The THAI Journal of SURGERY 2024;45(3):83-89.
Official Publication of The Royal College of Surgeons of Thailand

Case Report

Primary Papillary Mucinous Carcinoma of The Scalp: A Case Report and A Brief Review of Literature

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Abstract

Background: Primary papillary mucinous carcinoma of the skin is very uncommon and is seen mainly on the head and neck region. It is difficult to differentiate clinically and pathologically between these primary carcinomas of the skin and the more commonly found mucinous carcinoma distant deposits on the skin from malignancy in the breast and gastrointestinal system.

Case Presentation: We are presenting a case of a 64-year-old lady who presented with a slowly progressive, painful ulcero-proliferative growth on her scalp for 3 years. Incisional biopsy was suggestive of mucinous neoplasm. The patient underwent an oncological workup for another primary malignancy, but no other primary malignancy was detected. Subsequently, the patient underwent wide local excision with local flap reconstruction, and on the basis of the histopathology report, the diagnosis of a primary papillary mucinous carcinoma of the scalp was confirmed.

Discussion: It is a slowly progressive, low-grade carcinoma with the propensity of local tissue invasion and a high recurrence rate. Primary from head and neck, breast, gastrointestinal tract, and pelvic organ must be excluded. Treatment with wide local excision and 1 cm margin or Moh's microsurgery is advised as these are chemotherapy and radiotherapy-resistant.

Conclusion: Primary papillary mucinous carcinoma of the scalp is a rare tumor, and another primary site of mucinous neoplasm must be ruled out. Wide local excision with adequate margin is the mainstay of treatment.

Keywords: Primary papillary mucinous carcinoma, Rare carcinoma of scalp

Received for publication 2 April 2024; Revised 25 July 2024; Accepted 20 September 2024

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Introduction

Primary papillary mucinous carcinoma of the skin is an uncommon malignancy commonly seen on the head and neck. It is very important to differentiate between primary mucinous carcinoma of the skin and the more commonly found mucinous carcinoma metastatic deposits on the skin from primaries in the breast, ovary, prostate, lung renal, and gastrointestinal systems.

Presentation of Case

A 64-year-old lady presented to our tertiary care center at Lady Hardinge Medical College with a slow-growing, painful ulcer-proliferative growth on the scalp at the right parieto-occipital region for 3 years, measuring 3 × 2.8 cm (Figure 1). The growth was firm in consistency, non-compressible, non-reducible, and fixed to the skin but slightly mobile on the skull. On examination, no lymphadenopathy or other growth in the head, neck, breast, and abdomen were found. A wedge biopsy of the growth was taken, and it showed interstitial as well as perivascular chronic inflammatory infiltrate with plasma cells. The deeper dermis shows pools of mucin with chronic inflammatory infiltrate, with few papillary structures lined by pseudostratified columnar epithelium and mild atypia. Features were consistent with mucinous neoplasia. As it was important to identify the primary mucinous neoplasm of the scalp from metastatic tumor deposit, a whole-body evaluation by clinical and radiological (CECT) was done to look for primary malignancy in other head and neck regions, breast, and gastrointestinal tract. (Figure 2-4). The patient undergoes wide local excision with a 1 cm margin and local rotational flap reconstruction of the defect and drain insertion under general anesthesia (Figure 5-8). Histopathology (Figure 9) shows hyperkeratosis, keratotic plugging, and follicular plugging in the epidermis. The dermis shows a lobulated tumor composed of cells arranged in papillary architecture separated by pools of mucin. Tumor cells are lined by tall columnar cells showing stratification at some places, round to elongated nuclei with moderate pleomorphism, irregularly distributed chromatin, and 1-2 prominent nucleoli in many cells. IHC (Figure 10-13) shows ER: positive, PR: positive, GATA 3: positive, CK 7: focal positive, CK 20: negative, p63: negative, and CK5/6: negative with the impression of papillary mucinous carcinoma. All margins were uninvolved by the tumor. Follow-up after 3 months shows a healthy site.

