J Nepal Health Res Counc 2024 Oct.-Dec.;22 (65): 796-798

Rare presentation of Eccrine Porocarcinoma

Piyush Giri,[1] Shrinkhala Pokhrel,[1] Pratishtha Pandey,[1] Karuna Khadka,[1] Sushma Kunwar,[1] Laxman Banstola,[2] Narendra Vikram Gurung[1]

[1]Department of Surgery, Western Regional Hospital, Pokhara Academy of Health Sciences,

[2]Department of Pathology, Western Regional Hospital, Pokhara Academy of Health Sciences.

ABSTRACT

Eccrine porocarcinoma is a tumor of the sweat gland that mostly develops from an existing benign eccrine poroma. It is a rare cutaneous malignancy with a high risk of metastasis and recurrence. Known to affect the elderly population in general with no gender discrepancy, it presents as an ulcerative mass commonly in the trunk or head/neck region. Definitive diagnosis is made by histopathological findings of peculiar pleomorphic cells with nuclear hyperchromasia and mitotic activity, surrounded by ductal lumen. The standard treatment is complete surgical excision with appropriate surgical margins; adequate work-ups to look for metastasis and timely follow ups for chances of recurrence. Here we report a case of an 82-year-old patient who presented at our department with a long-standing exophytic lesion in the head that rapidly started growing in the past few weeks. After excisional biopsy and histopathology, the diagnosis of Eccrine porocarcinoma was made.

Keywords: Cutaneous; eccrine porocarcinoma; malignancy; rare.

INTRODUCTION

Eccrine porocarcinoma (EPC) is a rare malignant cutaneous adnexal tumor representing 0.005% of skin neoplasm which commonly arises from long standing benign Eccrine poroma and was first described by Pinkus and Mehregan in 1963 as epidermotropic Eccrine carcinoma later called "Eccrine porocarcinoma" in 1969.[1-3] Usually affects 61.5 to 73 years old ranging from 20 to 100 years and usually originates from lower portion of intraepidermal ductal portion of eccrine sweat gland and from pre-existing benign poroma.[1,2] Its histology resembles that of cutaneous squamous cell, hence the tendency to under or misdiagnose.[4]

CASE REPORT

An 82-Year-old male presented to the surgery department of our hospital with mass over the frontal scalp region for 51 years. It was more or less constant in size till about a few weeks back, when it started to rapidly grow in size. It was a single, pinkish, round, exophytic mass of size 3cmX3cm with regular well-defined borders and a narrow base, multilobulated with smooth, shiny surface. No active discharge or bleeding was seen (Figure 1). Patient was otherwise healthy. CT scan with contrast showed avidly enhancing soft tissue density lesions in the left frontal scalp with few areas of calcification. Full thickness wide local excision was done with 1 cm margin from site of induration with depth up to loose areolar tissue where the tumor was not adherent to the underlying bone. Rotational flap was done to cover the defect (Figure 2). Postoperative period was uneventful. Histological diagnosis of Eccrine Porocarcinoma was made. No growth seen in the follow up period up to 1 year done in every 3 months (Figure 3).

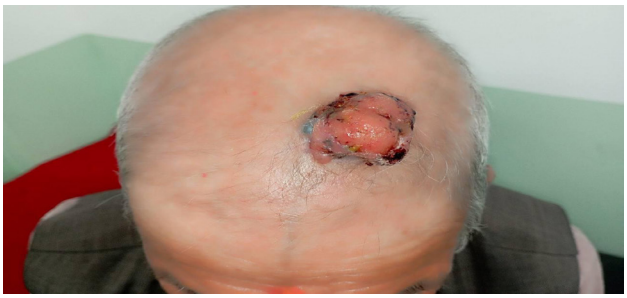

Figure 1. Preoperative findings of the patient with Eccrine porocarcinoma of the frontal scalp

Correspondence: Dr. Piyush Giri, Department of Surgery Western Regional Hospital, Pokhara Academy of Health Sciences, Pokhara, Gandaki, Nepal. Email: dr.piyushgiri@gmail.com, Phone: +9779745372953

Figure 2: Immediate postoperative finding after wide local excision and rotational flap.

