Eccrine Porocarcinoma: A Case Report

E Tan,* MBBS, E S W Ang,** MBBS, FRCS (Glas), C L Foo,*** FAMS, FRCS (Engl), FRCS (Edin)

Abstract

We report a 58-year-old Chinese female who presented with a pigmented skin lesion on the right thigh, associated with a single ipsilateral inguinal lymph node without distant metastases detected. A wide excision of the tumour, split skin graft coverage and an en-bloc ipsilateral superficial groin dissection was performed. Histopathology revealed malignant skin adnexal tumour or porocarcinoma, with nodal metastases. She is presently at six months follow-up without any recurrence.

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Key words: Early recognition, Intraepidermal growth, Lymphatic spread, Rare tumour

Introduction

Porocarcinoma is a rare malignant tumour, arising from the intra-epidermal ductal portion of the eccrine sweat gland, also known as "acrosyringium".[1] According to the histological typing of skin tumours,* it is an eccrine carcinoma that may arise de *novo* or develop in eccrine poroma, its benign counterpart, as an entirely intra-epidermal tumour or with an invasive dermal component. To date, there have been about 175 cases of porocarcinoma reported in the world's literature.[3]

The first malignant form of acrosyringium was reported in 1956.[4] However, the term eccrine porocarcinoma was only introduced in 1969 for the malignant counterpart of eccrine poroma.[5]

Since then, apart from single case reports and very small series of cases, three relatively large series of 27 cases (Shaw et al[6]), 18 cases (Mehregan et al[7]) and 24 cases (Poiares-Baptista et al[8]) have been reported.

We present here a case of eccrine porocarcinoma and a review of the literature.

Case Report

A 58-year-old Chinese female presented with a raised purple plaque-like lesion, 7 cm in diameter on the lateral aspect of the right hip for 5 months (Fig. 1). Subsequently, she also noticed the presence of a lump, about 3 cm, in the ipsilateral groin region. On palpation, it was found to be an enlarged lymph node. A fine needle aspiration biopsy of this lymph node showed poorly differentiated malignant cells, consistent with metastatic melanoma. The provisional diagnosis was malignant melanoma with metastatic lymph node. A metastatic workup with a liver ultrasound and bone scan showed no gross involvement of the liver or bone.

A wide excision of 3 cm surgical margin of the pigmented lesion and superficial groin dissection was performed. The resultant defect was reconstructed with a split thickness skin auto-graft.

The gross description of the specimen showed that the margins of the lesion were sharply demarcated from the adjacent normal skin, with subcutaneous fat containing multiple enlarged lymph nodes. The largest lymphnodes measured 3 cm in diameter and contained tumour metastasis. Cut section of the lesion showed tumour mostly confined to the epidermis, which was thickened to 2 to 4 mm.

Microscopic description of the tissue showed histological features consistent with malignant skin adnexal tumour or porocarcinoma. It showed a predominance of intraepidermal growth (Borst-Jodassohn effect) (Fig. 2). Monomorphous cells were arranged in compact nests or masses confined principally within the epidermis and were associated with ducts and microcysts. These tumour cells were composed of small cuboidal to polygonal cells with uniform nuclei, prominent nucleoli and scantybasophilic cytoplasm. Many mitoses were present. A moderate degree of melanin pigmentation was featured. The larger masses of tumour showed centrally located foci of necrosis with pkynotic nuclei and eosinophilic granular debris.

* *Medical Officer*
** *Senior Registrar*
*** *Senior Consultant Plastic Surgeon*
Department of Plastic Surgery
Singapore General Hospital
Address for Reprints: Dr C L Foo, Department of Plastic Surgery, Singapore General Hospital, Outram Road, Singapore 169608.

Fig. 1. Close-up view of the raised purple plaque-like lesion.

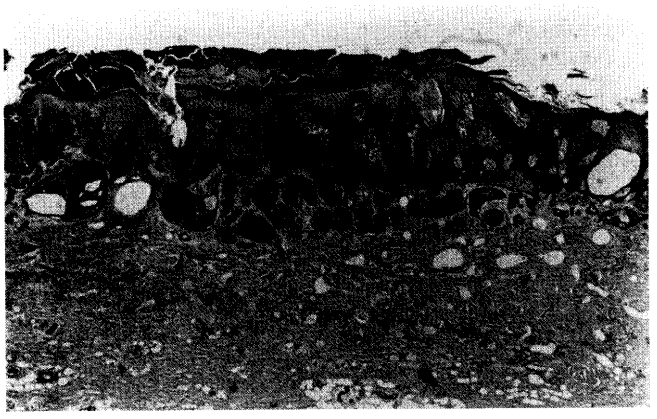

Fig. 2. Characteristic intraepidermal nests of tumour cells, thickening the epidermis to about 2 to 4 mm

This lesion showed focal invasion into the upper dermis, evidenced by focal breach of tumour beyond the confines of the basement membrane into the upper dermis as irregular nests and cords of tumour cells. There was associated intralymphatic spread where occasional tumour islands were seen within the dermal lymphatics. Nodalmetastasis was seen. Further adjuvant chemotherapy or radiotherapy was deemed unnecessary as there was a solitary lymph node involvement and there was no documented metastasis elsewhere. At six months follow-up, the patient showed no signs of recurrence. She will continue to be followed-up in the long term.