Figure 1 Showing ulceroproliferative growth on the scalp at the right parieto-occipital region

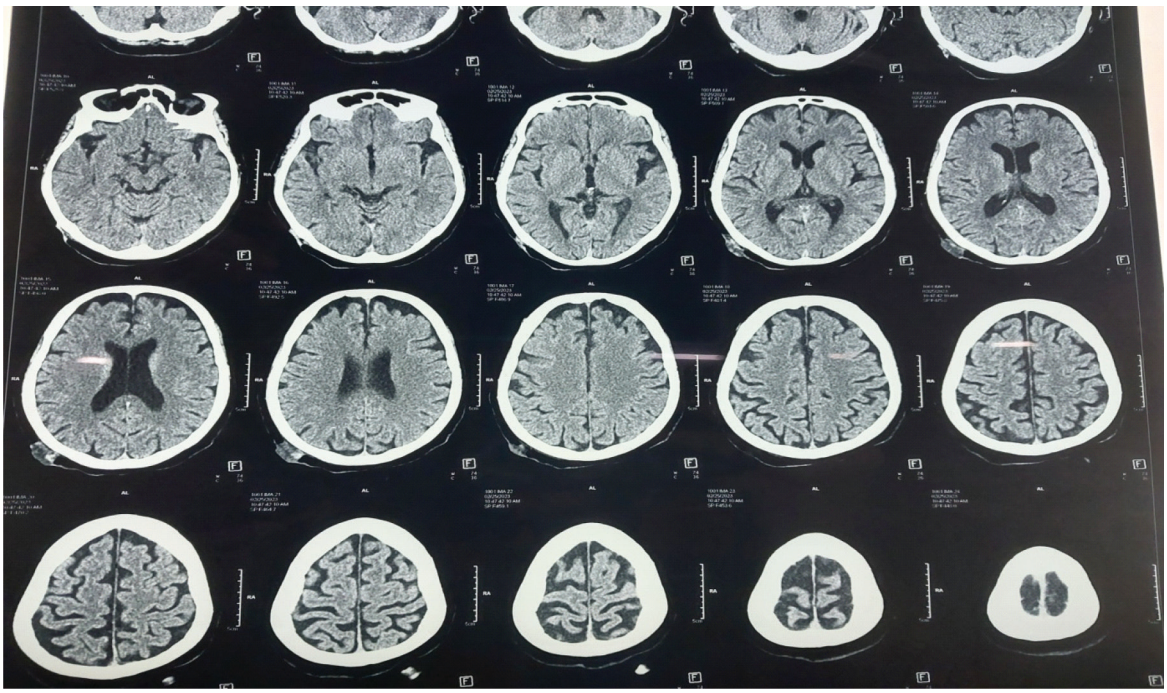

Figure 2 NCCT head showing no pericranium involvement by growth

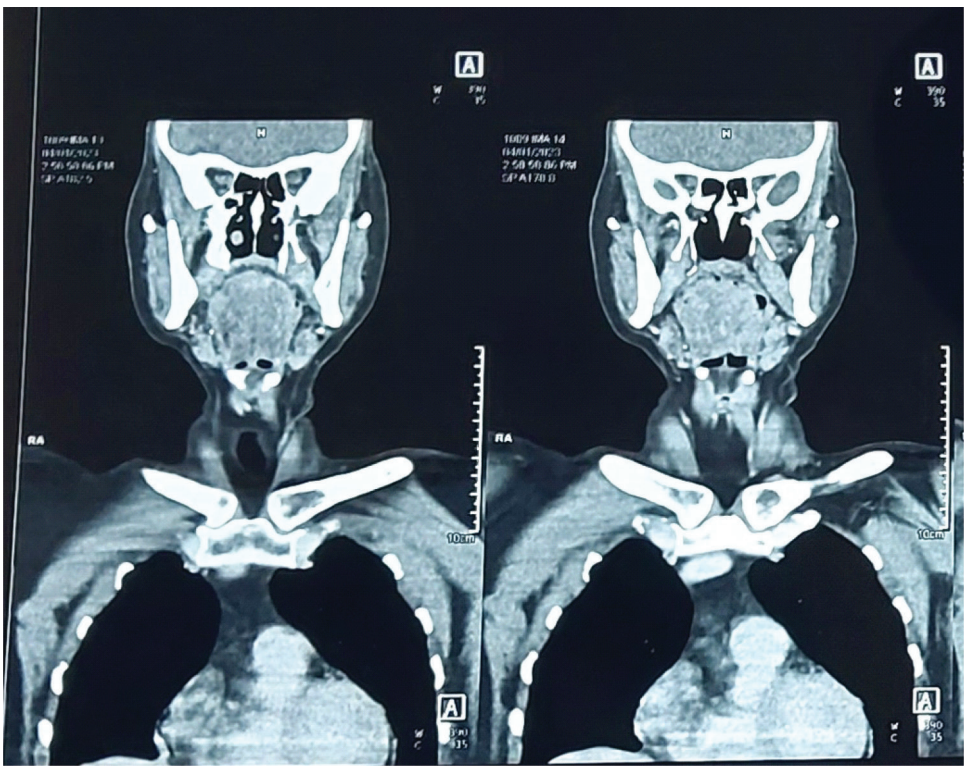

