Eccrine Porocarcinoma

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ABSTRACT

Eccrine porocarcinoma is an extremely rare malignant cutaneous neoplasm. This tumour has an indolent behaviour and misleading clinical diagnosis often leading to a delay in definitive treatment. The definitive diagnosis is made by histopathology since clinical findings often vary. The tumour must be considered in the differential diagnosis of elderly patients presenting with long standing skin lesion with a recent history of accelerated growth rate. Because of its reported high rate of local recurrence, a wide excision of the tumour with clear resection margins is mandatory. Close long term follow up is required. We present herein two cases of eccrine porocarcinoma.

Keywords: Eccrine porocarcinoma, complete excision, sweat gland tumour

INTRODUCTION

Eccrine porocarcinoma (EPC) is an extremely rare malignant cutaneous neoplasm. The tumour arises from the sweat glands and accounts for 0.005% of all cutaneous epithelial neoplasms. [1] It was first described in 1963 by Pinkus and Mehregan. [2] Since then, only around 200 cases of this tumour have been reported in the literature.

This tumour typically exhibits slow growth over a long period of time but often experiences an accelerated growth phase. The tumour may arise *de novo* from the intraepithelial portion of the eccrine sweat gland or more commonly from a malignant transformation of a pre-existing eccrine poroma.[3] We report herein two cases of EPC.

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CASE REPORTS

Case 1: A 67-year-old man presented with a five-year history of an enlarging lesion on the supraumbilical aspect of his abdominal wall. The lesion had been rapidly growing over the previous two months. The patient had some discomfort in the lesion. Examination showed a 4 x 3cm erythematous and indurated growth, with central ulceration. It was mobile and not fixed to underlying structures. There was no inguinal lymphadenopathy. The preoperative diagnosis was squamous cell carcinoma.

The patient underwent excisional bi–

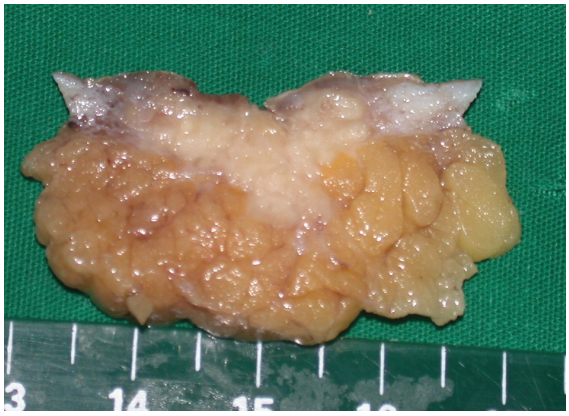

Fig. 1: The surface of lesion is ulcerated, with a diffuse grey white tumour extending 1.5 cm inferiorly.

opsy of the lesion. Macroscopically, the surface of the lesion was ulcerated, with a diffuse grey white tumour extending 1.5cm inferiorly (Figure 1). Histology showed an ulcerated tumour arising from the epidermis with nests and cords of cells extending diffusely into the dermis (Figure 2). Solid and cystic pattern was seen, with foci of squamous differentiation and clear cell change. The cells had atypical pleomorphic nuclei with increased mitotic activity. Pagetoid pattern of spread in the epidermis was seen. The stroma showed marked lymphocytic infiltrate. There was lymphovascular invasion. Margins of resection were free of tumour. The appearances were consistent with that of an eccrine porocarcinoma. The patient did not receive any adjuvant therapy and there was no evidence of recurrence after three years follow-up.

Case 2: A 75-year-old woman presented with an enlarging lesion on her right cheek. The lesion had been present for one year and had been rapidly growing over the previous two months. Recent discharge and malodour prompted her consultation visit. The patient had a circumscribed 3 x 2cm erythematous and exophytic growth that was mobile and not fixed to underlying structures. The pre-operative diagnosis was an infected pyogenic granuloma.

An excisional biopsy of the lesion was performed. Histology showed an ulcerated tumour of cuboidal cells extending from the epidermis as broad bands into the dermis. The nuclei of these cells were hyperchromatic and pleomorphic, and contained well-defined

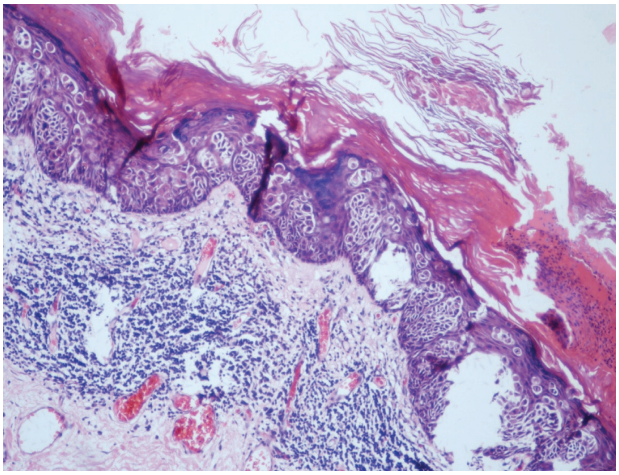

Fig. 2: Tumour arising from the epidermis with nests and cords of malignant cells expanding into the dermis. There is solid and cystic pattern of squamous differentiation with clear cell change. Mitotic activities are seen in the nuclei. Pagetoid pattern of spread in the epidermis and marked stromal lymphocytic infiltration is evident (H&E stain, x10).

nucleoli with many mitotic figures. The peripheral cells layer was haphazardly arranged. The cells on the surface showed clear cell change. There was marked lymphoplasmacytic infiltration at the base of the tumour. The appearances were consistent with that of an eccrine porocarcinoma. There was no vascular or perineural invasion and the lesion was completely excised. There had been no evidence of recurrence after five years follow up.

