

## A Case of Metastatic Eccrine Porocarcinoma with a Review of the Literatures

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### 전이성 에크린 한공암증 1예 및 문헌고찰

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에크린 한공암증은 흔하지 않으며, 전이성 에크린 한공암증은 매우 드문 암이다. 에크린 한공암증은 표피내 한관에서 기원하는 피부 부속기 악성 종양으로, 대부분 에크린 한공암증은 항암치료와 방사선치료에 효과가 없으며, 따라서 유일한 치료방법은 전이되기 전에 국소 및 광범위 절제술을 시행하여야 한다. 본 환자는 두피에 발생한 에크린 한공암증이 점점 진행하여 피부, 림프절, 폐 등으로 전이하였고, 다수의 광범위 절제술에 이어 항암치료와 방사선치료를 받았지만, 결국 폐전이로 악화되어 사망하였다. 본 저자들은 항암치료와 방사선치료에 불응한 에크린 한공암증 1예를 치료에 대한 문헌고찰과 더불어 보고하는 바이다.

**중심 단어** : 에크린 한공암증 · 항암치료 · 방사선치료.

### Introduction

Eccrine porocarcinomas are rare malignant tumors of the sweat glands arising from the acrosyringium. Between 30% and 50% of porocarcinomas originate from benign poromas. The first case was described in 1963 by Pinkus and Mehregan as an epidermotropic eccrine poroma.<sup>1,2</sup> The lower extremities are the most commonly affected site, followed by the trunk and the head. Some cases have multiple metastases to the skin, local lymph nodes, and visceral organs, which are fa-

tal. In Korea, with the exception of several cases of eccrine poromas with no metastasis, there have been few reported cases of metastatic porocarcinomas. With a review of the literature, we report a case of eccrine porocarcinoma that originated from the scalp that recurred repeatedly, despite performance of several surgical resections followed by chemotherapy and radiotherapy.

### Case Report

A 63-year-old woman sought evaluation at the Plastic Surgery Clinic in our hospital because of a mass that had been growing rapidly around the left parietal scalp since 2002(Fig. 1). In 2008, the 3×3cm sized mass was excised completely, and an eccrine porocarcinoma was diagnosed by biopsy (Fig. 2).

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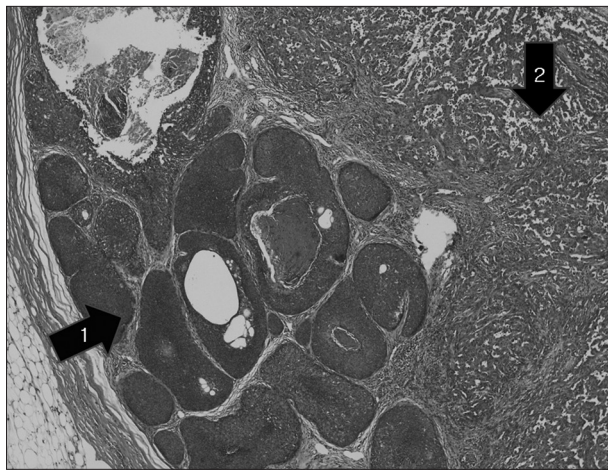
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Histopathologic examination showed an asymmetric neoplasm with poorly circumscribed margins and neoplastic cells with marked variation in size and shape. The tumor had an intra-epidermal and invasive intradermal component with an infiltrative growth pattern that involved the margin of re-



**Fig. 1.** There is a 3×3cm sized mass at the parietal region on the scalp.



**Fig. 2.** Microscopic finding of eccrine porocarcinoma with apocrine differentiation. The eccrine porocarcinoma area (Arrow 1) shows variable-sized tumor cell nests containing central tumor necrosis and ductal lumina. The right side of the figure (Arrow 2) shows papillary and tubular growth of pleomorphic cells representing apocrine differentiation (× 40, H & E stain).

section. Immunohistochemical staining of the ductal structures demonstrated carcinoembryonic antigen (CEA) and epithelial membrane antigen (EMA). In addition, results of cytokeratin 7, 20, and 5/6 stains were positive.

