PRIMARY MUCINOUS SKIN CANCER: A CASE REPORT AND REVIEW OF THE LITERATURE

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Primary mucinous carcinoma of the skin (PMCS) is an extremely uncommon skin cancer originating from sweat glands. We present a case of a male patient diagnosed with PMCS who was initially misdiagnosed with benign sebaceous cysts. A 62-year-old male patient presented to a private clinic with a chief complaint of a slowly progressing nodule located under his lower lip for the past two years; The nodule was locally excised . The histopathology analysis yielded a diagnosis of mucinous carcinoma with a positive circumferential margin. Reexcison was performed, and the final histopathology result was a small nest of mucinous carcinoma with a negative surgical margin. He underwent thorough investigation, including gastroduodenoscopy, colonoscopy, and computerized tomography (CT) scans of the chest, abdomen, and pelvis, and found no signs of primary tumor. He was discharged from our hospital and educated to follow up closely for local recurrence. This paper illustrates the rarity of this disease, its vague presentation, and a brief review of diagnosis and treatment.

Keywords: Primary mucinous carcinoma, skin.

I. INTRODUCTION

Primary mucinous carcinoma of the skin (PMCS) is an extremely uncommon skin cancer originating from sweat glands. The typical presentation of PMCS involves the development of a slow-growing, painless mass, which can mimic other conditions such as sebaceous cysts, hemangiomas, sebaceous carcinoma, and lipomas.[1] A conclusive diagnosis of PMCS necessitates a thorough investigative process to rule out potential primary tumors, primarily from the lung, breast, and gastrointestinal tract.[2] To the best of our knowledge, there was no case report of PMCS in Vietnam. Here, we present a case of a male patient diagnosed with PMCS who was initially misdiagnosed with a benign sebaceous cyst. We also aim to provide a comprehensive literature review concerning this rare medical condition.

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II. CASE PRESENTATION

In June 2023, a 62-year-old male patient presented to a private clinic with a chief complaint of a slowly progressing nodule located under his lower lip for the past two years (Figure 1). The patient expressed his intention to have the nodule removed if feasible. The medical record of that private clinic reported that the mass was found to be firm and mobile, without invasion into the mandible. The patient's medical history revealed that he was generally healthy, with no evidence of abnormal symptoms or chronic illnesses. After the clinical evaluation, an ultrasound of the mass was performed, revealing a heteroechoic mass measuring 7x12 mm. The mass exhibited regular and well-defined borders, with no signs of increased vascularity on color Doppler ultrasound (**Figure**

2). Additional diagnostic workup, including chest X-ray and abdominal ultrasound, was conducted and did not reveal any abnormal findings. The patient was diagnosed with benign sebaceous cysts based on the initial assessment. The treatment plan involved the removal of the mass under local anesthesia, as per the patient's preference and request.

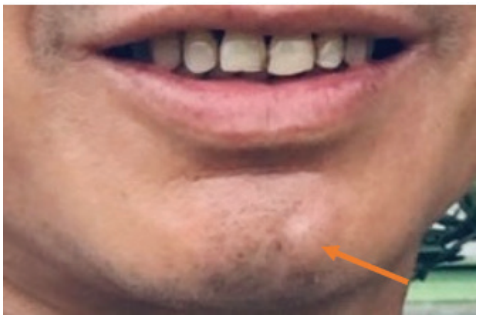

Figure 1. Clinical presentation of the tumor before resection (arrow)

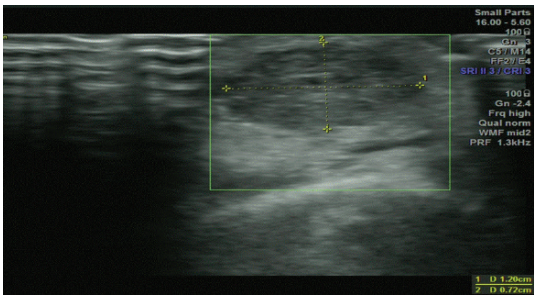

Figure 2. Ultrasound imaging of the tumor before resection

However, the histopathology analysis performed at Hanoi Medical University Hospital yielded a diagnosis of mucinous carcinoma with a positive circumferential margin. The macroscopic examination revealed a skin-covered lesion measuring 2x1.5x1 cm, which had been previously incised. Beneath the skin, a 1 cm diameter spot with a slightly mucous appearance was identified. Microscopically, the tumor was located in the mesoderm layer (**Figure 3**). The tumor cells exhibited round or oval nuclei and demonstrated relative uniformity in size. Sporadic cell division was also observed. The histological examination further revealed that the tumor cells were arranged in clusters, floating in pools of extracellular mucin, and separated by thin fibrous bands (**Figure 4**). The tumor exhibited infiltration into the muscular tissue (**Figure 5**). Notably, there was a residual tumor present at the circumferential margin (**Figure 6**). Hence, he was referred to our department for further assessment and treatment.

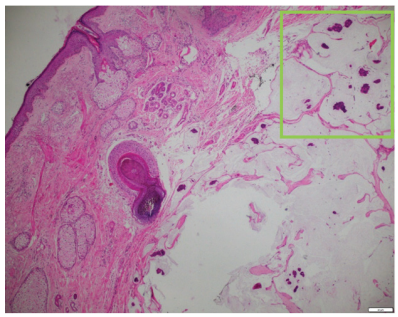

Figure 3. The tumor was located in the mesoderm layer (4x len)

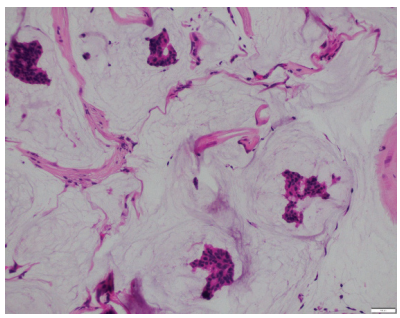

Figure 4. Islands of tumor cells floating in pools of extracellular mucin (20x len)

