Journal of the Egyptian National Cancer Institute (2016) **28**, 195–198

Cairo University

Journal of the Egyptian National Cancer Institute

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Case Report

Porocarcinoma scalp with high risk features treated with surgery and adjuvant radiotherapy: A case report and review of literature

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Received 5 April 2016; revised 15 May 2016; accepted 24 May 2016
Available online 11 June 2016

KEYWORDS

Porocarcinoma;
Scalp;
Radiotherapy;
Post operative

Abstract Eccrine porocarcinoma is a rare malignant sweat gland tumor arising from the intra dermal part of the gland and accounts for only 0.005% of all epithelial cutaneous tumors. Commonly involved site includes extremities and face. Scalp is a rare site for porocarcinoma with less than 20 reported cases so far. Wide local excision with clear margins remains the treatment of choice. Review of literature revealed a local recurrence rate of 37.5% and a nodal involvement risk of 20%. Porocarcinoma of the scalp is peculiar in that the primary tumor may be large at presentation, making surgery with adequate margins difficult. Adjuvant radiotherapy must be considered in a case to case basis due to the high local recurrence rates compared to other sites of porocarcinoma and should be given to all patients with close margins and extra capsular extension.

Introduction

Eccrine porocarcinoma is a rare malignant adnexal tumor arising from the intra dermal part of the sweat gland accounting for 0.005% of all epithelial cutaneous tumors. It is the malignant counterpart of common benign adnexal tumor (eccrine poroma) and is also termed as malignant hidroacanthoma simplex/eccrine poroepithelioma/malignant syringoacanthoma/dysplastic poroma. Eccrine porocarcinoma may arise de novo or as the malignant transformation of long standing benign poroma. Commonly involved site is lower extremity, though it has also been reported to arise from upper extremities, scalp and face also. Local recurrences and lymph node metastasis are seen in 20% of cases. Wide local excision with clear margins is the treatment of choice. The role of adjuvant radiotherapy has not been well defined. Scalp is a rare site for porocarcinoma with less than 20 reported cases. Here we present a case of porocarcinoma of the scalp treated with adjuvant radiotherapy and a literature review of cases of scalp porocarcinoma to evaluate the appropriate treatment in such cases.

Case report

A 42 year old male, without co-morbidities, presented with recurrent ulcer over the occipital region of scalp since 2011. Local examination revealed a 5 * 4 * 3 cm fungating ulcer pre-

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Peer review under responsibility of The National Cancer Institute, Cairo University.

http://dx.doi.org/10.1016/j.jnci.2016.05.004

sent over the right side of occiput. Examination of neck and systemic examination were normal. Magnetic resonance imaging (MRI) of brain showed a well marginated lesion in subgaleal soft tissue of the right occipital region with preserved occipital bone (Fig. 1). The lesion was hypo intense in T1 and T2 sequences and showed contrast enhancement. There was no intracranial extension or regional lymph nodal enlargement in MRI. Biopsy of the lesion was suggestive of eccrine poroma.

He underwent wide local excision with 1 cm margin all around. Post-operative histopathology revealed a tumor of size 4.5 * 3.5 * 1.5 cm and was suggestive of eccrine porocarcinoma with involved deep resected margin. The tumor cells showed nuclear pleomorphism, calcification, necrosis and occasional duct like structure lined by cuticular cells (Fig. 2). The photomicrograph showing patient was re-evaluated with contrast enhanced computed tomography which showed plaque like soft tissue thickening in the occipital region with thinning of occipital bone. There were multiple enlarged lymph nodes in the right posterior cervical region (largest 1.4 cm in short axis diameter). Fine needle aspiration cytology from cervical lymph node showed metastatic deposits from an adnexal tumor. The patient again underwent wide local excision with 1 cm margin all around with posterior neck node dissection. Histopathologically, it showed porocarcinoma in resected specimen with metastasis to cervical lymph nodes with extra capsular extension. The tumor cells showed mild nuclear pleomorphism with foci of calcification and necrosis. Atypical mitosis was also seen. The deep resected margin was also involved by tumor.