Figure 3 CECT showing no head, neck breast malignancy

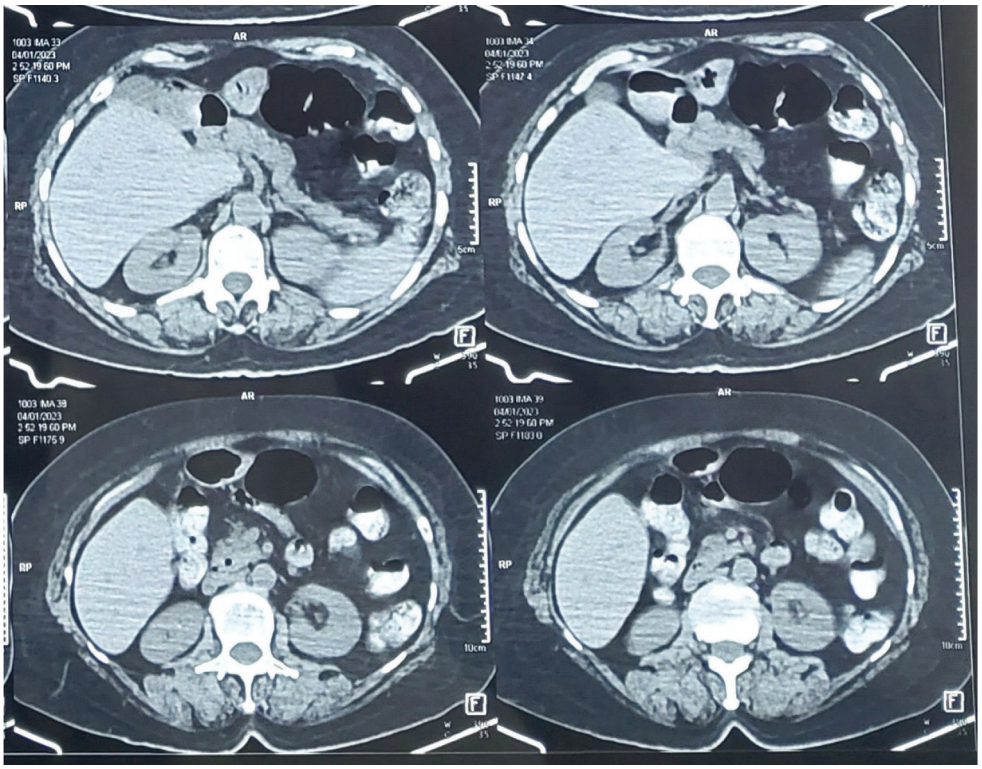

Figure 4 CECT showing no gastrointestinal tract malignancy

Figure 5 Showing preoperative marking for wide local excision with 1 cm margin and local rotational flap reconstruction

