



## Case Report

## Eosinophilic variant of eccrine porocarcinoma of the scalp: Case report and review of the literature

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## ARTICLE INFO

## Article history:

Received 19 December 2016

Received in revised form 13 June 2017

Accepted 13 June 2017

## Keywords:

Porocarcinoma

scalp

eosinophilic variant

## ABSTRACT

Porocarcinoma is a rare malignant neoplasm of the acrosyringium with metastatic potential that most commonly presents on the acral skin in older adults (mean age = 72 years). We present the case of a 43-year-old woman who developed a rapidly growing de novo porocarcinoma on the scalp with an unusual oncocyctic appearance. The tumor consisted of benign eccrine poroma that arose from the epidermis and broad pushing borders with minimal cytological atypia but ample eosinophilic cytoplasm with numerous mitotic figures. Although some tumors may appear deceptively bland, the histologic recognition of pushing/infiltrative borders and mitotic figures are helpful to make the appropriate diagnosis of carcinoma. This lesion was treated with Mohs micrographic surgery and the patient remained free of recurrence after more than 2 years. It is important to recognize the eosinophilic variants of eccrine porocarcinoma because it can histologically mimic a squamous cell carcinoma.

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## Introduction

Porocarcinoma is a rare malignant tumor of the acrosyringium that was first described by Pinkus and Mehregan (1963). Porocarcinoma has a reported true incidence as low as 0.004% of skin biopsy specimens (Mehregan et al., 1983) or as high as 3.5% of malignant skin tumors (Urso, 2013). To date, approximately 600 cases have been reported in the literature as single cases and relatively small case series (Urso, 2013). Notable for its aggressive potential, 19% of patients with porocarcinoma developed lymph node metastases, 11% developed distant metastasis or died, and 17% recurred locally in a large series of cases (Robson et al., 2001). Metastatic disease is often fatal because tumors are generally resistant to chemotherapy and radiation (Brown and Dy, 2008). The primary lesions present as verrucous or polypoid nodules or plaques, often with spontaneous bleeding and ulceration, and arise either from a preexisting eccrine poroma or de novo. The most common locations where lesions appear are the lower leg and trunk (68%) and less commonly on the head (18%) and upper limbs (11%; Robson et al., 2001).

Histologically, porocarcinoma is characterized by anastomosing cords and lobules of poroid cells, which are distinctly smaller than

epidermal keratinocytes. The tumor can be either entirely limited within the epidermis (in situ) or extend into the dermis with either a broad pushing border or infiltrative invasive strands (Robson et al., 2001). To support a diagnosis, tumors must have ductular structures, which can be intra- or extracellular. If these structures are not obviously visible with routine histology, ducts can be highlighted by epithelial membrane or carcinoembryonic antigen immunohistochemistry. In contrast to the bland monomorphic round clear cells that are observed with eccrine poroma, porocarcinoma cells may have large, pleomorphic, and vesicular nuclei and often display increased mitoses. They may be associated with adjacent poroma in approximately 11% of cases (Robson et al., 2001). Squamous cell carcinoma (SCC) is the most similar form of carcinoma histologically but distinguishes by its lack of ductular structures. Some researchers believe that the actual incidence of porocarcinoma is artificially low due to routine misdiagnoses of SCC (Urso, 2013). Histologic staining for cytokeratin 19, nestin, and cytokeratin 7 can be helpful to distinguish between porocarcinoma and SCC (Mahalingam et al., 2012).

The primary treatment for patients with porocarcinoma has traditionally been wide excision. Although sentinel lymph node biopsy (SLN) and Mohs surgery have been reported, there is no consensus on optimal treatment. Due to the small number of cases, the clinical utility of Mohs surgery and SLN biopsy are unknown. Here, we report on a case of porocarcinoma that displayed several rare features including a rapidly growing de novo lesion on the