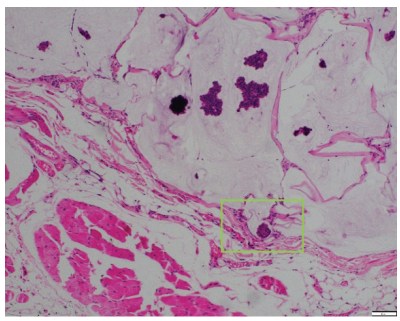

Figure 5. Tumor infiltration into the muscular tissue (10x len)

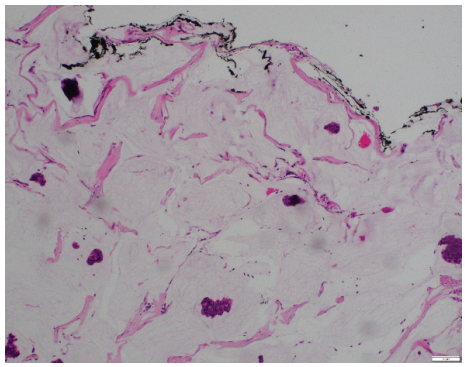

Figure 6. Positive margin (ink on tumor) (10x len)

Further investigation, including gastroduodenoscopy, colonoscopy, and computerized tomography (CT) scans of the chest, abdomen, and pelvis were found to be normal. Patient underwent re-excision with a wide margin. The final histopathology result was a small nest (< 0.5 mm) of mucinous carcinoma with a negative surgical margin. He was discharged from our hospital and will be followed up closely for local recurrence.

III. DISCUSSION

Primary mucinous carcinoma of the skin (PMCS) is an exceptionally rare cutaneous cancer arising from sweat glands.[3] It was first described by Lennox et al. in 1952 and later classified by Mendoza and Helwig in 1971.[4,5] To date, only around 200 cases have been described in the literature. There are no guideline for its management due to rarity.[6] To the best of our knowledge, this is the first case report of PMCS in Vietnam. PMCS predominantly affects individuals between 50 to 70 years of age, with a slightly higher occurrence in men than women.[6,7] Anatomically, the eyelid is the most commonly affected site (41%), followed by the scalp (17%), face (14%), axilla (9%), chest/abdomen (7%), vulva (4%), neck (2%), extremity (2%), canthus (2%), groin (1%), and ear (1%).[8–10] Our case was a 62-year-old man who falls into a few subgroups as a facial lesion.

Most cases of PMCS typically manifest as slow-growing, painless, and nodular lesions with colors that may vary from red and gray to purple. Due to their inconspicuous nature, patients may not readily seek medical attention, similar to the situation with our patient. Initial clinical evaluation often results in a broad range of potential diagnoses, including sebaceous cyst, melanoma, sebaceous carcinoma, cystic basal cell carcinoma, squamous cell carcinoma, neuroma, hemangioma, and lipoma.[1] Therefore, cytology or biopsy testing of the lesion might be necessary, especially for a growing mass like this case. This testing can assist the surgeon in planning for a wider margin during the initial definitive treatment, avoiding the result of a positive circumferential margin.

The rarity of PMCS underscores the importance of its differential diagnosis from other benign and malignant conditions, necessitating awareness and understanding of its clinical and pathological features for accurate identification and management. Immunohistochemical (IHC) staining is valuable for excluding metastatic colorectal mucinous carcinoma without CK20 expression.[11] Differentiating from breast carcinoma can be more intricate, though the occurrence of mucinous carcinoma from the breast metastasizing to the facial skin is notably rare. PMCS is typically positive for estrogen receptor, progesterone receptor, and GCDFP-15, which could also happen in primary mucinous breast cancer. [1,7] In these cases, inclusive staining for myoepithelial cells (positive for p63 and CK5/6) in an in-situ tumor component can assist in excluding metastatic mucinous breast carcinoma.[12] Nevertheless, the definitive diagnosis can only be established following a comprehensive examination that

eliminates the possibility of a more frequent primary mucinous carcinoma originating from the breast, lung, gastrointestinal tract, gall bladder, prostate, or ovary. Thus, IHC results are informative but inconclusive. Our patient is a male with no family history of breast cancer, so we performed gastroduodenoscopy, colonoscopy, and CT scans of the chest, abdomen, and pelvis to exclude the above-mentioned possible sites of primary disease. This approach saved him time and money from taking IHC testing before re-excision.

The management approach for mucinous carcinoma involves performing a local excision. Due to its notable recurrence rate, it is advisable to carry out a thorough excision with generous margins, typically at least one cm.[1,7,8] Mohs micrographic surgery has demonstrated effectiveness in various documented cases.[1,7,13] While chemotherapy and radiation are generally not utilized in treating these tumors, it is crucial to educate patients about the significance of regular follow-up appointments to monitor potential local tumor recurrence and the emergence of regional lymphadenopathy.[7]

IV. CONCLUSION

To summarize, although primary mucinous carcinoma of the skin (PMCS) is a rare disease, it does happen. Thus, physicians should always be aware of this differential diagnosis in approaching a growing tumor and carefully consider the optimal surgical margin. PMCS is usually slow-growing, never regresses, and is partly immobile. Physicians should also understand potential primary tumor locations to reach a definitive diagnosis. Thorough investigation and IHC testing may help differentiate PMCS from metastatic mucinous carcinoma originating from other sites.

Ethical approval and patient consent

This report was approved by the Head of Oncology and Palliative Care Department, Hanoi Medical University Hospital. The patient agreed to publicize his case without detailed personal information and gave written informed consent.

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