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Original Research Paper

General Surgery

A SURGEON'S SURPRISE TO VERRUCOUS GROWTH OVER CHEEK - CASE REPORT ON MALIGNANT ECCRINE POROMA

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ABSTRACT Malignant eccrine poroma is a rare malignancy of eccrine glands , whose diagnosis is an index of high suspicion and challenging to a clinician. A case of 67 year old female with swelling over left cheek since 20 years with no radiation exposure and no comorbidities , was investigated and wide local excision with 5mm margins from the tumor was performed , and specimen sent for biopsy - which revealed tumor cells arranged in lobules with peripheral palisading , ductal and squamous differentiation - suggestive of Porocarcinoma.Wide surgical excision is the mainstay of treatment, resulting in cure rates of 70–80% when the margins are clear. Some studies also reported good outcomes with Mohs micrographic surgery, and this technique could also be considered 1. Other treatments such as chemotherapy and radiotherapy have also been reported in the literature without clear standard guidelines.

KEYWORDS : ECCRINE POROMA ; BASAL CELL CARCINOMA ; MELANOMA

INTRODUCTION :

Eccrine porocarcinoma (EPC) is a rare malignant cutaneous tumor with high rates of extracutaneous spread. It was first described by Pinkus and Mehregan in 1963 [2] . It is a malignant tumor arising from intraepidermal eccrine ducts [2,3] . It may arise de novo [2,3] , but usually develops in a long standing eccrine poroma [9,7,10,15] . In some instances , malignant eccrine poroma manifests itself as nodule , plaque or ulcerated tumor [3,15,7,8,10] .

In 18% cases , tumor arises from preexisting benign eccrine poroma [3] . Incidence is 0.01 to 0.005% of all cutaneous tumors [5-8] . Females are affected more than males .

Here , we report a case of malignant eccrine poroma in a 67 year old female , managed at our institution.

CASE PRESENTATION -

A 67 year old female patient presented with swelling over left cheek , since 20 years - gradual onset , progressive in size, eventually formed a growth with blackish discoloration. No history of sudden increase in size of swelling. No history of Trauma / Radiation / Drug intake / Smoking . No history of loss of appetite or loss of weight . No family history of similar complaints . On Examination - 5x4 cm firm ,verrucous growth , non tender , non warm is noted on the left cheek area , 3 cm away from the left angle of mouth with hyperpigmented , crusted surface and restricted mobility. Speech and Mastication movements are normal. No deviation of mouth.

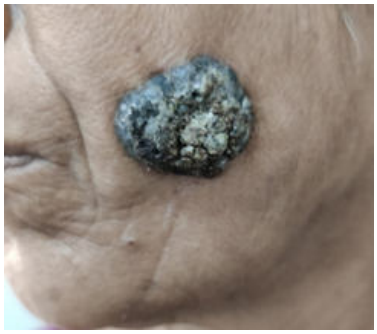

Figure 1 : Clinical presentation of the patient

CLINICAL COURSE -

BLOOD TESTS	Hb - 12.2 gm% WBC - 6,580/mm3 PLC - 2.66 lakh/mm3 Creatinine - 0,6 mg/dl RBS - 101 mg/dl Total Bilirubin - 0.67 mg/dl
USG ABDOMEN AND PELVIS	Liver normal size and echotexture Left kidney shows 1.6x1 cm anechoic cystic lesion(simple cyst)
2D ECHOCARDIOGRAPHY	EF - 60% Normal sized chambers and good LV systolic function
HISTOPATHOLOGY REPORT	Section from the growth shows ulcerated epidermis and tumor tissue , arising from the basal layer of epidermis and extending upto deep dermis. Tumor tissue is arranged in lobules and solid nests and anastomosing cords , with peripheral palisading with follicular structure, squamous differentiation and ductal differentiation with central necrosis in some tumor nests. Individual cells show moderate eosinophilic cytoplasm , round to oval vesicular nuclei with coarse chromatin. Brisk mitotic activity 3-5/10 hpf noted. Areas of sebaceous differentiation , cystic change , myxoid change and retraction clefts noted. Stroma shows desmoplasia , along with lymphoplasmacytic infiltrate. All resected margins , including deep resected margin are free from tumor.

DISCUSSION -

A malignant tumor arising from intra-epidermal eccrine ducts [2,3] ; 18% cases , tumor arises from preexisting benign eccrine poroma [4] . Incidence is 0.01 to 0.005% of all cutaneous tumors [6,7,8] . Among reported cases, the 8th decade of life is the ideal age of affection followed by the 7th decade (Table 1). Females are affected more than males . The lesion presents as endo

exophytic growth , often ulcerated , seen mostly on lower limbs (44%) , may attain large size and is frequently long standing.

Clinically, EPC appears as an asymptomatic erythematous or violaceous nodule or mass, although it can sometimes be ulcerative and painful [20]. Differential diagnoses include pyogenic granuloma, amelanotic melanoma, seborrheic keratosis, Bowen's disease, fibroma, verruca vulgaris, or metastatic adenocarcinoma.

AGE GROUP (in years)	NUMBER OF REPORTED CASES/%
0-20	0/0
21-30	3/5
31-40	0/0
41-50	6/11
51-60	9/16
61-70	12/21
71-80	18/32
81-90	8/14

Table 1 [21]: Age wise distribution of reported cases

Robson et al.[4] presented a review of the disease with the largest series of 69 patients, describing the clinicopathologic features of EPC. Histology is important to confirm the diagnosis of EPC.