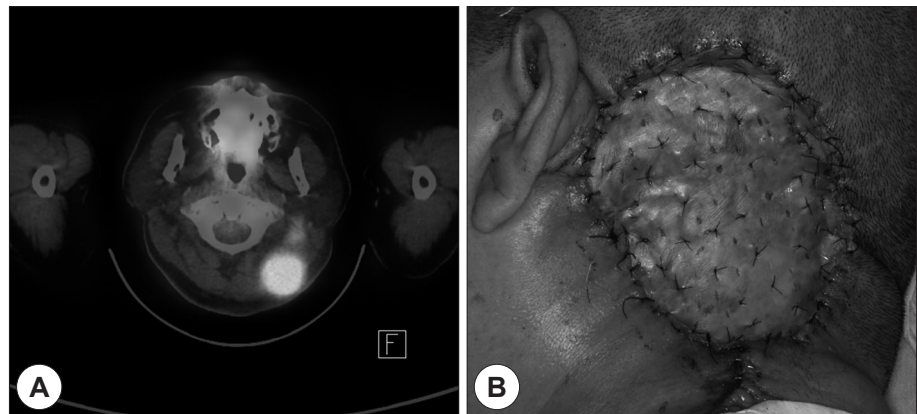
Radiation was delivered to the post-operative scalp area (2,200cGy/11 fraction). Seventeen months later, the tumor recurred in the left suboccipital area with regional lymph node metastasis out of the past radiation area (Fig. 3A). There were no other metastasis on image study including PET CT and neck CT. Wide excision and modified radical neck lymph node dissection were performed (Fig. 3B). Nevertheless, recurrences occurred again at the left parietal and frontal areas within two months. Wide excision was performed repeatedly.

We decided to administer infusion 5-FU chemotherapy (900mg/m<sup>2</sup> for three days every three weeks). Just after the second cycle of 5-FU, disease progression was confirmed by neck CT, which showed new recurrence in the left parotid and posterolateral neck around the second operation bed. Adriamycin (50mg/m<sup>2</sup> for one day every three weeks) as the second-line chemotherapy was administered. Nevertheless, small nodules developed in the scalp and gradually around the neck. Weekly paclitaxel (80mg/m<sup>2</sup> for one day every week) as the third-line chemotherapy was given. Lung metastasis



**Fig. 4.** Chest X-ray showed multiple hematogenous lung metastasis in both lung fields.

**Fig. 3.** A : PET (positron emission tomography) CT showed an abnormal hypermetabolic lesion in the left suboccipital area and left cervical level II lymph node. B : Postoperative picture showed split-thickness skin graft after wide excision of left suboccipital mass and modified radical neck dissection.



developed during the third line chemotherapy(Fig. 4). As a result, we discontinued the chemotherapy and added palliative radiotherapy(2,800cGy/14 fraction) to the painful neck area. She ultimately expired due to refractory respiratory failure from lung metastasis aggravation.

## Discussion

Eccrine porocarcinomas mainly affect individuals in the 6<sup>th</sup>–8<sup>th</sup> decades, and there is no difference in prevalence between males and females. The lower extremities(44%) are the most commonly affected site, followed by the trunk(24%) and head(18%) and have no relationship with the density of the eccrine glands of the skin.<sup>4)</sup> The tumor can appear as a nodule or a verrucous, dome-shaped, infiltrating, or erosive plaque, or as a polypoid growth that is frequently ulcerated. The average size of the primary lesion is 2.4cm, ranging from 1–10cm.<sup>5)</sup>

Histopathologically, the tumor may remain completely intra-epidermal(in situ porocarcinoma), but it is often associated with an invasive intradermal component, as in our case.<sup>6)</sup>

Some reports have evaluated different histologic findings that may provide a guide to prognosis. Those reports have concluded that a more aggressive clinical course may be indicated by >14mitoses per 10 high-power fields, lymphovascular invasion by tumor, and depth >7mm. It has also been suggested that tumors presenting with an infiltrative advancing margin are particularly prone to local recurrence.<sup>4,7)</sup> In our case, histopathological analysis exhibited high mitotic activity(65 mitosis per 10 high-power fields) and showed an infiltrative advancing margin.

