

## Case Report

# A case of recurrent porocarcinoma of the scalp requiring scalp reconstruction

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## ABSTRACT

Eccrine porocarcinoma (EPC) is a rare malignant tumour, first reported by Pinkus and Mehregan in 1963. It can develop from the eccrine duct epithelial component or transform from an underlying eccrine poroma. The tumor usually presents as an exophytic growth and has a female predilection. Rarely it can occur in scalp, face, ear, genitalia and eye lids. Treatment modalities have included standard excision, Mohs micrographic surgery, chemotherapy, and radiation therapy. Therapy of choice is surgical excision with clear margins with 70-80% cure rates. Chemotherapy is of no known significance and radiation might help in selective cases. Majority of cases are managed with excision and closure of defects but in selective cases there may be need to reconstruct the scalp defects. The methods of reconstruction may be local flaps, free tissue transfer and partial/full thickness skin grafts. Regional flaps are very useful in patients requiring cover of large defects.

**Keywords:** Eccrine porocarcinoma, Scalp reconstruction, Trapezius myocutaneous flap

## INTRODUCTION

Eccrine porocarcinoma or malignant eccrine poroma, can develop from the eccrine duct epithelial component or transform from an underlying eccrine poroma. The tumor usually presents as an exophytic growth and has a female predilection. Majority of the cases are seen in middle or old age.

The most common locations are the lower extremities, trunk, head, and upper extremities. It is notoriously known for recurrence and regional lymph node metastasis. Scalp is a rare site with 20 cases of porocarcinoma been reported on the scalp so far, only two are involving the occipital region.<sup>1,2</sup>

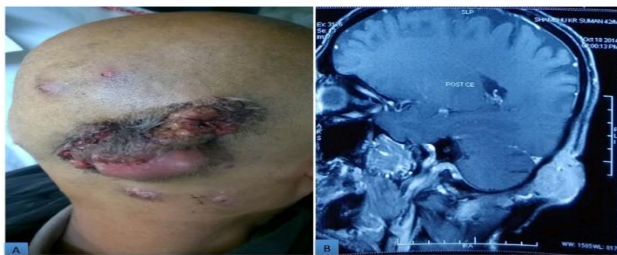
In this case report we present a case of a middle-aged man with recurrent porocarcinoma of the occipital region with involvement of the occipital bone, for which

craniotomy was done followed by myocutaneous flap reconstruction of the skull.

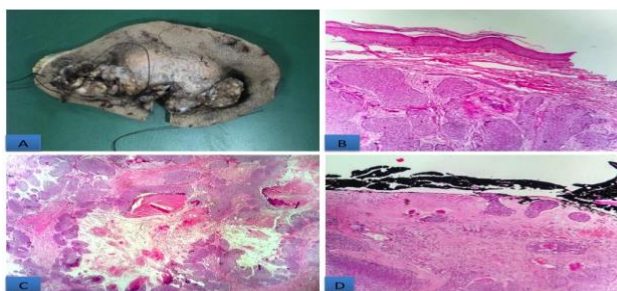
## CASE REPORT

A 46 years old male patient noticed a small painless swelling in the occipital region in the year 2008. The swelling gradually increased over the course of next 3 years. Resection of the swelling was done elsewhere in the year 2011 and the histopathology came as skin adnexal tumour. After one-year patient had recurrence at the same site. A second resection was done; the histopathology again came as skin adnexal tumour. Next time, the patient presented to our surgical OPD with a large bosselated ulceroproiferative mass in the occipital region (Figure 1A), measuring 7×4.3 cm without any regional lymphadenopathy. On evaluation, MRI (magnetic resonance imaging) reported as a lobulated sub-galeal mass in the right occipital region (Figure 1B). Wedge biopsy from the tumour margin revealed a lymphoplasmacytic stromal infiltration with no necrosis

or pleomorphism. The differentials were, benign adnexal tumour or an eccrine poroma. The patient was taken up for third surgery, wide local excision with 2 cm margin with peri-cranial shave. The gross specimen was 8×4.5×2.6cm ulcero-proliferative (Figure 2A) with microscopic examination revealing mild nuclear pleomorphism with atypical mitosis and necrosis. There was occasional duct like structures lined with cuticular cells. All the findings were suggestive of malignant tumour of adnexa, porocarcinoma Figure 2 (B and C). Only the deep margins taken as the peri cranial shave was positive for the malignant cells (Figure 2D). CECT head and neck done as part of metastatic work up, showed multiple enlarged lymph node in right posterior cervical chain, with largest node measuring 14 mm with no necrosis or calcification. FNAC from these nodes revealed malignant cells same as the primary tumour of the scalp. The patient underwent a second stage surgery i.e. craniotomy of the underlying occipital bone (Figure 3B) along with modified radical neck dissection. The raw area with exposed dura was covered with a trapezius myocutaneous flap in the same sitting Figure 3 (C and D). The histopathology of the neck nodes was negative for all the nodes. This was followed by radiotherapy in the form of 64 Gy/32#/6 weeks to primary site and bilateral neck by direct electron field and bilateral photon fields respectively. After a follow up of 4 years, there is no local recurrence or distant metastasis.



