

A Case of Metastatic Eccrine Porocarcinoma

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Sir,

Primary adenocarcinomas arising from the eccrine sweat glands are rare and represent approximately 0.005% of epithelial cutaneous neoplasms (1). Eccrine porocarcinoma, otherwise known as malignant eccrine poroma, is the most common variant among them (2). It was first described by Pinkus & Mehregan in 1963 (3) as epidermotropic eccrine carcinoma. However, in 1969, Mishima & Morioka (4) introduced the term "eccrine porocarcinoma". Approximately 20% of cases of eccrine porocarcinoma recur locally after removal (5) and another 20% of cases metastasize to the regional lymph nodes (6). More rarely, widespread visceral metastases may occur (7–11).

CASE REPORT

A 42-year-old Korean man visited his local clinic in February 1998 for the treatment of an erythematous verrucous plaque on his right palm. The lesion was completely excised at that time, but a pathological examination was not performed. In November 1999, a few erythematous papules developed on his right palm and wrist with right axillary lymphadenopathy. The lesion on his palm was excised and a fine needle aspiration was performed on his right axillary lymph node. A diagnosis of malignant nodular hidroadenoma was made at that time. Some of the lesions were excised widely and right axillary lymphadenectomy was performed. Adjuvant chemotherapy, with cyclophosphamide, cisplatin, and doxorubicin, was then given for 6 months. Over the next 2 years, several erythematous papules and plaques developed on his right wrist, arm and right axilla. From 2002 to 2004, wide excisions were carried out 4 times.

In February 2006, he presented to our department with mental deterioration. A physical examination revealed multiple, various sized, erythematous papules and plaques on his face, right arm, right axilla and trunk (Fig. 1A and B). Some of them were eroded

and bleeding. A neurological examination showed that he had a deep drowsy mentality and left hemiparesis. A brain computed tomography (CT) revealed multiple brain tumours with tumour bleeding. Emergency craniectomy was performed to remove the tumours. Histologically, the brain tumour consisted mainly of basaloid cells with ductal structures being found within the nest. After surgery, he became alert and was evaluated for multiple distant metastases. Abdominal CT, chest CT, T-spine magnetic resonance imaging, and skin biopsy were performed. The radiological findings demonstrated multiple metastases to the lung, pleura, bone, muscle and spinal cord. Histological examination of the skin lesion on his arm, revealed basaloid tumour cells with continuity with the surface epithelium and downward infiltrating growths with broad anastomosis. Ductal differentiation and intracytoplasmic lumen formation were found on the tumour nest (Fig. 2A). The tumour also showed nuclear pleomorphism and abnormal mitotic activity focally. Immunohistochemical staining of the ductal structures demonstrated the carcinoembryonic antigen (CEA) (Fig. 2B) and epithelial membrane antigen (Fig. 2C). A diagnosis of eccrine porocarcinoma was therefore made.

In May 2006, the patient was treated with gamma knife radiosurgery and radiotherapy for the metastatic brain lesions, and palliative radiotherapy for the metastatic spinal cord lesions. He refused further treatment and was placed in palliative care.

DISCUSSION

Eccrine porocarcinoma is a rare malignant tumour of the sweat gland arising from the acrosyringium (1). These lesions are most commonly found on the lower extremities, followed by the head, scalp, upper extremities, trunk and abdomen (8, 12). Eccrine porocarcinoma affects mainly elderly people (14). It can appear as a nodule or a verrucous, dome-shaped, infiltrated or erosive plaque or as a polypoid growth that is frequently ulcerated. Multi-nodularity, ulceration and rapid growth may be associated with either local recurrence or metastasis. The average size of the primary lesion is 2.4 cm, ranging from 1 to 10 cm (8). Our patient presented with multiple erythematous papules, plaques and nodules, some of which were eroded and bleeding.

Histopathologically, the tumour arises from terminal cells of the intra-epidermal portion of the eccrine sweat duct (5, 14). The intra-epi-



Fig. 1. Multiple erythematous papules, plaques and nodules, (A) on the face and (B) on the right axilla and arm.