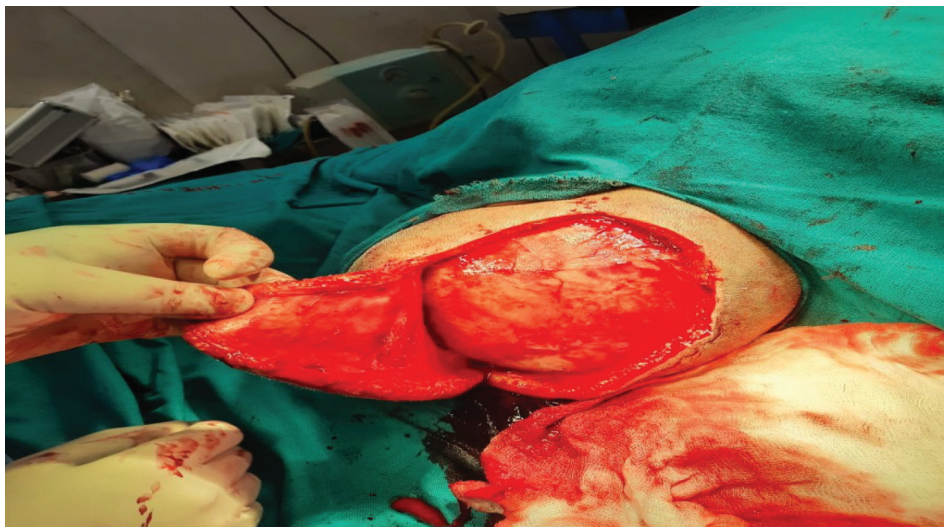

Figure 6 Showing excision of growth and creation of local rotational flap

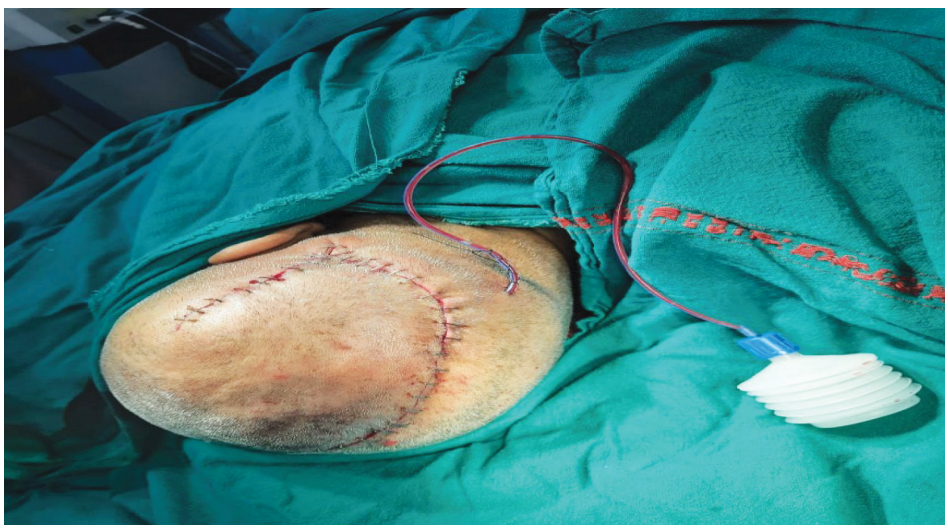

Figure 7 Showing the final reconstruction and suction drain placement beneath the flap