**Figure 1: (A) Large bosselated ulcero-proliferative mass in the occipital region and (B) MRI showing a lobulated sub-galeal mass in the right occipital region.**



**Figure 2: (A) the gross specimen was ulcero proliferative growth of size 8×4.5×2.6 cm, (B) revealing mild nuclear pleomorphism with atypical mitosis and necrosis, (C) presence of occasional duct like structures lined with cuticular cells were also seen, and (D) peri-cranial shave was positive for the malignant cells.**



**Figure 3: (A) Marking of trapezius myocutaneous flap, (B) exposed dura mater following occipital craniotomy, (C) posterior view of trapezius myocutaneous flap during follow up, and (D) lateral view of trapezius myocutaneous flap during follow up.**

## DISCUSSION

Eccrine porocarcinoma (EPC) is a rare malignant tumour, first reported by Pinkus and Mehregan in 1963. It is also known as malignant hidroacanthoma simplex, eccrine poroepithelioma, malignant poroepithelioma, dysplastic poroma, sweat gland carcinoma. Reported first by Mehregan used the term 'epidermotropic eccrine carcinoma'. Eccrine porocarcinoma was introduced by Mishima and Morioka in 1969. The etiopathogenesis EPC is yet to be eluded upon. This malignant tumor can arise denovo or some pre-existing lesions like "benign poroma" after a rather long latency period have been shown to transform to EPC. The other major risk factors include chronic sun exposure and states of immunocompromise, such as diabetes, transplantation and HIV.<sup>3</sup>

The most common presentation is nodule that may ulcerate, itch and may spontaneously bleed. The most common site of origin is lower extremities followed by trunk, head and neck and upper extremities. Rarely it can occur in scalp, face, ear, genitalia and eye lids. Spread to regional lymph nodes occurs in a significant number of cases. The definitive way to make the diagnosis is histopathological confirmation<sup>2</sup>. Histopathological diagnosis is based on nests of characteristic small poromatous basaloid epithelial cells with foci of ductal differentiation in the epidermis. These nests can reach deep into the dermis. Areas with clear cell, ductal components, and squamous cell differentiation may be found in the dermis. More than two-thirds of cases have matured eccrine ducts with cuticle with eosinophilic lumen. The cells of EPC are positive for p16, CEA, CK5/6, EMA and pan cytokeratin. They are negative for CK20 and S-100.<sup>4</sup>

Porocarcinoma of the scalp, frequently involves the occipital followed by frontal and parietal areas. Involvement of the underlying bone is very rare.<sup>3</sup> Treatment modalities have included standard excision, Mohs micrographic surgery, chemotherapy, and radiation therapy. Therapy of choice is surgical excision with clear margins with 70-80% cure rates.<sup>2</sup> Chemotherapy is of no known significance and radiation might help in selective cases. Due to paucity of literature no standardised treatment regimens are available. Lymph node exploration is only advised if involvement is proven or in patients with large tumor size (more than 5 cm). The use of adjuvant local radiotherapy to reduce local recurrence in cases with inadequate margins. Even though the response rate remains questionable, chemotherapy and radiotherapy are advised in metastatic lesions and cases with extra capsular extensions.<sup>5</sup>

There are two main goals to be considered while doing Surgical excision i.e. margin negativity and cosmesis. The various methods to reconstruct the scalp defects are, local flaps, free tissue transfer and partial/full thickness skin grafts. Regional flaps are very useful in patients needing large tissue but not for patients with impaired wound healing, radiation therapy history and palliative care setting.<sup>6</sup> In our case since the shaving from the occipital bone showed involvement, occipital bone was excised exposing the dura. Thus, necessitating the need for reconstruction. This was done by a rotational trapezius myocutaneous flap, which showed excellent cosmetic outcome on long term follow up.

## CONCLUSION

The EPC remains a challenge, primarily because of its rarity, morphologic peculiarity and similarity to another carcinoma. Thus, guidelines and strong recommendations are difficult to formulate and hence, are not available. To standardise the protocols publications from multicentric groups are needed. This will facilitate the ease of making clinical decisions when dealing this intriguing neoplasia.

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