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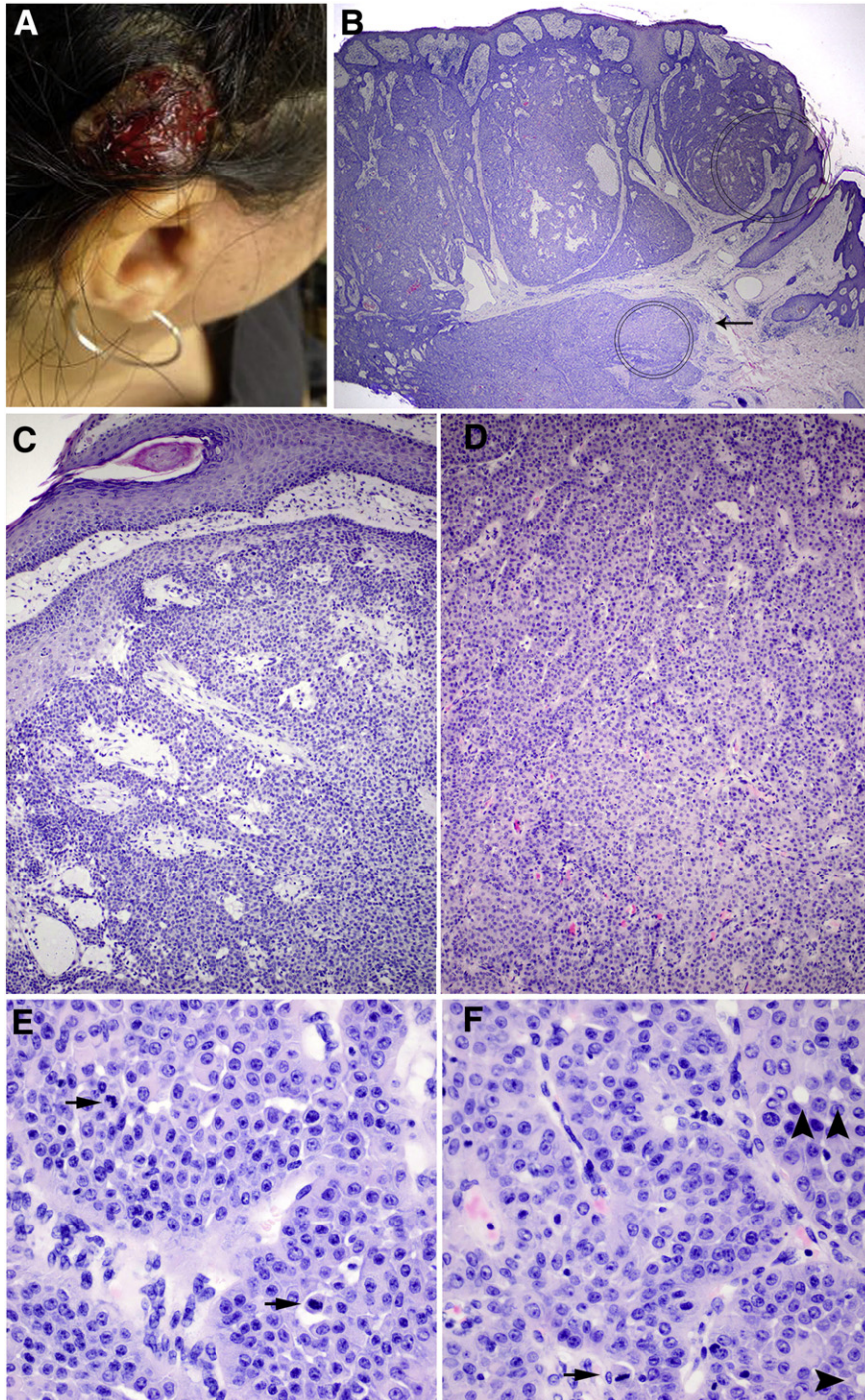
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scalp of a 43-year-old woman with a previously underrecognized oncocytic differentiation. The patient remained free of recurrence after treatment with Mohs micrographic surgery, which supports its use in the management of patients with porocarcinoma.

### Case

A 43-year-old woman presented with a painful, 2.3 cm by 2 cm, firm, ulcerated nodule on the right temporal scalp that had been

growing rapidly over the previous 10 months (Fig. 1A). The results of a physical examination were negative for cervical or occipital lymphadenopathy. The patient denied any constitutional symptoms (e.g., fever, night sweats, malaise, and fatigue) and was only prescribed levothyroxine for hypothyroidism. She had a tubal ligation and breast reduction 5 years prior. The patient reported no previous personal history of malignancy. Her family history of malignancy was positive only for her father, who was a smoker and had been diagnosed with laryngeal cancer.



**Fig. 1.** Porocarcinoma of the scalp. A) Ulcerated tumor on the right parietal scalp. B) Lobular tumor composed of monomorphic basophilic cells that originate from the epidermis with pushing border (arrow). C) Higher magnification (large circle in B) showed benign-appearing intraepidermal poroma. D) In contrast, the deep dermal nodule (small circle in B) showed a solid growth pattern. E) High-power magnification revealed cells with eosinophilic cytoplasm, slight nuclear pleomorphism, vesicular nuclei, prominent nucleoli, and mitoses. F) Numerous ductal structures were identified.



