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ECCRINE POROCARCINOMA OF SCALP: A CASE REPORT OF AN UNUSUAL TUMOR AT AN UNUSUAL SITE

General Surgery

Jagadish B. Hedawoo — Associate Professor, Department of General surgery, Government Medical College Nagpur

Vipul Gupta — Junior Resident Department of General surgery, Government Medical College Nagpur Corresponding Author

ABSTRACT

Eccrine porocarcinoma is a rare malignant adnexal tumour of ductal portion of eccrine sweat gland. It commonly occurs in older age group, common site being lower extremities and rarely in scalp, face, ear, trunk and upper extremities. Local recurrence and metastasis to skin, lymph nodes, viscera and bone may occur. Surgery is the treatment modality of choice. Metastatic lesions can be treated with chemotherapy. We report an unusual case of eccrine porocarcinoma of scalp in a 85 years old female patient arising over right fronto-parietal region. Diagnosis is quite challenging by clinical examination alone, hence histopathology and immunohistochemistry form the mainstay for early diagnosis and treatment. Till date, only one case of eccrine porocarcinoma on fronto-parietal region has been described in literature akin to our case.

KEYWORDS

Eccrine porocarcinoma, scalp, fronto-parietal

Eccrine porocarcinoma is a rare malignant adnexal tumour which arises from intra-epidermal ductal portion of sweat gland (4). The first case was reported by Pinkus and Mehregan in 1963 after which few more cases have been documented (1). These rare tumours account for around 0.005% of all epithelial cutaneous tumours and have been termed as malignant hidroacanthoma simplex, malignant intra-epithelial eccrine poroma, eccrine poroepithelioma, malignant syringoacanthoma, dysplastic poroma and sweat gland carcinoma (1,4,6). It is the malignant counterpart of eccrine poroma, a common benign adnexal tumour. Eccrine porocarcinoma may arise denovo or due to malignant transformation of its long standing benign counterpart (1). Most common location of eccrine porocarcinoma is lower extremities. Other less common sites are scalp, face, ear, upper extremities, trunk etc (1, 2, 4). Though uncommon, local recurrence and lymph node metastasis both may occur in eccrine porocarcinoma. Wide local excision with negative marginal status is the key management and post operative chemotherapy may be necessary in case of lymph node metastasis (1, 6).

In this case report, we are presenting a rare case of eccrine porocarcinoma of scalp in a 85 year old female.

Case Report

An 85 years old female patient presented to us with a bosseleted mass of 10 years duration over right fronto-parietal region. It was firm, painless, freely mobile, tender, and bled on touch. There was no clinical evidence of any lymph node involvement or distant metastasis. CT scan showed no intracranial extension, no bony erosion and no calcification. Wedge biopsy suggested malignant skin adnexal tumour favouring sebaceous carcinoma. Wide local excision was done and preauricular lymph node sampling was done. Pericranium was free of tumour. The resultant defect was covered with split thickness skin graft. Cut section of tumour was greyish white, friable with areas of necrosis and cystic changes. Microscopic examination showed stratified squamous epithelium lined tissue with underlying neoplasm arranged in the form of lobules, islands, nests and groups with a characteristic infiltrative borders. Tumor cells are polyhedral to fossaform with variable cytoplasm, hyperchromatic nuclei, distinct nucleoli and indistinct cell borders. Mitosis is brisk (20/10 high power fields) with few atypical forms. Squamous differentiation and clear cell change is seen. Histopathologically margins were free of tumour infiltration and there was no evidence of lymph node metastasis. Diagnosis was confirmed with immunohistochemistry which showed tumour cells positive for p63, ck 5/6 with ki-67 proliferation index of 80%.

Figure 1 Gross examination showed a reddish yellow mass of size 11x9x4 cm with multinodular surface and foci of necrosis.