Patients with metastasis to the viscera or the regional lymph nodes show detrimental prognosis. The survival duration for such patients has been reported to be between 5 and 24 months. Distant metastasis occur in the lung, breast, retroperitoneum, peritoneum, liver, bladder, ovary, and femur.<sup>8)</sup>

Due to the propensity to local recurrence, the treatment of choice is wide local excision with histologic confirmation of the tumor-free margins. Sentinel node biopsy may be indicat-

ed when risk factors are present, including deep invasion, high mitotic activity rate, and lymphovascular infiltration on cutaneous cancer of the head and neck.<sup>9,10)</sup> A wide excision of the primary lesion is curative in 70–80% of cases. Lymph node dissection is recommended in the case of a positive lymph node biopsy and clinical lymph node involvement.<sup>11)</sup>

We report a rare case of eccrine porocarcinoma that showed aggressive features and was refractory to chemotherapy and radiotherapy. The optimum treatment for metastatic eccrine porocarcinomas has not been standardized. Eccrine porocarcinomas have generally been refractory to multiple treatment regimens, as in our case. Radiotherapy has not been effective, and the benefits of chemotherapy are generally poor.

The cases recurred together with lymphadenopathy and were given combined treatment based on 1) excision of the lesion, 2) regional lymphadenectomy, and 3) radiotherapy or chemotherapy with palliative or adjuvant intent.<sup>12-17)</sup> Although long term overall survival has been rarely reported, there are several cases that showed favorable responses to chemotherapy or radiotherapy and long term progression free survivals longer than 10months(Table 1). The first complete response case was reported in a patient with cutaneous and lymph node metastasis following melphalan and intra-arterial infusion of 5-fluorouracil(5-FU) associated with regional hyperthermia. Another case of complete response was reported in a patient with lymph node, pleural, and pericardial metastasis, though the response lasted for only three months following 5-FU in continuous infusion.<sup>14)</sup> A case with complete response for 16months was reported following combination chemotherapy consisting of doxorubicin, cisplatin, mitomycin C, and vincristine.<sup>13)</sup> Although single-agent treatments such as docetaxel, paclitaxel and 5-fluorouracil(5-FU) have some antitumor activity in metastatic disease, combination chemotherapy demonstrating prolonged response would be considered in the first-line treatment.<sup>12-16)</sup> Recently, concurrent chemoradiotherapy was shown to induce complete remission in some cases of metastatic porocarcinoma and should be considered in patients of advanced disease.<sup>14,17)</sup> In particular, ra-

**Table 1.** Treatment outcomes of recurrent metastatic eccrine porocarcinoma after surgery

Age/Sex	Met.	Treatment	Response	PFS(Mon.)	Reference
42/M	–	Cyclophosphamide/Cisplatin/Doxorubicin	PR	21	2
67/M	–	Paclitaxel/IFN-a	PR	>7	15
45/F	Lung	Docetaxel	PR	>5	12
71/M	–	Isotretinoin+RTx, Isotretinoin/Tegafur	CR SD	11 > 66	14
61/M	–	5-FU/Cisplatin+RTx	CR	>10	17
20/M	Lung, Bone	Paclitaxel/Carboplatin+RTx	PR	16	16
45/F	Lung, Bone	Doxorubicin/Mitomycin-C/Vincristine/Cisplatin	CR	16	13

Met. : Distant metastasis, RTx : Radiotherapy, PFS : Progression free survival, IFN-a : Interferon-a, CR : Complete remission, PR : Partial remission, SD : Stable disease

diotherapy and oral isotretinoin, subsequently substituted by tegafur and oral isotretinoin, achieved a long term survival without distant metastasis.<sup>14)</sup> As in this case, oral isotretinoin may be considered as maintenance therapy after chemotherapy or radiotherapy. Nevertheless, no standard therapeutic protocols for metastatic porocarcinoma have been established. Therefore, not only prospective studies but retrospective studies are warranted to further investigate the pathogenetic mechanisms, prognostic factors, and management of this rare and aggressive tumor.

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#### Conflicts of Interest

Conflict of interest relevant to this article was not reported.

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