A diagnostic shave biopsy specimen that was analyzed histopathologically revealed an ulcerated large lobular tumor that was composed of well-defined, monomorphic nodules emanating from the epidermis and a lower tumor edge with primarily smooth, rounded borders. Focally, a pushing and infiltrative border was observed (Fig. 1B). A closer examination showed a typical poroma with small bland keratinocytes from within the epidermis (Fig. 1C). In the area of the pushing border, the growth pattern was solid and cells exhibited eosinophilia (Fig. 1D). A high-power magnification of the specimen revealed tumor cells with ample eosinophilic cytoplasm and brisk mitotic activity, which is consistent with a rapid clinical growth. There were slight nuclear pleomorphism, vesicular nuclei, prominent nucleoli, and five to six mitoses per 10 40X-fields (Fig. 1E). Numerous ductal structures were identified and the cytoplasm appeared more eosinophilic and granular than expected (Fig. 1F). No lymphovascular invasion was identified as the tumor's depth of invasion was measured at 6 mm. Mohs surgery was performed and required two fresh frozen stages to achieve negative margins. The defect was closed in a linear fashion. The patient had no signs of local or distant recurrence at all subsequent visits to date (at 3, 6, 12, 18, and 24 months).

## Discussion

Our case represents a rare clinical presentation that occurred on the scalp of a relatively young patient with an unexpected histopathologic finding of cytoplasmic eosinophilia. To date, only 16 cases of porocarcinoma have been reported on the scalp (Ekmekci and Lebe, 2013; Grimme et al., 1999; Kim et al., 2012; Kose et al., 2006; Matloub et al., 1988; Permi et al., 2011; Puttick et al., 1986; Ritter et al., 1999; Shaw et al., 1982; Sigal et al., 1997; Tarkhan and Domingo, 1985; Urso et al., 2001; Vessels et al., 2011). Of these 16 cases, three of the porocarcinomas metastasized, two demonstrated a recurrent local invasion, and one had an intracranial extension.

Due to its rarity and a lack of prospective studies, there is little consensus with regard to an optimal treatment for patients with porocarcinoma. In the largest case series to date (69 cases), investigators defined histologic features that are associated with poor prognostic value, which include more than 14 mitoses per high power field, lymphovascular invasion, invasive depth of >7 mm, and infiltrative borders that necessitate a "wide excision with close attention to surgical margins by the surgical pathologist" (Robson et al., 2001). However, in a retrospective study of 24 cases by Belin et al. (2011), a frequent recurrence of infiltrative and pagetoid tumors were observed despite a wide excision. Extrapolating from other recurrence prone tumors such as dermatofibrosarcoma protuberans, the researchers advocate for modified Mohs micrographic surgery to treat patients with tumors with an infiltrative or pagetoid histology while also advocating for a simple excision with negative margins for tumors with pushing borders (Belin et al., 2011).

Our case represents the seventeenth reported case using Mohs micrographic surgery to treat a patient with porocarcinoma. To date, all cases treated with Mohs surgery achieved negative margins without local recurrence (Cowden et al., 2006; D'Ambrosia et al., 2004; Johr et al., 2003; McMichael and Gay, 1999; Snow and Reizner, 1992; Vleugels et al., 2012; Wildemore et al., 2004; Wittenberg et al., 1999); however, one patient died of distant metastatic disease 2 years after surgery, presumably due to micrometastases at the time of surgery (Vleugels et al., 2012). On the basis of these reported cases, Mohs micrographic surgery has a 94% cure rate. As a malignant cutaneous neoplasm on the scalp, porocarcinoma meets the appropriate use criteria for treatment with Mohs micrographic surgery. In addition to its high cure rate, the tissue-sparing aspect of Mohs on the scalp requires less substantial reconstruction to close larger defects.

The salient histological features of porocarcinoma can include anastomosing cords and lobules of poroid cells within the epidermis. An important clue is a broad pushing border or infiltrative, invasive strands that can clue the pathologist to examine the tumor more closely for mitoses, cytological atypia that includes nuclear pleomorphism, and ductular structures. Although eosinophilic cytoplasm has been reported (Robson et al., 2001), it is a rare histological variant that should be recognized. This feature may mimic SCC; however, a ductal differentiation and the presence of intraepithelial poroma should aid in differentiating porocarcinoma from SCC.

## Epilogue

Jane's leadership style has been intentionally unconventional. When I joined her department in 2009, I thought she would be the usual chair of an academic department (i.e., manage the unit like a company's CEO). Soon after, I learned that her style of management was nurturing and almost motherly, which I found to be not only refreshing but very effective. She supported faculty members to pursue what they were genuinely interested in, akin to a caring mother who wanted the best for her children. In my case, she helped me pursue my interest in noncoding ribonucleic acids in melanoma progression and especially focus my work to benefit patient care. I appreciate how tirelessly she supported me to overcome obstacles to establish molecular diagnostics in our department. For all of us involved with teaching and mentoring students and residents, Jane has left us with a unique model of mentorship.

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