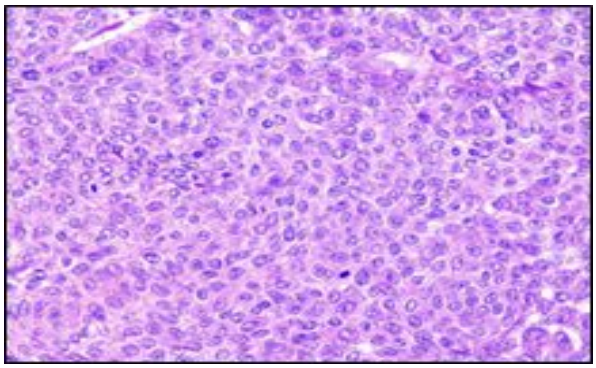
Discussion

Eccrine porocarcinomas are very rare malignant adnexal tumours arising from intra-epidermal ductal portion of eccrine sweat gland. Around 50% cases involve lower extremities (6). In few cases, it may also involve head and neck region, upper limbs, trunk and abdomen (4,6). The tumours have been found to commonly affect elderly patients of more than 60 years, though cases have also been reported in younger age group (2). Eccrine porocarcinoma may either arise de-novo or secondary to any long standing pre-existing lesions like eccrine poroma, nevus sebaceous, chronic lymphocytic leukaemia and actinic keratoses (3,6,9). The commonest presentation is reddish nodular cauliflower like growth or infiltrative verrucous plaque which often shows superficial ulceration & bleeding due to trivial trauma (3). Clinically lesions of the extremities mimic seborrheic keratoses, pyogenic granuloma, amelanotic melanoma, squamous cell carcinoma and basal cell carcinoma and verruca vulgaris and hence should be differentiated from them (3,10). Clinical features of eccrine porocarcinoma of scalp also mimic cylindroma, eccrine poroma, sebaceous adenoma, sebaceous carcinoma, pilar tumour and metastatic carcinoma (6).

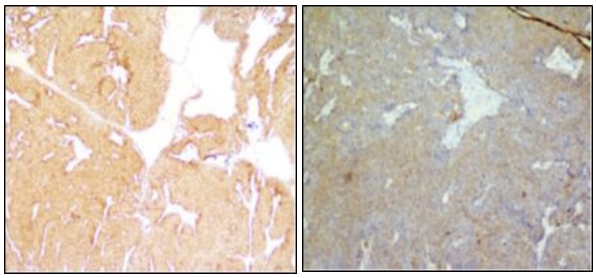
Matlaub, Cunningham, Yousif, Sanger & Romano (1988) (6) reported a case of ulcerative porocarcinoma of occipital region involving pericranium. Ritter, Graham, Amaker, Broaddus, Young (1999) (9) have reported one case of eccrine porocarcinoma in occipital region with intracranial extension. They mentioned 6 cases of scalp porocarcinoma as described in literature. Rana, Verma, Puri, Baliarsing (2005) (6) described a case of porocarcinoma in fronto-parietal region similar to this case.

Microscopically eccrine porocarcinoma shows intra-epidermal nests or cords of polygonal anaplastic cells which invade downwards into dermis and subcutaneous tissue. Epidermotropism and ulceration of epidermis are often seen in eccrine porocarcinomas. The tumour cells contain clear cytoplasm, large hyperchromatic nuclei and prominence of nucleoli. Frequent mitosis and necrosis are also seen (7). Most of the sweat gland carcinomas exhibit immunoreactivity for CK,

CEA and EMA (5). In our case diagnosis was confirmed with immunohistochemistry which showed positivity of tumour cells for p-63 and ck-5/6 with ki-67 proliferation index of 80%.



A



B C

Figure 2 Microscopy examination: Histopathology findings (A); Immunohistochemistry findings (B) p63 © ck-5/6

Treatment is done by wide local excision. If there is involvement of regional lymph nodes, regional lymph node dissection should be done. For covering such large skin defects plastic surgery assistance is needed. Metastatic lesions should be treated with chemotherapy (6). In the present case, after 6 months of follow up there is no local recurrence as well as no clinical evidence of metastasis.

Around 100 cases of porocarcinoma have been described in literature (6), of which around 10 have been of scalp porocarcinoma. There is only one case in literature of porocarcinoma on fronto-parietal region akin to our case.

Conclusion

The present case is being reported in view of its rarity. Although a rare diagnosis, it should be considered as a differential diagnosis while evaluating any exophytic tumour of scalp. Further studies are needed to get a better understanding about the lesion.

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