

Eccrine Porocarcinoma of the Face

Dr. Y. P. Fung

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extended fairly close to the deep resection margin. Immuno-histochemical studies showed that the tumor cells were positive for CAM5.2 (low molecular weight cytokeratin) and the ductal structure was highlighted by carcinoembryonic antigen (CEA) and AE1/AE3 (high molecular weight cytokeratin) stains. The histopathology was that of eccrine porocarcinoma.

CASE SUMMARY

History

A 74-year-old housewife presented to South Kwai Chung Skin Clinic with one month history of an ulcerating nodule on her right cheek. It was non-itchy and painless. It was slowly enlarging and had bled occasionally. There was no history of trauma or prolonged sunlight exposure. Apart from mild chronic bronchitis, the patient was fit and well.

Physical examination

A non-specific ulcerating nodule, with a regular border, measuring 0.5cm in diameter, was noted on the right cheek (Figure 1). It was non-tender and there was no subcutaneous infiltration clinically. There was no lymphadenopathy or organomegaly.

Differential diagnosis

The differential diagnoses included seborrhoeic keratosis, pyogenic granuloma, basal cell carcinoma, squamous cell carcinoma, amelanotic melanoma and other epithelial tumors.

Investigations

Her chest X-ray, complete blood count, renal and liver function tests were all normal. Excisional skin biopsy showed a circumscribed nodular growth in the dermis, composing of anastomosing nests and cords of cells separated by fibrous stroma (Figure 2). Focal glandular differentiation with ductal structure was noted. The tumor cell nuclei were round to oval, with coarse chromatin, and small but distinct nucleoli. Mitotic figures were frequently seen. Moderate nuclear atypia and high nucleo-cytoplasmic ratio were noted in places. No perineural or vascular invasion was seen. The overlying epidermis was not involved. The tumor

Diagnosis

The diagnosis of eccrine porocarcinoma was made.

Follow-up

Her wound healed well and there was no evidence of recurrence. She was arranged to be followed up every three months.

Discussion

The diagnosis of eccrine porocarcinoma was made by the combination of clinical history, examination and histology. Histology alone was insufficient because it was essentially that of an adenocarcinoma. If the patient was cachexic and had multiple cutaneous nodules, the diagnosis of metastatic secondary deposits had to be considered. The latter is more common than eccrine porocarcinoma. The primary lesion could be from the breasts, pancreas, prostate and large intestine.

REVIEW ON ECCRINE POROCARCINOMA

Eccrine porocarcinoma is a rare malignant tumor arising from the acrosyringium (intraepidermal ductal portion of the eccrine sweat gland). Its benign counterpart is the eccrine poroma. Since eccrine poroma is a rather common condition, it is surprising to find that eccrine porocarcinoma represents only 0.005% of all epithelial skin neoplasms.¹ There were less than 200 reported cases in the world literature. It was first reported by Pinkus and Mehregan in 1963 as 'epidermotropic eccrine carcinoma'. Since then, numerous terminology had been used, including epidermotropic eccrine carcinoma, eccrine porocarcinoma, malignant hidroacanthoma simplex, poroepithelioma, malignant intraepidermal eccrine poroma and malignant syringoacanthoma.² It has been suggested that the



Figure 1: A non-healing ulcerating nodule on the right cheek

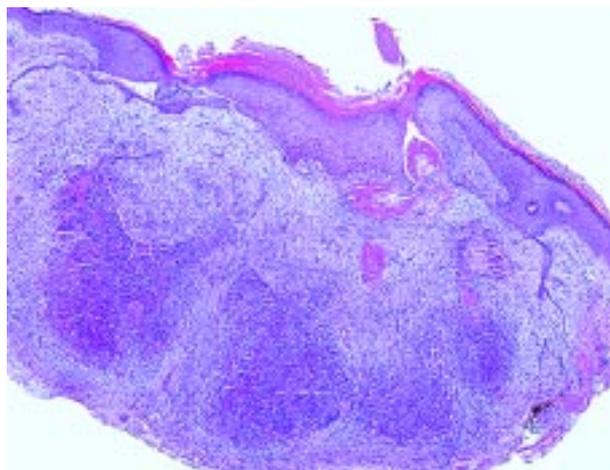


Figure 2: Low power view (40x) showing multiple islands of tumour cells with irregular border in the dermis. The surrounding stroma is desmoplastic. (By courtesy of Dr. K. C. Lee, Department of Pathology, PMH)

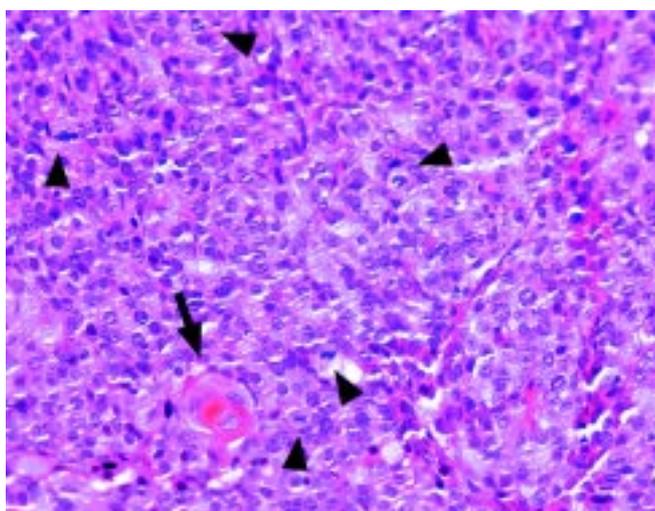


Figure 3: High power photomicrograph (400x) showing round to oval tumour cells with scanty cytoplasm and numerous mitotic figures (arrow heads). Focally there is ductal differentiation (arrow). (By courtesy of Dr. K. C. Lee, Department of Pathology, PMH)

