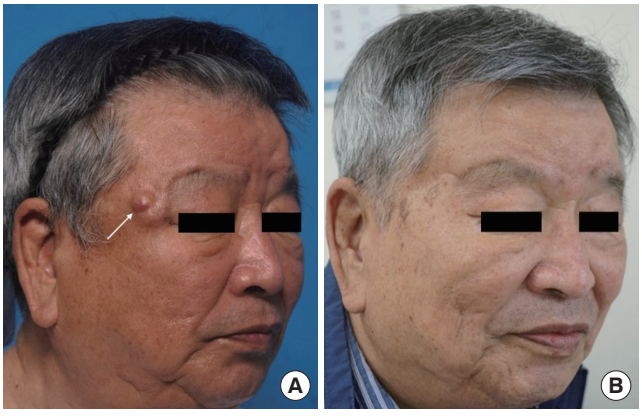

Fig. 1. Preoperative and postoperative photographs of a 76-year-old man with a primary cutaneous mucinous carcinoma. (A) A preoperative photograph showing a 3.0×1.5 cm mass in right temple area (white arrow). (B) A postoperative photograph taken after 2 years. A rotation flap was performed for periorbital reconstruction after wide excision.

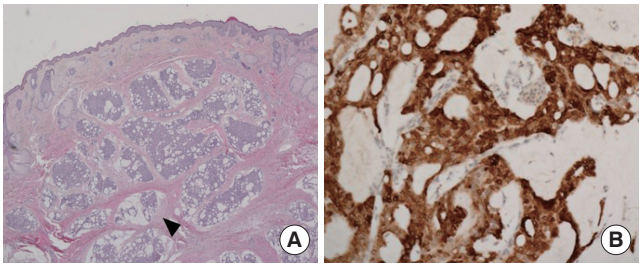

Fig. 2. Histologic and immunohistochemical analyses of the second surgical specimen from a 76-year-old man with primary cutaneous mucinous carcinoma. (A) Histologic examination showing clustered tumor cells with features of a mucin-producing sweat gland carcinoma and large mucinous pools referred to as "floating islands of tumor cells" (black arrowhead) (hematoxylin and eosin, ×20). (B) Immunohistochemistry showing positivity for cytokeratin 7 (×100).

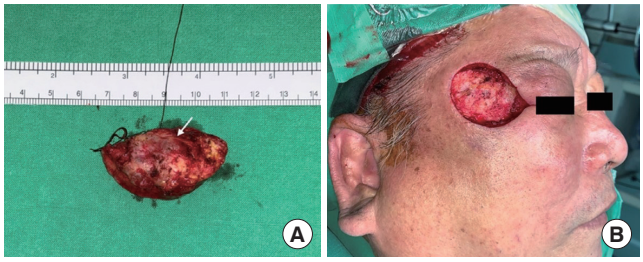

Fig. 3. Clinical photographs of a 76-year-old man with a primary cutaneous mucinous carcinoma. (A) Gross photograph showing the translucent morphology of the mass (white arrow). (B) Intraoperative photograph right after wide excision.

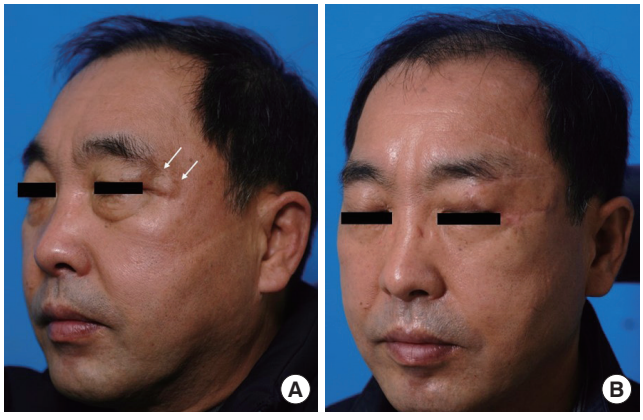

Fig. 4. Preoperative and postoperative photographs of a 61-year-old man with a primary cutaneous mucinous carcinoma. (A) Preoperative photograph showing two masses measuring 1.0×1.0 cm on the left upper eyelid. Satellite lesions are present, with the smaller lesion near the edge of the principal lesion (white arrows). (B) Postoperative photograph taken 5 months after surgery. A transposition flap was performed for periorbital reconstruction after wide excision.

with a 5-mm safety margin followed by an additional excision of the same width was performed. The resulting defect was reconstructed using a transposition flap.

Each patient received follow-up every 3 months, for a total of 3 and 2 years, respectively, to detect any local recurrence or distant metastasis. There was no evidence of recurrence or metastasis, and neither patient experienced postoperative complications such as lagophthalmos, ptosis, or severe deformity following periorbital reconstruction (Figs. 1B, 4B). In the first case, the patient exhibited symptoms of facial nerve palsy immediately postoperatively, but significant recovery was observed at the 6-month follow-up.

