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Case Report

Eccrine Porocarcinoma: A Challenging Case Series

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Abstract: Eccrine porocarcinoma (EPC) is a rare malignant tumour of sweat glandorigin representing only 0.005-0.01% of epithelial cutaneous neoplasms. EPC is an unusual tumour to diagnose both clinically and histologically. Several reports describe aggressive behaviour. Due to its rarity, specific guidelines for management are not readily available. We report a series of four cases of EPC analysed in terms of clinical features, histology and aggressiveness of the tumour. The unusual highlights of this report are the rarer sites of occurrence of all the four tumours.
Keywords: Eccrine porocarcinoma (EPC), malignant tumour

INTRODUCTION

Adnexal tumours are epithelial tumours with differentiation towards theadnexal structures of skin, namely hair follicles, sebaceous, apocrineand eccrine glands. Eccrine porocarcinoma is a malignant tumour of sweat gland origin. It represents only 0.005-0.01% of epithelial cutaneous neoplasms. It is seen predominantly in elderly people, with a reported female predominance. The most common site is the lower limb followed bytrunk, head & neck and upper limbs. Among the reported series, majority describe aggressive behaviour. Due to its rarity, specific guidelines and recommendations for management are not readily available[12,1]. This study is a retrospective analysis of four cases of eccrine porocarcinoma diagnosed in our centre during a period of four years.

MATERIALS &METHODS

A retrospective audit of malignant cutaneous neoplasms during the period from 2010 to 2014 was done. It showed total of 133 cases out of which six were malignant appendageal tumours(0.045%). Eccrine porocarcinoma constituted four of these six cases, representing 0.037% of all cutaneousmalignancies. These four cases were analysed for their clinical presentation, histologic features and biologic aggressiveness.

RESULTS

All four patients were males. The age of presentation ranged from 51 to 90 years. The predominant site was head & neck with one case being on the anterior abdominal wall (hypogastrium and inguinal area).The size variedfrom 1-20 cm, the largest lesion measuring 20cm was seen on the anterior abdominal wall. The lesions in the head & neck were clinically diagnosed as Basal cell carcinoma or sebaceous cyst whereas the lesion in the lowerabdominal wall was called Squamous cell carcinoma. Following initialbiopsy, wide excision with adequate margins was performed (except inlesion on the external auditory canal). Histologically all tumours appeared as lobular and trabecular epidermal downgrowths growing into the dermis with pushing margins. Ductular formations as seen histologically or immunohistochemical staining for CEA& EMA were seen in all four cases. Tumour depth varied from 3-8mm. Mitosis varied from 10-70/10hpf. Ki-67 ranged from 1-6%. Lymphovascular emboli and tumour necrosis were present in one case each. Perineural invasion was absent in all four. Regional lymphnodes were positive in the case with lesion on the scalp, anterior abdominal wall and external auditory canal.

Table 1: Comparative features

Age	Sex	Site	Size(in cm)	Clinical diagnosis	Regional lymph nodes	Tumour depth in mm	Mitotic count/10 hpf	LVI	PNI	Tumour necrosis	Ductal differentiation by histology + IHC for EMA & CEA	Ki-67
83	M	Lower Abdominal wall	20x25	SCC	Present	5	70-80	A	A	P	Ducts seen	5%
51	M	Scalp	4x4	BCC	Absent	6	40-50	A	A	A	Ducts seen	1%
90	M	Scalp	2x2	Sebaceous cyst	Absent	8	10-20	P	A	A	Ducts seen	3%
69	M	External Auditory canal	1.5x1	BCC	Absent	3	10-20	A	A	A	Ducts seen	2%

Fig-1: Two pigmented and ulcerated plaques on the scalp posteriorly, 4x4 and 2x1 cm respectively, clinically diagnosed as Basal cell carcinoma