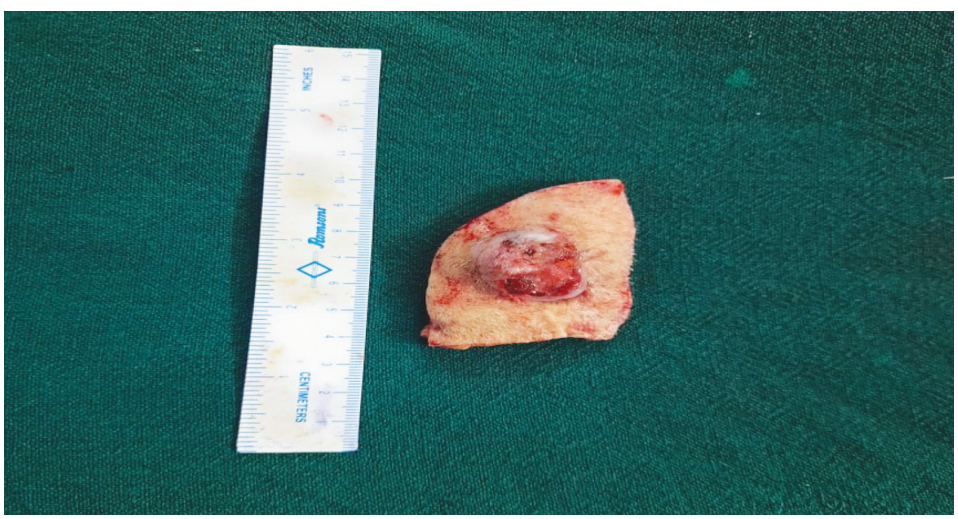

Figure 8 Showing a wide local excised specimen with 1 cm margin

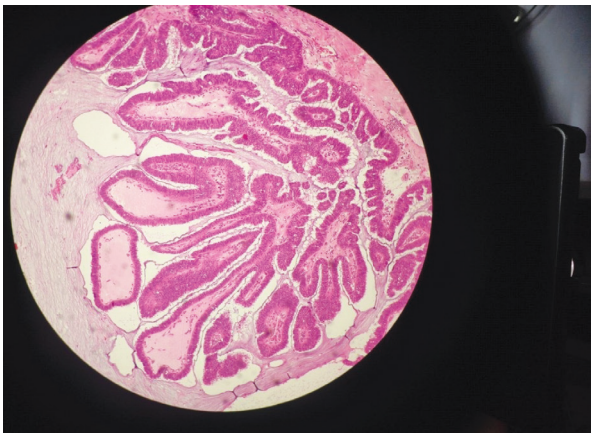

Figure 9 Histopathological image of the final resected specimen showing papillary mucinous carcinoma

