Large Cerebriform Eccrine Porocarcinoma: A Case Report

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Abstract
Porocarcinoma is a rare, slow-growing malignant neoplasm that may arise de novo or evolve from a pre-existing benign eccrine poroma. This lesion typically affects females over the age of 60 and represents about 0.005% of all cutaneous tumors. Porocarcinoma can be difficult to recognize. We present the case of a 46-year-old male with an unusually large, exophytic, cerebriform mass requiring multiple biopsies prior to a diagnosis of porocarcinoma.

Introduction
Porocarcinoma is a rare, malignant tumor that develops from the intraepidermal ductal portion of the eccrine sweat gland. It first described by Pinkus and Mehregan in 1963 as an epidermotropic eccrine carcinoma.1 With fewer than 250 cases reported worldwide in 2009, this condition accounts for about 0.005% of all cutaneous tumors.2

Affected individuals are typically elderly people over the age of 60, with a female predominance.2-4 Lesions primarily appear on the lower extremities, though they can occur anywhere else on the body including the trunk, head, upper limbs, and neck.5,6 Local and distant metastasis occurs in 10% to 20% of cases and often leads to poor prognostic outcomes, with a mortality rate of up to 80%.5 A lack of specific clinical findings and the presence of benign poroid features make this condition difficult to diagnose. In the presence of a benign poroma, clinicians should always evaluate the likelihood of the lesion undergoing a malignant transformation in order to minimize the morbidity and mortality associated with this neoplasm.

Case Summary
A 46-year-old male presented with a large, slow-growing, non-painful, exophytic mass on the anterior aspect of his left thigh that had been present for more than 19 years. It had started as a small, 5 mm red papule and remained relatively small until about five years prior, when it began expanding in size and leaking clear fluid. Despite its unusual transformation, the lesion remained untreated due to the patient not having any health insurance.

Upon physical exam, the tumor was nodular, ulcerated, and cerebriform in appearance, measuring 7 cm x 6 cm (Figure 1). The tumor had restricted mobility in all directions, and its borders were poorly demarcated, with a bulk of the lesion buried deep in the dermis. General examination revealed no apparent signs of lymph-node involvement, and there were no other significant physical findings associated with this lesion.

A biopsy was performed with a generously wide shave of the ulcerating mass. However, despite the size of the sample provided, the lesion displayed mostly poroid cells with uniform nuclear features consistent with a poroid neoplasm (Figures 2-3). Due to the extensive involvement of the lesion and the lack of a firm diagnosis for a malignant finding, a second biopsy was performed with deeper samplings utilizing a punch instrument. Review of the deeper sections of the second biopsy displayed a more infiltrative pattern with nuclear enlargement and mitotic activity (Figures 4-5). Even though the cytomorphology of the overall sample was uniform and lacked anaplastic features, there was enough evidence of cytological atypia and stromal infiltration at the base for it to be classified as a porocarcinoma (Figure 6).

Due to the unusually large size of the lesion and its poor prognostic features, the patient was referred to a general surgeon and an oncologist for further evaluation and treatment. The general surgeon performed a wide excision of the lesion followed by a left femoral lymphadenectomy of two nodes. The lesion was removed in its entirety, with the margins narrowly cleared on the deep surface by 3 mm. Both nodes removed from the left leg were negative for tumor.
As of this report, the surgical defect has been left open to heal by secondary intention with negative-pressure wound therapy in preparation for split-thickness skin grafting. Several radiologic and laboratory studies have also been performed and will be followed closely by the oncologist for any recurrence or metastasis.

Discussion

Establishing a solid diagnosis for porocarcinoma can be challenging. Unlike most malignant neoplasms, porocarcinomas have no specific clinical features and can vary in size, shape, and appearance. Tumors may vary greatly in size, ranging from less than 1 cm to 10 cm. Due to the condition’s rarity and nonspecific appearance, a clinical diagnosis may never be accurate without proper histologic correlation, and it can easily be misdiagnosed as a squamous cell carcinoma, basal cell carcinoma, seborrheic keratosis, or metastatic adenocarcinoma.2,3

Although porocarcinomas have no specific clinical features to identify, the sudden transformation of any lesion, becoming nodular, infiltrative, ulcerated, or polypoid, is often indicative of an underlying malignant process. Since porocarcinomas have been known to develop both de novo and from benign poromas, diagnosis of this condition can take many years depending on when the lesion underwent malignant transformation. As a result, it is common to find porocarcinomas arising from adjacent poromas histologically. The pathogenesis and neoplastic behavior of this condition explains why the biopsy samples taken from our patient displayed the cytologic uniformity of a benign process rather than a malignant one.

The classic histopathologic features of porocarcinomas typically involve intra-epidermal and dermal nests with features of cellular atypia and increased mitotic activity. These tumor masses form clearly demarcated nests of polygonal cells with hyperchromatic irregular nuclei, prominent nucleoli, and scant eosinophilic cytoplasm. Immunochemical staining techniques may be used to aid in the diagnosis of porocarcinoma, but results can vary depending on the histologic presentation of the lesion. Two of the main markers for this condition include epithelial membrane antigen (EMA) positivity and carcinoembryonic antigen (CEA) negativity. However, CEA staining for the evaluation of porocarcinoma can be misleading because it displays positive markers in tumors containing well-formed ducts, which are often found in significant portions of the lesion.

Many histologic features of porocarcinoma have proved helpful in assessing prognosis. The presence of lymphovascular invasion, a tumoral depth greater than 7 mm, and a mitotic index of more than 14 mitotic cells often indicate a worse prognosis compared to cases without these particular findings. Local and regional lymph-node metastases are observed in approximately 20% of patients, and distant metastases to the viscera and bone arise in 10% of patients with this condition. Prognoses of patients with metastatic porocarcinomas are usually poor, with a mortality rate of up to 80%. Due to the rarity of this condition and the subsequent lack of opportunities to evaluate new treatment options, surgical excision of the primary lesion continues to be the treatment of choice, with a cure rate of up to 80% and a recurrence rate of less than 20%. Any evidence of local or distant metastatic involvement would warrant regional lymph-node dissection in addition to close follow-up with an oncologist. Postoperative radiation or chemotherapeutic agents have been shown to improve patient outcomes and reduce the likelihood of recurrence. Mohs micrographic surgery is another viable treatment option and has been shown to produce moderate success without recurrence at two years to four years postoperative follow-up. However, if patients have multiple metastatic lesions or are poor surgical candidates, chemotherapeutic agents such as tamoxifen and docetaxel can provide effective, systemic alternatives to surgery.

Conclusion

Definitive diagnosis of porocarcinoma requires the collaboration of expert pathologic and oncologic studies. The case presented here shows that clinical findings are just as important as histological findings in diagnosing a suspected porocarcinoma. Upon receiving a diagnosis of benign poroma, every clinician should reevaluate the lesion in terms of its likelihood to undergo a malignant transformation. Early intervention can help minimize the morbidity and mortality of this potentially dangerous neoplasm.

References