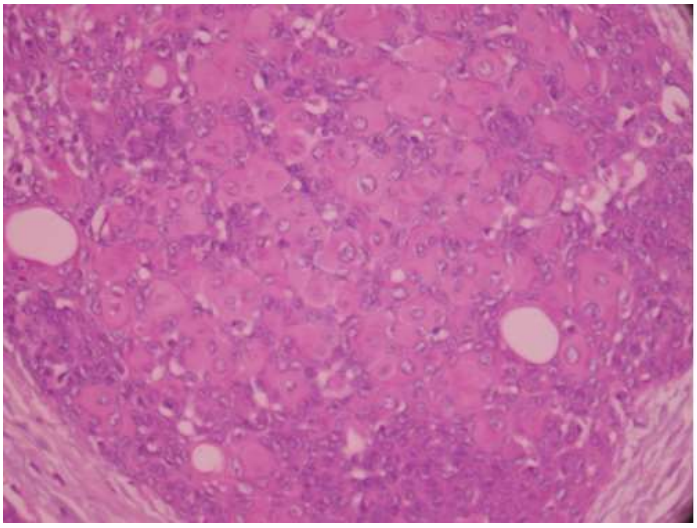

Fig-2: Skin with large tumour lobules growing into dermis and subcutis-H & E,40x

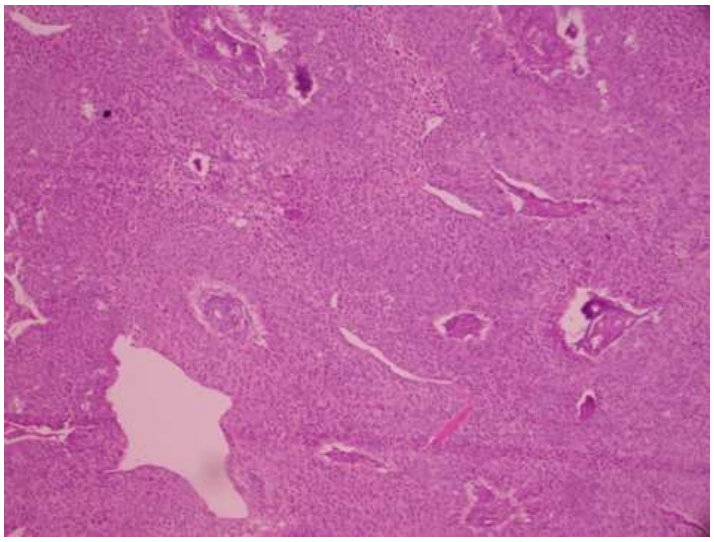

Fig-3: Sheet of proliferated monotonous appearing basaloid cells with scant fibrovascular tissue in between-H & E,400x

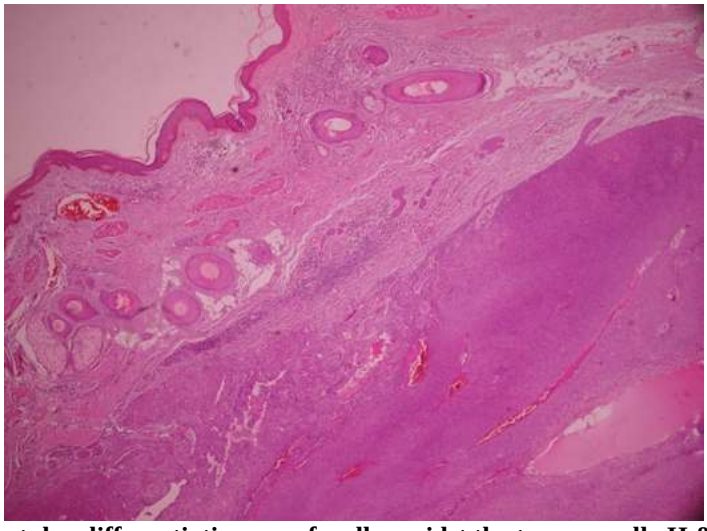

Fig-4: Ductular differentiation seen focally amidst the tumour cells-H & E, 400x

DISCUSSION

Majority of studies on adnexal neoplasms has been reported in the Western countries. Adnexal carcinomas can result from actinic damage asin Basal cell carcinoma to those that seem to those that seem to have little relationship with sun exposure. Eccrine porocarcinoma(EPC) is a malignanttumour related to the sweat gland duct with both intraepidermal and dermal components. The first reported case in 1963 was attributed to Pinkus and Mchregan. The term EPC was introduced in 1969 by Mishima & Morika[2, 3].