LITERATURE REVIEW

PCMC is a rare malignancy of the sweat glands that most commonly affects the periorbital region, followed by the scalp, face, and axilla [1]. The highest incidence rate is observed in individuals aged between 50 and 70 years [2]. Clinically, PCMCs are characterized as slow-growing, with a duration ranging from several months to years, painless, and typically measuring 0.7–2.5 cm in diameter [5]. Due to their clinical presentation, which can vary among patients, PCMCs are often misdiagnosed as benign soft tissue tumors.

The diagnosis of PCMC begins with ruling out metastasis because the majority of mucinous carcinomas in the skin are not primary cancers [6]. The gastrointestinal tract is the most common origin of metastatic mucinous carcinomas, although other sites including the breast, salivary glands, lacrimal glands, nose, and paranasal sinuses have also been reported [7]. The differentiation between primary and secondary mucinous carcinomas initially relied on histological findings and immunohistochemical patterns, which required an excisional biopsy. Requena et al. [8] identified distinctive histological features of metastatic tumors, such as larger clusters of cohesive neoplastic cells, fewer quantities of mucin, a high ratio of epithelium to mucin, and the absence of fibrous septa. The expression pattern of CK can also be a useful indicator, since metastases from the gastrointestinal tract show CK-7 negativity and CK-20 positivity, whereas PCMC exhibits the opposite pattern. A comprehensive clinical investigation, including abdominal and thoracic CT scans and positron emission tomography-CT, is then required to exclude metastasis and determine the primary site.

The primary treatment for PCMC is wide excision of the tumor due to its high recurrence rate, which can reach up to 40% in the periorbital region [1], and resistance to chemotherapy and radiotherapy [9]. However, debate continues regarding appropriate surgical margins. While some sources recommend wide excision with generous margins of at least 10 mm, there is no consensus in the literature supporting this as a definitive margin to prevent recurrence [10-12]. Choi et al. [13] recommended a safety margin of 5 mm, taking into account factors such as tumor size, frozen biopsy results, age, and ethnicity. However, it is important to note that the disease duration in their study was relatively short, and the tumor size was small, measuring less than 6 mm. Furthermore, since re-excision occurred 3 weeks after the initial surgery, various factors could have influenced the choice of the safety margin. The prognosis of PCMC is better than that of other skin cancers after complete wide excision, and metastasis to other sites is rare [6]. Nevertheless, due to the variable local recurrence rates at different sites, it is crucial to establish a proper safety margin and ensure close follow-up by the physician.

DISCUSSION

In our cases, the masses were located in the periorbital region and were initially mistaken for benign lesions because of their translucent appearance (Fig. 3). However, the first patient presented with a mass measuring 3.0 cm, while the second patient had two adjacent tumors, each approximately 1.0 cm in size. As a result, the treatment of both cases led to relatively large defects that required reconstructive surgery, utilizing a cheek flap and a transposition flap for coverage, respectively.

For accurate diagnosis and appropriate treatment, it is essential to initially confirm PCMC through an excisional biopsy. PCMC exhibits unique pathological findings, such as tumor cells encased in mucin and a characteristic CK expression pattern on immunohistochemistry [14]. Subsequently, it is important to rule out metastasis by conducting abdominal and thoracic CT scans, as well as positron emission tomography-CT. The cases we report herein adhered to this diagnostic protocol and demonstrated similar CK expression patterns and pathological features. Furthermore, because PCMC is an adnexal tumor, assessing the precise surgical margins through physical examination is inherently challenging [15]. Therefore, imaging studies, including CT or magnetic resonance imaging, are vital for determining the extent of infiltration and checking whether surrounding structures are involved [16]. For periorbital lesions, it is especially important to maintain an appropriate safety margin in order to minimize tissue loss, which has a significant impact on the risk of postoperative complications, including notching, trichiasis, conjunctivalization, entropion, retraction, ptosis, and ectropion [17].

After confirming that major structures were not involved, we initially established a safety margin of 5 mm and planned additional resection based on the results of the intraoperative frozen

biopsy. Intraoperative frozen biopsy enables simultaneous diagnosis and management [18]. Performing further resection only in the direction indicated by the frozen biopsy, rather than in all directions, enables a tissue-sparing approach. By excising tissue in only one direction, we minimized tissue loss, ultimately achieving a 10-mm margin. The patients were monitored for at least 2 years, during which they underwent a thorough diagnostic process and had their surgical margins appropriately delineated. The significance of these cases is that they demonstrate the feasibility of effective treatment with narrow surgical margins, achieved through the use of imaging modalities and a proper treatment protocol—even though our patients had a longer disease duration and larger tumors compared to the patients described in previous studies. Our findings highlight the potential for using narrow surgical margins, supported by the application of suitable imaging techniques and the information provided by frozen biopsies.

NOTES

Conflict of interest

No potential conflict of interest relevant to this article was reported.

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Ethical approval

The report was approved by the Institutional Review Board of Daegu Catholic University Medical Center (IRB No. CR-23-057-L).

Patient consent

The patients provided written informed consent for the publication and use of their images.

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