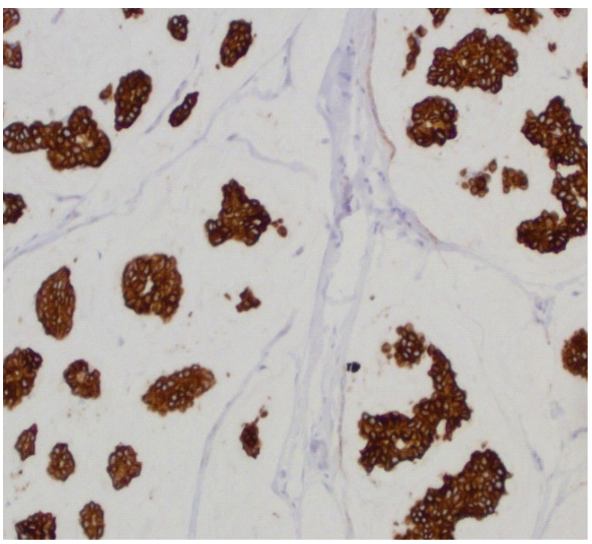

Figure 10 CK7 focal positive

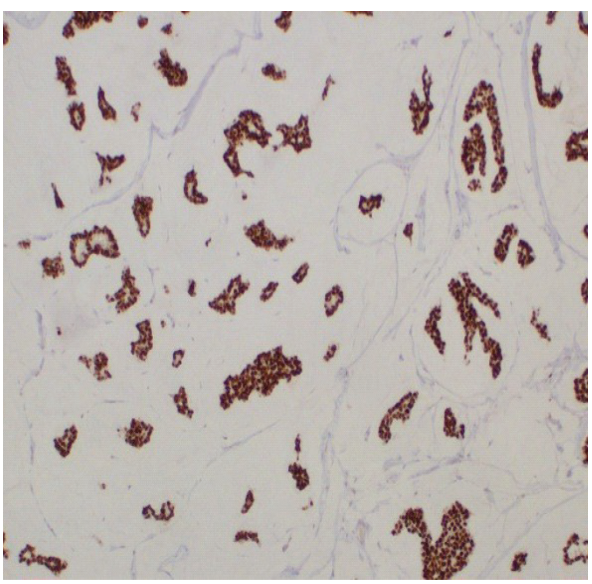

Figure 11 GATA3 positive

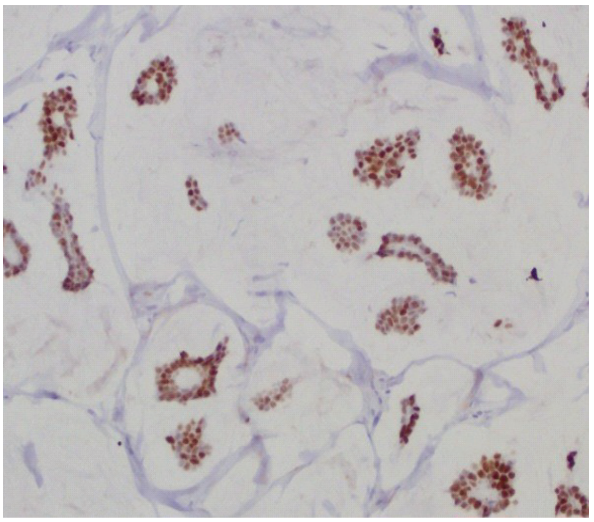

Figure 12 ER positive

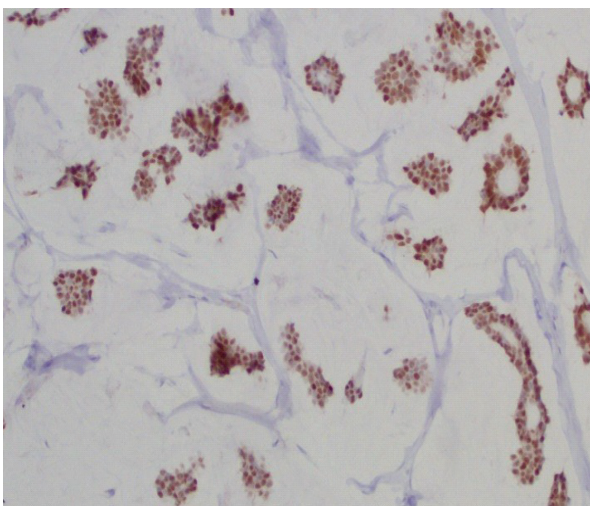

Figure 13 PR positive

Discussion

Primary papillary mucinous carcinoma of the scalp is a rare malignancy. These tumors are slow-growing, painless, soft to firm, indurated, and have ulcerated growth for a longer duration before presentation.[1] These are low-grade malignancies with high recurrence at the local site (19.6%) and a distant metastasis rate of (6.1%).[2] Metastases are commonly seen in the loco-regional lymph nodes.[3,4] In 2–7 % of cases, distant metastases have been reported.[5] Mucinous carcinoma invades local tissues by direct extension, by satellite islands of tumor, and by loco-regional lymph node involvement.[6,7] Mortality is seen due to multiple recurrences and metastatic disease.[8]

Primary skin neoplasms must be identified and differentiated from more common mucinous metastatic deposits on the skin from primaries in the breast, prostate, ovary, lungs, gastrointestinal system, and renal system.[9,10] Differentiation of secondary deposits from primary mucinous skin malignancy should be done by clinical examination, radiological evaluation, and histopathology. Surgical-wide local excision of primary with negative margin is the mainstay of treatment. To prevent a recurrence, a margin of at least 1 cm) is advised for wide local excision. Moh's micrographic surgery is a very useful procedure for achieving negative margin resection. It has been seen that these tumors are resistant to radiotherapy and chemotherapy; hence, surgery is the mainstay of treatment.[9,11]

Conclusion

Primary papillary mucinous carcinoma of the scalp is an uncommon malignancy of the scalp, and primary mucinous carcinoma of the breast, gastrointestinal system, ovary, prostate, and the renal system must be ruled out as most skin mucinous carcinoma are metastatic deposits from these primaries. It is a low-grade, slowly progressing tumor with a tendency for multiple local recurrences, regional lymph node involvement, and a low distant metastasis rate. Wide local excision with adequate margin is the mainstay of treatment as these are chemo & radioresistant.

Conflicts of Interest

There is nothing to declare.

Sources of Funding

There is nothing to declare.

Acknowledgement

Department of Pathology, Lady Hardinge Medical College, for providing histopathological images.

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