Pathologically [4,9,10,11] , the tumor shows multiple connections to epidermis ; in-situ lesions are seen occasionally [4,12] . Tumor infiltrates dermis and subcutaneous tissue in nests and lobules , composed of relatively small cells that don't have basaloid appearance. Ductal differentiation is necessary for diagnosis to be made . Very often , comedo necrosis is present . In some cases , this tumor is localized , manifesting itself as nodule , plaque , ulcerated tumor [3,7,8,10,15] . Clear cell change , Squamous differentiation and rarely sarcomatoid differentiation is seen.

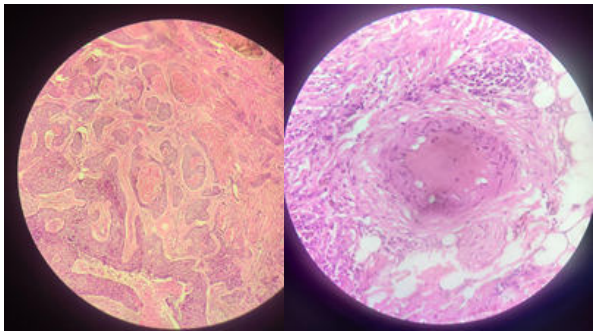

Figure 2 : Nest of tumor cells with central comedo necrosis.

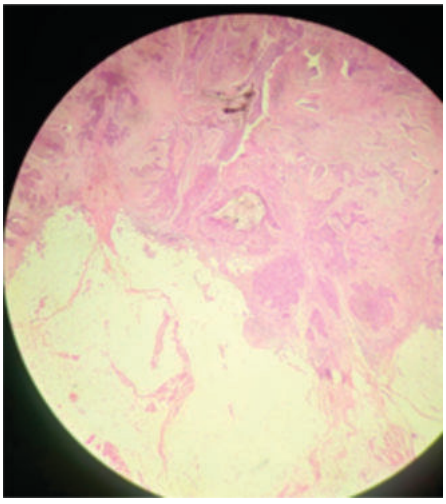

Figure 3 : Tumor cells infiltrating the subcutaneous (adipose) tissue

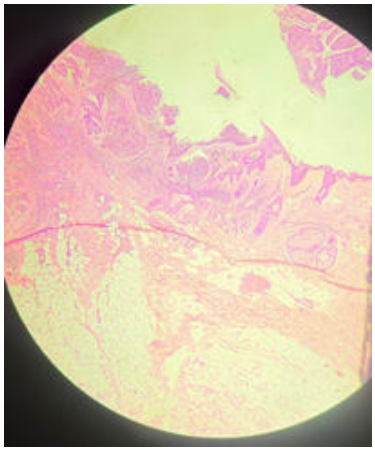

Figure 4 : Tumor tissue (above) infiltrating the normal epidermis (below)

Treatment should always be considered, as it is an aggressive disease with a high potential for morbidity and mortality. Many treatment options have been reported, with no clear standard. Wide surgical excision is the mainstay of treatment, resulting in cure rates of 70–80% when the margins are clear. Some studies also reported good outcomes with Mohs micrographic surgery, and this technique could also be considered [1].

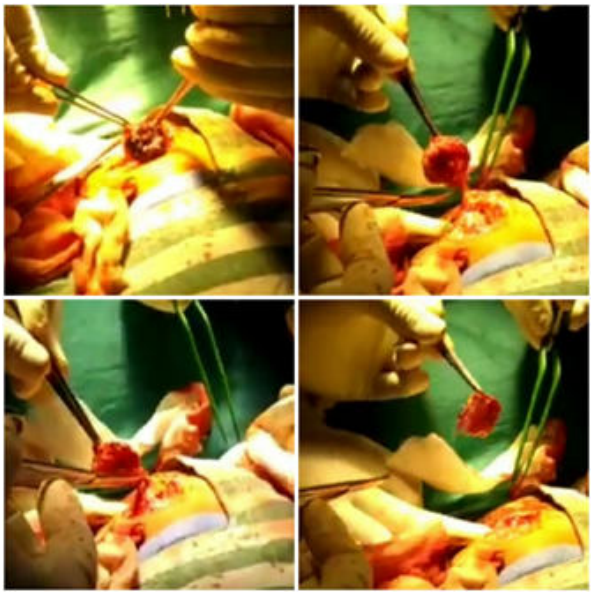

Figure 5 : Intra - operative pictures of wide local excision of the verrucous growth.

Given that porocarcinoma displays retraction artifacts and focal Ber EP4 positivity , distinction from basal cell carcinoma can be challenging with partial biopsy.

Poor prognosis is associated with large mitotic figures, lymphovascular invasion , tumor depth >7mm .

Local recurrence is seen in 17% cases, Regional lymph node metastasis is seen in 19 % cases with a mortality of 67% ; systemic metastasis is seen in 11 % cases ; Distant metastasis is rare [13] .

A case of cutaneous metastatic EPC treated with topical 5-FU and intra-arterial docetaxel has also been reported [17]. Plunkett et al. [16]showed some benefit using a single cytotoxic agent with docetaxel. More recently, responses have been reported with the use of epidermal growth factor receptor targeted

therapy and pembrolizumab [18,19].

This patient was treated by wide local excision with 5mm margins from the tumor , and specimen sent for biopsy - which revealed tumor cells arranged in lobules with peripheral palisading , ductal and squamous differentiation with brisk mitotic activity (3-5/10 hpf) ; All resected margins including deep resected margins were free from tumor.

CONCLUSION -

This case is being presented because of its rarity. It raises suspicion of malignant melanoma and basal cell carcinoma , owing to its site. On Wide local excision and biopsy , it was identified as Malignant eccrine poroma (or porocarcinoma) . Patient was followed up post operatively for 1 week and 6 months and had an uneventful course with no recurrence and no local growth .

Wide local excision with follow up are required ,in such cases of malignant eccrine poroma [14] .

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