In view of presence of positive margin and presence of extra capsular extension in the nodal area, the patient was planned for adjuvant radiotherapy. Radiotherapy to the occipital region was planned with 12 MeV electrons and to neck by photons to a dose of 64 gray in conventional fractionation till. The patient developed acute grade II mucositis, grade II dysphagia and grade I dermatitis during treatment. Twelve months post treatment, the patient remains free of disease in local and nodal sites. The patient had chronic grade II xerostomia at the last follow-up.

Discussion

Eccrine porocarcinoma are extremely rare malignant tumors arising from the intra dermal part of sweat gland. It was first described by Pinkus and Mehregan in 1963 [1]. Mishisma and Moriko later introduced the term "eccrine porocarcinoma" in 1969 [2]. Porocarcinoma occurs equally in both sexes and predominantly in elderly. It presents as an ulcerated nodule or plaque, in lower extremities (50%), trunk (24%) or head and neck (24%). Prognosis is variable with wide local excision showing curative results. Lymph node metastasis and distant metastasis are associated with poorer prognosis. There are many reports that this cancer may arise from eccrine poroma and in our patient also it arose from a pre-existing poroma. Bone involvement is rare in this tumor and in our case also bone was free.

Microscopic appearance usually shows atypical tumor cells arranged in cords and lobules which may involve both dermis and epidermis. Tumor cell shows nuclear atypia with frequent mitosis and necrosis. These findings were evident in our patient. Robson et al. reported few histopathological factors that are predictive for poorer clinical outcome and death.[3] These are (1) presence of more than 14 mitosis per high power field (2) lymphovascular invasion (3) tumor depth >7 mm (4) infiltrating margins.

Wide excision or Mohs micrographic surgery is the treatment of choice. Surgery alone has shown curative results in 70% of cases although a recurrence rate of 20% is seen. Regional lymph nodes should be assessed as porocarcinoma has shown propensity to invade dermal lymphatics which cause lymph nodal disease in about 20% of the cases.[4] In our patient wide local excision was done with lymph node dissection in view of lymph node involvement.

A review of cases of porocarcinoma of the scalp in which the clinical details were available is summarized in Table 1 [5–13]. Clinical summary of 10 patients could be retrieved. The median age of diagnosis was 52 years. 40% of the cases arose from a pre-existing poroma. The median tumor size was 5 cm (range 0.4–8 cm). Bone involvement was not seen

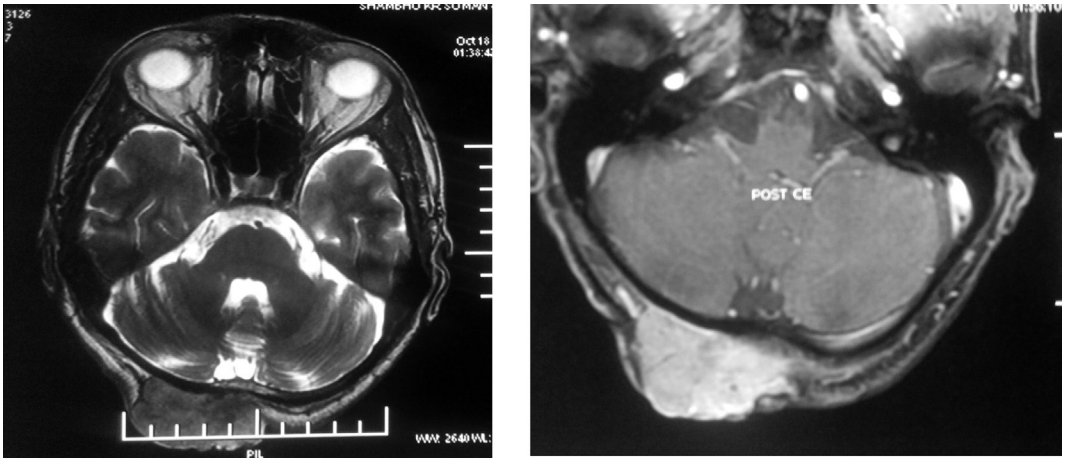

Figure 1 Magnetic resonance imaging of the patient showing a well marginated contrast enhancing lesion in the right occipital region.