Figure 3. Superior view of the frontal scalp; healed wound and no apparent growth at 12-month follow-up period.

DISCUSSION

EPC mostly occurs in lower limbs (33%) followed by head and neck.[5,6] The diagnosis of eccrine porocarcinoma is based on clinical evaluation, histopathological examination, and immunohistochemical studies. Clinical appearance of the tumor has been described as a reddish or brown nodular lesion with some being ulcerative. Important differential diagnoses include basal cell carcinoma, amelanotic melanoma and verrucae vulgaris. Histology of tumor is characterized by basaloid pleomorphic cells with nuclear hyperchromasia and mitotic activity, surrounded by mature ductal lumen, necrosis and is PAS positive and glycogen-rich.[5] Based on marginal patterns, the histological findings of EPC lesions have been classified into three variants- "infiltrative", "pushing" and "pagetoid". The infiltrative variant is characterized by malignant clusters infiltrating the dermis and/or hypodermis, pushing form exhibits a clear lower dermal limit and pagetoid lesion is characterized by intraepidermal spreading of neoplastic cells, with probability of relapse in descending order is the pushing, infiltrative and pagetoid variant.[3] On immunohistochemistry study, both EPC and benign poroma express CEA and EMA, helping distinguish it from other tumors.[4] It may demonstrate a local, horizontal (radical) intraepidermal growth pattern and a vertical dermal-subcutaneous growth pattern in addition to regional and distant metastases. Salih et al. reported 31% metastasis with most common being nearby lymph node (58.5%), followed by lungs (12.8%).[6]

The treatment is wide local excision with at least 0.5 cm to 3 cm of margin of the primary lesion along with the involved lymph nodes which ensures minimum loss of healthy tissue while effectively removing the involved margins.[3] Other modes of tumor removal described by Brown et al. are electro fulguration, electro cautery, simple excision, radiation and amputation, however, Aaribi et al. states at cost of high local recurrence.[1,7]

Additional treatment modalities such as radiation therapy can be used in order to reduce the risk of local recurrence after surgical excision and systemic chemotherapy such as cisplatin-based regimens has benefits in case of advanced or metastatic disease. The study for efficacy of this treatment however is limited. Aaribi et al. suggest the use of Docetaxel as a second line treatment for metastatic carcinoma as well as in cases of platinum resistant or refractory, and 5-FU before Moh's micrographic surgery.[7] Meanwhile Miyamoto et al. highlights the emergence of immunotherapy to target cancer cells selectively with pembrolizumab. Although remission for a 16 months period after the administration of pembrolizumab in a

chemo radiation resistant case, immunotherapy in some cases has shown as an up-and-coming treatment modality, the disestablishment of its large- scale clinical trials and studies due to its rarity, questions its efficacy.[4]

Regular follow up and surveillance allows for timely detection of disease progression and facilitates instantaneous initiation of additional therapies as necessary as interval from excision of the primary lesion to regional metastasis ranges from 1 month to 6 months.[8]

Accurate prediction of outcomes of EPC is challenging and which depends on degree of spread to surrounding tissues, especially distant metastasis as the prognosis is comparatively poor.[4] However; early diagnosis and surgical treatment are good prognostic factors. High mitotic index, lymph vascular invasion and a tumor depth more than 7mm are factors that may be associated with a poor prognosis.[9] In an analysis of 563 EPC cases, Scampa et al. reported 5-year overall survival (OS) as 74.8%. [10] Similarly Le et al. reported a 3-year overall survival and regional recurrence rate as 70.3% and 19.0%.[8] As for lymph node metastasis, Robson et al. found the 1-year and 3-year overall survival of 88.9% and 39.5% respectively while Song et al. mentioned a high mortality rate of 67% with distant metastasis rate of 11-12%. [9]

CONCLUSIONS

Eccrine porocarcinoma is a rare and aggressive cutaneous carcinoma originating from the eccrine sweat glands. Early diagnosis and prompt treatment are crucial in managing this malignancy effectively. Surgical excision remains the primary treatment; along with advancements in targeted therapies and immunotherapy offer potential new avenues for improved outcomes. Close follow-up is crucial to check for recurrence and possible metastasis.

