Eccrine Porocarcinoma of Scalp: a Rare Case Report

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ABSTRACT
Eccrine porocarcinoma is a rare malignant adnexal tumor of ductal portion of eccrine sweat gland. It occurs commonly in the lower extremities and rarely in scalp, face, ear, trunk and upper extremities. This survey presents a classic case of eccrine porocarcinoma of scalp in a 58 yr old male patient, presenting as cauliflower like growth over parietal aspect of scalp.

Keywords: Eccrine porocarcinomas, Scalp, India

Introduction
Eccrine porocarcinoma is a rare malignant adnexal tumor arising from intra-epidermal ductal portion of sweat gland (1). The first case was reported by Pinkus and Mwhregan in 1963 and after that few cases have been documented (2). These rare tumors account for 0.005% of all epithelial cutaneous tumors and have been termed as malignant hidroacanthoma simplex, malignant intra epithelial eccrineporoma, eccrine poroepithelioma, malignant syringoacanthoma, dysplastic poroma and sweat gland carcinoma (1-3). It is the malignant counterpart of eccrineporoma, a common benign adnexal tumor. Eccrine porocarcinoma may arise denovo or as a result of malignant transformation of long standing benign counterpart (2). Commonest location of eccrine porocarcinoma is lower extremities. Other uncommon 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 excision with negative marginal status is the key management and chemotherapy may be necessary (2,3).

In this case report we are presenting a rare case of eccrine porocarcinoma of scalp in a 58 year old male.

Case Report
A 58 yr old male patient presented with gradually
increasing painless lobulated exophytic mass at scalp over left parietal region (Fig. 1). It was (5×4×4) cm in dimension and not fixed with underlying structures. No lymphadenopathy was noted at cervical, pre-auricular and post auricular regions. CT scan of brain did not show any intracranial extension and no calcification was noted in the tumor. All other routine investigations were within normal limit. Wide surgical excision and primary closure of the scalp wound were done. Excised specimen was sent for histopathological evaluation. Multiple sections were examined from the tumor tissue. The sections showed well defined tumor lobules or nests of polygonal to cuboidal cells invading laterally and deep into dermis & subcutaneous tissue (Fig. 2, 3). Individual tumor cell had clear cytoplasm, large pleomorphic and hyper chromatic nuclei with prominent nucleoli. Necrosis and frequent mitosis were also found in the examined sections (Fig. 4). Histopathological examination confirmed the diagnosis of eccrine porocarcinoma. Margins were free from tumor involvement and the lesion involved >7mm deep in dermis. The patient received 6 cycles of chemotherapy and had no recurrence during follow up period of one year.
Discussion
Eccrine porocarcinomas, are very rare malignant adnexal tumors arising from intra-epidermal ductal portion of eccrine sweat gland. More than 50% cases involve lower extremities (3). On rare occasion, it may involve head and neck region, upper limbs, trunk and abdomen (1,3). The tumors commonly affect elderly patients of more than 60 years, though cases have been reported in younger age group as well (4). Eccrine porocarcinoma may arise as de-novo or secondary to any pre-existing lesions like eccrine poroma, nevus sebaceous, chronic lymphocytic leukemia and actinic keratoses (3,5,6). The commonest presentation is reddish nodular cauliflower like growth or infiltrative verrucous plaque lesion which frequently shows superficial ulceration & bleeding due to trivial trauma (5). In the present case, it was illustrated with a reddish-yellow, lobulated cauliflower like mass without superficial ulceration. Clinically lesions of the extremities should be differentiated from seborrheic keratoses, pyogenic granuloma, amelanotic melanoma, squamous cell carcinoma and basal cell carcinoma and verruca vulgaris (5,7). In their large series, Robson et al. found that specific clinical diagnosis were never correct (7). Clinical features of eccrine porocarcinoma from scalp also mimic cylindroma, eccrine poroma, sebaceous adenoma, sebaceous carcinoma, pilar tumor and metastatic carcinoma (3). In our case it was clinically diagnosed as benign adnexal tumor of scalp. Searching the English literature revealed that only few cases of porocarcinoma are involving scalp (3) which was similar to this survey. But involvement of occipital region was documented by Ritter et al. (6). 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 tumor cells contain clear cytoplasm, large hyperchromatic nuclei and prominence of nucleoli. Frequent mitosis and necrosis are also seen. Our case showed classical histological features of eccrine porocarcinoma including invasion into dermis, necrosis & increased rate of mitosis. Histopathological differential diagnosis includes metastatic adenocarcinoma, trabecular carcinoma and Merkel cell carcinoma (4). Metastatic adenocarcinomas often exhibit nests of tumor cells in glandular or acinar pattern in epidermis and dermis with hyperplasia of overlying squamous epithelium. Lack of such pattern and absence of any features of primary carcinoma helps to differentiate our case from metastatic adenocarcinoma (8). Trabecular carcinoma and Merkel cell carcinoma exhibit trabecular pattern with classical nuclear features of fine granular chromatin, nuclear moulding, fragmentation and abundant mitosis which were lacking in the present case. Occasionally the neoplastic cells of eccrineporoma may exhibit low grade cytological atypia but the lesions lack infiltrative growth pattern, necrosis and frequent mitosis (7). Cytoplasm of tumor cells in porocarcinoma show PAS reactive, diastase labile glycogen (1). Immunohistochemical features like positivity for CEA, EMA, CK-7 and negative staining for S-100, CK-20 help in diagnosis of difficult cases where histopathological findings are not conclusive (1,3,4). Treatment of eccrine porocarcinoma is wide local excision with histopathological confirmation of clear margin (1,3,6,7). In case of regional lymph node involvement, lymph node dissection should be considered. In contrast to radiotherapy, chemotherapy is suitable and effective for management of metastatic lesions (1,3). Regional lymph node metastasis has been reported in 20% cases and local recurrence in 25% cases (2,3,5,9). Distant metastasis is uncommon but reported in previous literatures (9). Prognosis of eccrine porocarcinoma varies on lymph vascular invasion, tumor margin status after resection, mitotic count (>14/HPF), depth of invasion in deeper tissue (>7mm) (7). All of the above features are poor prognostic factors. In our
case no nodal metastasis was found and tumor margins were free but depth of the tumor was >7mm and average mitosis was 12/HPF. Eccrine porocarcinoma of scalp is a rare malignant adnexal neoplasm. Local excision with negative margin is the cornerstone of management. Our case is a classical example of this rare malignancy and emphasizes inclusion of the lesion in the list of differential diagnosis during evaluation of any exophytic tumor of scalp.

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References