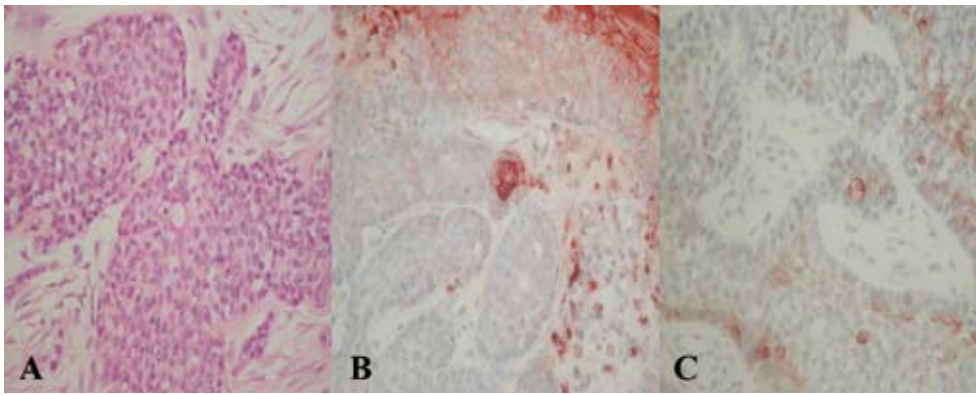


Fig. 2. (A) Basaloid cells showed a downward infiltrating growth pattern with ductal differentiation and intracytoplasmic lumen formations (H&E $\times 400$). Ductal structures shown by (B) carcinoembryonic antigen staining (CEA $\times 400$) and (C) epithelial membrane antigen staining (EMA $\times 400$).

dermal portion is recognized by the presence of basaloid cells with typical ductal lumina that are associated with pleomorphism, nuclear hyperchromatism and mitotic activity. Invasive eccrine porocarcinoma shows continuity with the surface epithelium and might be associated with a broad pushing deep margin or a more obvious infiltrative lower border. Ductal differentiation and intracytoplasmic lumen formation is invariably evident and is rendered more obvious by immunohistochemical staining, such as for epithelial membrane antigen (EMA) and CEA (4, 6, 15). The histological findings in our case revealed a striking infiltrative growth pattern, basaloid cells with ductal differentiation and intracytoplasmic lumen on the skin and brain tumours. They were highlighted by EMA and CEA staining. Eccrine porocarcinoma can occur as lymphovascular invasion and pagetoid extension of the neoplastic cells (4, 5, 7–9). This pattern of invasion leads to regional cutaneous metastases and lymph node invasion, with the subsequent risk of an internal metastasis (7–9). In our case, although lymphovascular invasions could not be found on the histology section, axillary lymph node invasion and distant metastases were present.

The prognosis of eccrine porocarcinoma varies. In a majority of cases it grows slowly and initial surgical treatment is usually curative. However, approximately 20% of these tumours recur locally and/or metastasize to the regional lymph nodes, resulting in an elevated mortality rate (3, 5, 6–8). Less frequently, distant metastases to the internal organs can occur. The prognosis of widespread metastatic eccrine porocarcinoma is generally fatal regardless of treatment (7–11). In our case, new lesions recurred locally several months after removal of the primary lesion. Metastases to the axillary lymph node and multiple distant metastases to the lung, spinal cord, muscle and bone were found.

The optimum treatment for metastatic eccrine porocarcinoma has not been standardized. Because of the propensity to develop local recurrences, the treatment of choice is a wide local excision of the primary tumour with histological confirmation of the tumour-free margins (16). Prophylactic lymphadenectomy in eccrine porocarcinoma is controversial. Nevertheless, it is be-

lieved that the excision should be performed if there is evidence of lymphadenopathy or when there are recurrent tumours with signs of histological aggressiveness or intralymphatic permeation (6, 8). In our case, cancer cells were detected in the fine needle aspiration of the right axillary lymph node and an axillary lymphadenectomy was performed.

The benefits of chemotherapy in the treatment of metastatic eccrine porocarcinoma are generally poor. Several chemotherapeutic agents, including docetaxel (13), have shown a partial response. In our case, the patient was treated with 6 cycles of chemotherapy with cyclophosphamide, cisplatin and doxorubicin, but with an unsatisfactory response.

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