DISCUSSION

Our two case reports of EPC emphasised the major problems in the management of this particular type of cutaneous tumour. The tumour has an indolent behavior and misleading clinical diagnosis, which often leads to a delay in definitive treatment.

As seen in our cases, EPC most commonly occurs in elderly patients; with most cases reported in the fifth decade of life. [4, 5] Adults of either sex are affected, with a female predominance in most reported series.[4,5] The tumour has a propensity to arise on the lower limbs (44 to 62%), trunk (17 to 24%) and head and neck (18 to 19%). [5, 6] Other rare affected sites reported are penis [7] and nail fold. [8] Time to diagnosis is usually long, with up to 50 years reported, because some of these tumours could have arisen from pre-existing benign eccrine poroma.[5] Clinically the cutaneous lesion vary greatly in size, from less than 1 cm up to 10 cm in diameter.

Macroscopically, the lesions may present as solitary circumscribed nodules or plaques, pinkish or erythematous in appearance. The surface could be ulcerated. Many are asymptomatic, but they can be associated with bleeding, pain, pruritus and/or recent rapid growth rate. Mild local symptoms were often reported by patients. The clinical differential diagnoses of this lesion include seborrheic keratosis, pyogenic granuloma, verruca vulgaris, amelanotic melanoma, squamous cell carcinoma, basal cell carcinoma and skin metastasis of neoplasms. [4, 5]

The definitive diagnosis is made by histopathology since clinical findings often vary. These tumours originate from cells of the eccrine duct epithelium, usually in the region of the dermoepidermal junction. [9] The tumour is typically formed of cohesive basaloid epithelial cells. Neoplastic cells have a polygonal shape with clear cytoplasm, multi–ple nuclei or a large, hyperchromic and irregular nucleus. Ductal structures, Paget-like growth of atypical cells in the epidermis and mitotic figures are usually observed. The epidermis may show acanthosis as a result of numerous well-defined nests of intertwined tumour cells. [9] The histological diagnosis may be difficult to make. The abundance of glycogen granules in the cytoplasm of the tumour cells is detected by the periodic acid Shiff (PAS). Lack of keratinisation and clusters of ducts have been reported to be peculiar characteristics of EPC. [4, 7]

Other prognostic indicators observed histopathologically has been described. A high mitotic index of more than 14 mitotic cells per HPF, lymphovascular invasion and a tumour depth exceeding 7mm have been associated with a poor prognosis. [5] The tumour margin has also been shown to have a dramatic influence on local recurrence [5] tumours displaying an 'infiltrative' advancing margin has an increased predictive risk of local recurrence

compared to those tumours with "pushing" advancing margins. [5]

Our two cases of EPC showed no evidence of recurrence or metastasis after three and five years follow up respectively; despite lymphovascular invasion seen on histology in Case 1. The majority of reported series described this tumour with aggressive behaviour. [4, 5] Eccrine porocarcinoma seems to metastasize readily via the lymphatics in the dermis. [8] At presentation, regional lymph node metastases have been reported to be present in about 20% of cases. [5] Distant metastases have been seen in about 10% of patients in reported series.[10] The prognosis of this tumour is difficult to establish due to its rarity and the different follow-up periods reported in the literature. In the series report by Robson et al, [5] 17% of the patients experienced a local recurrence, 19% developed lymph node metastases and 11% experienced distant metastases or death on follow-up.

Due to the high rate of local recurrence, a wide excision of the primary tumour with histologically clear resection margins is mandatory. [6, 9] If clinically indicated, block dissection of regional lymph nodes have been advocated. [8] The role of adjuvant radiochemotherapy remains controversial. [11] There are few reports addressing the use of chemotherapy in the treatment of EPC without metastases. [12] No standard therapy protocol exists for metastatic EPC. Anecdotal reports have shown some benefits in the treatment of metastatic EPC with chemotherapy where there have been minor remissions and long-term stability of the disease. [13, 14] Much less is known about the role of radiation therapy in the treatment of ECP with metastasis. There have been few reports showing benefit. [15]

In conclusion, EPC is a very rare malignant cutaneous neoplasm. Because of its reported aggressive behavior, healthcare professionals should be aware of this disease. Eccrine porocarcinoma must be considered in the differential diagnosis of elderly patients who present with long standing skin lesion and a recent history of accelerated growth rate. Because of its reported high rate of local recurrence, a wide excision of the tumour with histologically clear resection margins is mandatory. Close long term follow up is required. The role of chemotherapy and radiotherapy in the adjuvant setting still remains unclear.

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