Discussion

Histology of Eccrine Sweat Glands

Skin appendages or adnexa include eccrine and apocrine sweat glands, hair follicles and sebaceous glands. There are two kinds of sweat glands, eccrine sweat glands and apocrine sweat glands.

Apocrine glands are found in the axillary, areolar, and anal region and are embedded in the subcutaneous tissues. Their ducts open into hair follicles and they produce a viscous secretion that is initially odourless but develops a distinctive odour after bacterial decomposition.

Eccrine (merocrine) sweat glands are simple, coiled tubular glands whose ducts open at the skin surface. They produce hypotonic sweat, which evaporates after its release on the surface of the skin, cooling the surface. These glands are distributed throughout the entire surface of the body except at mucocutaneous junction, but are highly concentrated on the palms, soles, axilla and forehead.

The ducts are in the dermis and epidermis. The intradermal portion of the duct is straight and is known as the syrinx. The intraepidermal portion of the eccrine duct, which spirals to its opening on the skin surface, is the acrosyringium. Tumours of the syrinx are called syringomas, and those of the acrosyringium are called poromas.

A wide spectrum of tumours may originate from acrosyringium, both benign and malignant. The benign forms are known as poromas and include variants of eccrine poroma, hydroacanthoma simplex.[9] The first malignant form was described by Smith and Coburn, naming them hydroacanthoma simplex "in-situ" and "invasive" hydroacanthoma simplex.

Clinical Features of Porocarcinoma

Porocarcinoma has been reported in patients between the ages of 19 and 94 years with the average age of 67 years.[10] There is no apparent sex predilection. The majority of the patients had noticed the primary lesion for 1 to 5 years but a variable period of 2 months to 50 years have been reported.[7,10] Known for its biological aggressiveness, the slow evolution supports the hypothesis formulated by many that majority of the porocarcinomas develop from the "malignant transformation" of a preexisting poroma.[11,12]

The majority of the porocarcinomas are situated on the lower extremities, accounting for approximately 55%. Twenty per cent occur on the trunk, 15% on the head and 10% on the upper limbs.[13] The tumour can appear in the form of a verrucous plaque or polypoid tumour that is often ulcerated, at least focally. Most reach diameters between 1 and 5 cm[7] and show evidence clinically of being malignant-asymmetrical, poorly circumscribed, and fixed to the subcutaneous tissues. The lesions are often red, but can also be light brown or flesh-coloured.[14] They are usually clinically diagnosed as pyogenic granuloma, squamous cell carcinoma, amelanotic melanoma or basal cell carcinoma.[6]

Porocarcinomas have two characteristic biologic features, which are invasion of lymphatics in 30% of cases and marked epidermotropism in 20% of cases.[15] Invasion of the lymphatic vessels may result in metastasis to the regional lymph nodes. Lymphangitis carcinomatosa associated with lymphoedema has been reported.[7] Epidermotropism explains the frequent cutaneous metastasis that is observed in patients. These cutaneous

lesions may be skin-coloured or reddish papules or nodules, which can appear distant from the original site. Less commonly, visceral metastases have been reported.[14]

Histopathology of Porocarcinoma

The histological characteristic of porocarcinoma is the presence of intraepidermal nests with round, deeply basophilic cells in acanthotic epidermis[9] (Fig. 3). The nuclei are large with hyperchromatic mitoses.[16] Abundant clear cytoplasm is seen. Characteristically, the cells are rich in glycogen, Periodic acid Schiff (PAS) positive,[16] and contain carcino-embryonic antigen (CEA), differentiating the tumour from squamous cell carcinoma and basal cell carcinoma.[17] Melanin pigmentation, just as in our patient has been reported.[18]

Many rounded oblong, or slit-like lumina similar to eccrine duct may be found in the epidermis. Necrosis en masse is not uncommon,[19] as in our patient.

The tumour may remain completely intraepidermal (malignant hydroacanthoma *in situ)* but is usually intradermally invasive, occasionally involving the subcutaneous fat.[7] Invasive porocarcinoma is characterised by downgrowths of broad anastomosing bands of epithelium with small cells united by intercellular bridges and absence of peripheral palisading.