CONFLICT OF INTEREST

The authors declare no conflict of interest.

REFERENCES

1. Brown CW Jr, Dy LC. Eccrine porocarcinoma. Dermatol Ther. 2008 Nov-Dec;21(6):433-8. doi: 10.1111/j.1529-8019.2008.00243.x. PMID: 19076620.

2. Huet P, Dandurand M, Pignodel C, Guillot B. Metastasizing eccrine porocarcinoma: report of a case and review of the literature. J Am Acad Dermatol. 1996 Nov;35(5 Pt 2):860-4. doi: 10.1016/s0190-9622(96)90105-x. PMID: 8912607.

3. Fionda B, Di Stefani A, Lancellotta V, Gentileschi S, Caretto AA, Casà C, et al. The role of postoperative radiotherapy in eccrine porocarcinoma: a multidisciplinary systematic review. Eur Rev Med Pharmacol Sci. 2022 Mar;26(5):1695-1700. doi: 10.26355/eurrev_202203_28238. PMID: 35302218.

4. Miyamoto K, Yanagi T, Maeda T, Ujiie H. Diagnosis and Management of Porocarcinoma. Cancers (Basel). 2022 Oct 25;14(21):5232. doi: 10.3390/cancers14215232. PMID: 36358649; PMCID: PMC9659280.

5. Kim HJ, Kim A, Moon KC, Seo SH, Kim IH, Kim A, Baek YS. Eccrine Porocarcinoma: A Multicenter Retrospective Study with Review of the Literatures Reported in Korea. Ann Dermatol. 2020 Jun;32(3):223-229. doi: 10.5021/ad.2020.32.3.223. Epub 2020 Apr 24. PMID: 33911741; PMCID: PMC7992626.

6. Salih AM, Kakamad FH, Essa RA, Rauf GM, S A M, H M S,. Porocarcinoma: A systematic review of literature with a single case report. Int J Surg Case Rep. 2017;30:13-16. doi: 10.1016/j.ijscr.2016.10.051. Epub 2016 Nov 18. PMID: 27898349; PMCID: PMC5128823.

7. Aaribi I, Mohtaram A, Ben Ameur El Youbi M, Kharmoum J, El Kabous M, et al. Successful management of metastatic eccrine porocarcinoma. Case Rep Oncol Med. 2013;2013:282536. doi: 10.1155/2013/282536. Epub 2013 Oct 26. PMID: 24282649; PMCID: PMC3825131.

8. Le NS, Janik S, Liu DT, Grasl S, Faisal M, Pammer J, et al. Eccrine porocarcinoma of the head and neck: Meta-analysis of 120 cases. Head Neck. 2020 Sep;42(9):2644-2659. doi: 10.1002/hed.26178. Epub 2020 Apr 21. PMID: 32314845.

9. Belin E, Ezzedine K, Stanislas S, Lalanne N, Beylot-Barry M, Taieb A, et al. Factors in the surgical management of primary eccrine porocarcinoma: prognostic histological factors can guide the surgical procedure. Br J Dermatol. 2011 Nov;165(5):985-9. doi: 10.1111/j.1365-2133.2011.10486.x. Epub 2011 Sep 20. PMID: 21711331.

10. Scampa M, Merat R, Kalbermatten DF, Oranges CM. Head and Neck Porocarcinoma: SEER Analysis of Epidemiology and Survival. J Clin Med. 2022 Apr 14;11(8):2185. doi: 10.3390/jcm11082185. PMID: 35456278; PMCID: PMC9024621.