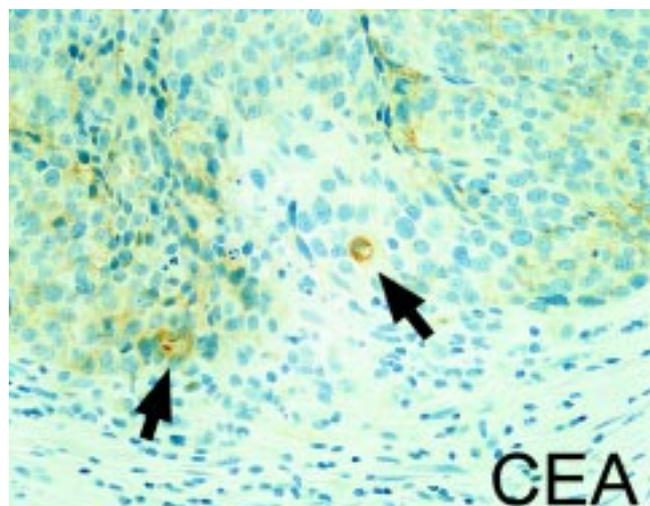


Figure 4: The tumour is immunoreactive for carcinoembryonic antigen (CEA), especially around the cuticular cells (arrows). (By courtesy of Dr. K. C. Lee, Department of Pathology, PMH)

condition might be under-reported because of the confusing terminology. Eccrine porocarcinoma is the term used in most large dermatology textbooks nowadays.

Clinical features

Eccrine porocarcinoma tends to affect older people (average 67.5 years) but the age range can be wide (from 19-94 years).¹ Sex incidence is equal. Typically there is a rather lengthy duration from its onset till treatment

(average 8.5 years). Because of such a long duration, it has been postulated that eccrine porocarcinoma arises from malignant transformation of pre-existing benign lesion. This theory, however, cannot be substantiated because the distribution of porocarcinoma and that of poroma are totally different. Eccrine poroma tends to concentrate at sites where there is a high concentration of sweat glands, especially the palms and soles. Eccrine porocarcinoma however is rarely found at these sites. They were reported to be found at the legs (55%), head (20%), upper limbs (12%), trunk and abdomen (10%)

and the rest (3%).¹ It may present as a nodule (red, light brown or flesh-coloured), plaque (verrucous, infiltrative, erosive) or polypoid growth. Like other epithelial tumors, multi-nodularity, rapid growth and ulceration are signs of recurrence or metastasis.

Histopathology

Due to its origin, eccrine porocarcinoma tends to have epidermotropism. It can invade locally and downwards into the dermis and subcutaneous tissue. On reaching dermal lymphatics, it can re-invade the epidermis. Presenting as intraepidermal proliferation of well defined nests of small anaplastic tumor cells, the epidermis may be ulcerated. Tumor cell has pleomorphic and hyperchromatic nucleus and clear cytoplasm. Small squamoid cells that form ducts lined by a cuticle are often found. PAS-positive, diastase-labile glycogen deposits can be found in the cytoplasm. In immunohistochemical studies, it shows positive staining towards CEA (ductal lumina and periluminal cells), epithelial membrane antigen and cytokeratins, but negative staining towards S-100 marker.

Prognosis

The prognosis is difficult to be estimated. In general, those patients who have lesions completely excised and have no metastasis do well. It has been reported that 20% of eccrine porocarcinoma recur locally at 4 months to 12 years after excision. In addition, 20% of the reported cases have metastases to lymph nodes and these are associated with a mortality rate of 67%.² Less commonly, porocarcinoma may invade large organs such as lungs, peritoneum and liver. This is associated with poor prognosis.

Treatment

Removal of the primary tumor is the mainstay of treatment. Wide local excision with histological confirmation of clear margin is the most accepted method. Prophylactic lymph node dissection is sometimes considered if the porocarcinoma is poorly-differentiated and there is intra-lymphatic permeation in histology. Lymphadenectomy is performed when there is regional lymph node involvement. Radiotherapy is unhelpful as the tumor is generally not radio-sensitive. In metastasized disease, intensive combination chemotherapy with doxorubicin and cyclophosphamide, or epirubicin, mitomycin, vincristine, carboplatin and 5-fluorouracil had been used. Huet et al reported success in clearing eccrine porocarcinoma metastasis in a patient by using interferon alfa-2a (9 million units, 3 times per week) for 9 months and isotretinoin (2mg/kg/day) for 2 months.

Learning points:

In immunohistochemical studies, eccrine porocarcinoma shows positive staining to CEA, epithelial membrane antigen and cytokeratins, but negative staining to S-100.

References

1. Huet P, Dandurand M, Pignodel C, et al. Metastasizing eccrine porocarcinoma: report of a case and review of the literature. *J Am Acad Dermatol* 1996;35(5 Pt 2):860-4.
2. Snow SN, Reizner GT. Eccrine porocarcinoma of the face. *J Am Acad Dermatol* 1992;27(2 Pt 2):306-11.