Approximately 300 cases have since been published in medical literature [3, 4]. Predominantly observed in elderly population, it usually arises denovo or rarely in a pre-existing eccrine poroma. The incidence in one large series was 0.004%. Fifty percent cases arise in the lower extremities, with next frequent sites being the trunk(24%) and head(24%) [2, 3].The rarity in our case series is that all the four cases were not on the lower extremity which is the usual site of presentation, but on uncommon sites like anterior abdominal wall, scalp and external auditory canal.The case on the anterior abdominal wall appeared to be an extensive lesion. There have been rare reports of similar lesions on the thorax[4, 5, 10,11].

It usually presents as a verrucousnodulo-ulcerative plaque resembling several of the other cutaneous neoplasms.The rarity and non-specificappearance of EPC makes an accurate clinical diagnosis unlikely and it is usually labelled as seborrheic keratosis, verruca vulgaris, squamous cell carcinoma(SCC),basal cell carcinoma(BCC) or amelanotic melanoma. In particular a small biopsy can be easily misdiagnosed.A long clinical historywith slow

progression is typical in EPC which suggests the possibility of malignant change in pre-existing eccrine poroma [6, 9].

In our cases the clinical diagnosis wasSCC,BCC or sebaceous cyst with EPC not being considered in the differential diagnosis. Poromas and Porocarcinomas usually lack melanocytes and melanin pigmentation within the lesion but rare incidence of pigmented porocarcinoma has been reported which can clinically and histopathologically be misdiagnosed as malignant melanoma [7, 8].

The histology of EPC suggests development from acrosyringium and is characterised by well demarcated nests and islands of basaloid cells, with cytoplasmic glycogen and irregular,mitotically active nuclei. Keratinsation is absent. Histologically ductal differentiation is characteristic,although poorly differentiated EPC may not show obvious duct formations. The ducts like lumina are formed intracellularly or by separation of neighbouring cells [6, 9].

Immunohistochemically, the poroid cells stain with antibodies to pan-cytokeratin. CEA and EMA are seen to be positive at the rim of the ductal lumen. Presence of scattered dendritic cells positive for both S-100 and HMB-45 are seen in both benign and malignant poromas[2,8].

All the fourcases in this series showed the classic histology with evidence of ductular differentiation seen without and with IHC Increased number of mitosis, lymphovascular invasion and a tumour depth of more than 7mm are associated with a relatively poor prognosis. There is limited knowledge about the genetic mutations implicated in adnexal neoplasms with the exception of multiple tumours which occur as apart of autosomal dominant syndromes. Genetic studies in EPC have shown mutation of p53 with loss of its suppressor function.There are no standard treatment protocols for EPC[4,3].

As a fairly high rate of local recurrence has been observed, wide surgical excision of the primary tumour with histologic clear margins is the treatment of choice. Adequate surgical resection achieves curative outcomes in 70-80%[4]. Therapeutic lymphadenectomy should be performed in cases of regional lymphadenopathy[5]. About 20% of EPC will recur locally and about 20% will metastasize.Carcinomas with eccrine differentiation have a known propensity to metastasize to skin.None of our cases had any evidence of metastatic disease.In general,adnexal carcinomas of small size, low histologic grade and whencompletely excised have an excellent prognosis[1]. Increased number of mitosis, lymphovascular invasion and a tumour depth of more than 7mm are associated with a relatively poor prognosis. The high rates of recurrence and metastasis warrants a close follow up and sometimesadjuvant therapy is indicated. Clinical trials using electroporation (electrochemotherapy) have shown good response rates in patients with primary or metastatic skin cancers[7]. Post operative radiotherapy is generally reserved for palliative care and tumour response is seen to be partial and inconsistent.

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