Fig. 3. Higher magnification demonstrating characteristic monomorphous appearance of eccrine porocarcinoma. The cells are round and deeply basophilic, with large nuclei and hyperchromatic mitoses.

Porocarcinoma may invade the dermal lymphatics, spread within them, and finally reinvade the epidermis. This could explain the extensive dissemination of cutaneous metastasis within a circumscribed skin region and late recurrence of internal metastasis.[14] In the metastasising original nodule, dermal nests and lymphatic involvement are observed. Dermal infiltration as well as epidermotropic growth within the epidermis may be seen in metastatic deposits.[20] The neoplastic cells may have a pagetoid pattern within the epidermis where there is scatter of neoplastic cells throughout the entire thickness of the epidermis. This is also called the Paget's phenomenon and is seen in Paget's disease and in some cases of cancer en *cuirasse*.[19]

Only a few porocarcinomas have been studied by electron microscopy.[5,7,12] Tonofilaments and amounts of glycogen in neoplastic cells, cell membranes with complex interdigitating microvilli-like processes, and rare tight junctions have been described. Vacuoles within the neoplastic cells are a characteristic feature. Mature melanosomes have been detected within neoplastic cells and melanocytes have been identified.

Prognosis and Treatment

The prognosis of porocarcinoma is difficult to assess. Tumours that are present for only a few weeks can be accompanied by both regional and visceral metastases. On the other hand, some tumours can be present for up to 30 years without any metastasis.

A mortality rate cannot be determined with certainty.[14] According to available information, most patients with metastasis died despite treatment.[14] It has been estimated that carcinoma of the sweat glands without involvement of the lymph nodes usually have a poor prognosis, with a survival rate at 5 years of 67% and 10 years of 56%. With local regional metastasis (lymph node or cutaneous) or at a distance (lung, bone, etc), the survival rates were reduced to 29% and 9% at 5 years and 10 years, respectively.[21]

Because of its propensity to develop local recurrence, it has been recommended that wide excision of the primary tumour with histologic confirmation of tumour-free margins is the treatment of choice.[14] Prophylactic lymphadenectomy should be done if regional nodes are enlarged, as in our patient, or when a recurrent or poorly differentiated tumour with intralymphatic permeation is present.[14] However, it has been reported that, in spite of radical surgical excision, there is still a local neoplastic recurrence in 25% of patients with porocarcinoma.[6,13]

The benefits of radiotherapy and systemic chemotherapy are uncertain as metastatic porocarcinoma is

generally refractory to these modalities.[14]

In view of the high incidence of local recurrences and dissemination in the regional lymph nodes and skin, it has been suggested that radiotherapy should be used to complement surgical removal of the tumour so as to sterilise eventual residual tumours and lymphatic micrometastasis.[13] Radiotherapy may also have a place in the treatment of inoperable adnexal tumour or recurrences.** However, sweat gland carcinomas demonstrate a radioresistance in most cases, and doses of more than 70 Gy are considered necessary for a local control of the neoplasm.[21]

The exact indication and limits of chemotherapy in the treatment of malignant tumours of the sweat glands are not clear.[21] 5-fluorouracil, cyclophosphamide, doxorubicin, melfalan, vincristin, crisplatin and bleomicin have been used.[13] Regional hyperthermia used in combination with chemotherapy has been attempted.[13] Systemic chemotherapy and intraarterial infusion have been tried. Current literature show that there is a relative resistance of metastatic tumour of the sweat glands, but polychemotherapy appears to be more efficient with respect to monochemotherapy.[13]

Retinoids were shown to have some benefit in one patient, but failed in another. Combination of retinoid with interferon has been tried with a result of a lo-month remission in metastasis of lymph nodes, skin and retroperitoneum. A singular result showed a 3-year response of lymph nodes and bone metastasis of an estradiol receptor-positive eccrine adenocarcinoma to tamoxifen. Complete regression of multiple cutaneous metastasis was achieved with. perilesional interferon alfa in combination with interleukin.[13]

Conclusion

Porocarcinoma is a rare malignant tumour, arising from the intraepidermal ductal portion of the eccrine sweat gland. Porocarcinoma has a tendency to invade the lymphatics and to metastasise to the skin. The primary treatment is complete surgical excision. Local recurrences are common and metastatic porocarcinoma is generally resistant to radiation, chemotherapy and biotherapy. Survival rate is dependent on extent of spread on presentation and patients with metastases to lymph nodes, lungs or bones have grim prognosis. Therefore, early recognition and complete treatment by wide surgical excision using histological confirmation are the strategy to improve survival for the patients.

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