American Journal of Otolaryngology and Head and Neck Surgery

Case Report
Published: 12 Mar, 2021

Porocarcinoma with Extramammary Paget Disease: Case Presentation and Review of the Literature

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Introduction

Eccrine Porocarcinoma is a rare primary skin tumor accounting for 0.005% of malignant epithelial neoplasms, most often presenting in the elderly population after the sixth decade of life [1]. Extramammary Paget's disease is a rare dermatologic condition that frequently presents in areas where sweat glands are abundant, notably, male and female genitalia, perineal, and axillary regions. Here we present a case encompassing extra-mammary Paget's disease arising from an underlying porocarcinoma. The concurrent presentation of both of these entities in a single area is rare with less than five reported cases dating from 1963 to current day.

Case Presentation

In May 2020, an 86-year-old woman presented with a long-standing history of a persistent scaly plaque-like lesion on her left cheek. The initial clinical diagnosis was suspicious of squamous cell carcinoma arising from the skin. Punch biopsies were performed and sent for microscopic examination which demonstrated a poorly differentiated malignant epithelial neoplasm involving the dermis, comprised of invasive nests of malignant cells with tumor necrosis and numerous mitosis however definite keratinization is lacking (Figure 1).

While most of the lesion demonstrated a malignant neoplasm in the dermis, the overlying epidermis demonstrated malignant cells with a pagetoid appearance (Figure 2). The malignant cells with pale cytoplasm in the dermis were positive for immunostains p16 (Figure 3), EMA pancytokeratin, and AE1/AE3 (Figure 3). The pagetoid cells in the epidermis were positive for immunostain CK7 (Figure 5). The tumor cells are negative for SOX-10, Mart-1, and S-100 ruling out melanoma. Additionally, synaptophysin and CEA were negative. The H&E and immunohistochemistry were most consistent with porocarcinoma.

Operatively, a wide excision with lymph node dissection was undertaken. The specimen was dissected to reveal fixation to the parotid gland, involving both the superficial and deep portions of the gland. Due to longstanding inflammatory changes from the cancer, the usual anatomic planes were not identifiable. The specimen was eventually resected in an en bloc fashion including the tumor, masseter muscle, and a portion of the parotid gland. Adjacent lymph nodes from the periparotid region and deep parotid were isolated and dissected out.

The skin and subcutaneous tissue excised from this individual demonstrated a 5.5 cm necrotic mass invading into the soft tissue and muscle to a depth of 2.1 cm and all resection margins were negative for carcinoma. The left periparotid lymph node was involved by metastatic porocarcinoma measuring 25 millimeters in greatest dimension with extracapsular extension. The intraparotid lymph node was positive with a focus measuring 3 millimeters.

Discussion

Pathology

Sweat glands are appendages of the integumentary system. There are two main categories including, eccrine and apocrine sweat glands. Eccrine glands are present throughout the human body most abundantly on the palms and soles. They are specialized units that provide the body with a well-established mechanism for thermoregulation and homeostatic control *via* a process of evaporative heat loss [2]. Porocarcinoma is the most frequent member of the malignant sweat gland neoplasms [3]. The lower extremities are the most commonly involved location for these

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Received Date: *09 Feb 2021*
Accepted Date: *09 Mar 2021*
Published Date: *12 Mar 2021*

Citation:
Ahmed Z, Shawn Liu S, Harrison Howard J, Bodie BF, Phung T, Knowles KJ. Porocarcinoma with Extramammary Paget Disease: Case Presentation and Review of the Literature. Am J Otolaryngol Head Neck Surg. 2021; 4(2): 1124.

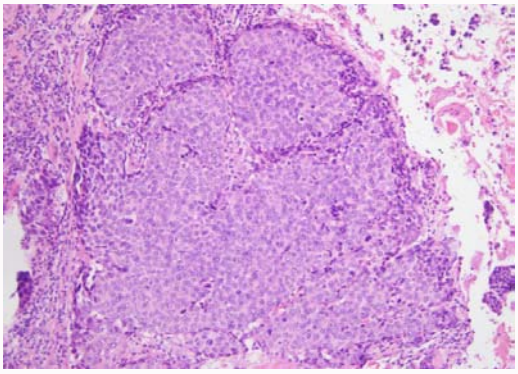

Figure 1: Porocarcinoma, H&E, 100x Nests of malignant tumor cells with necrosis and numerous mitoses.

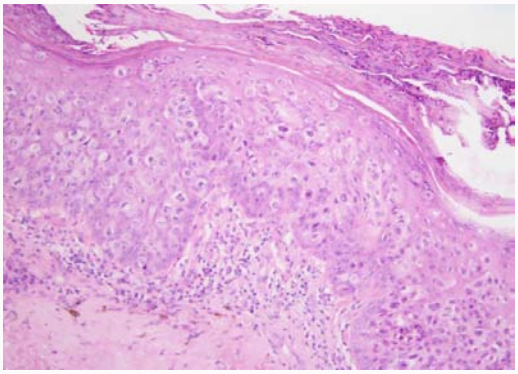

Figure 2: Paget's disease, malignant cells in the epidermis, H&E, 400x malignant epithelial cells with pale cytoplasm in the dermis with pagetoid appearance.

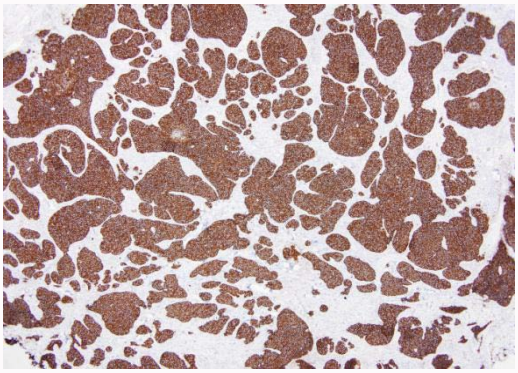

Figure 3: IHC for p16, 100x malignant cells displaying strong reactivity to p16.

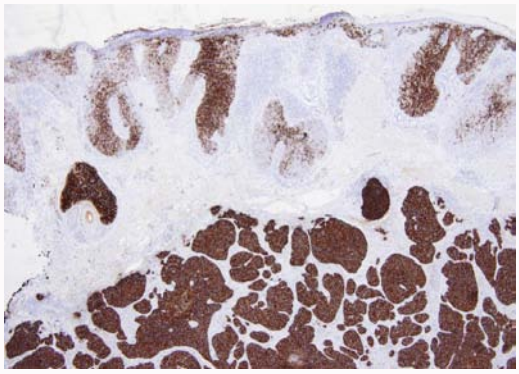

Figure 4: IHC for EMA, 100x malignant cells displaying strong reactivity to EMA.

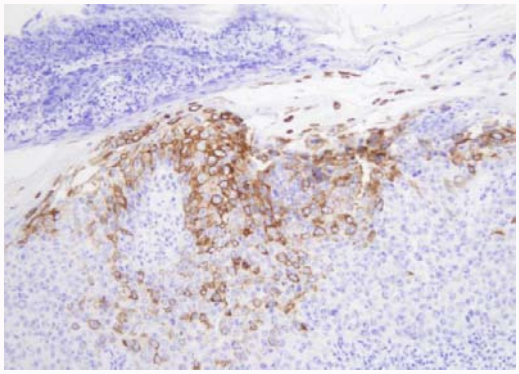

Figure 5: Paget's cells positive for CK7, 200x malignant cells are displaying strong positivity to CK7 in the epidermis.

neoplasms however other body regions are not entirely excluded [4]. Extramammary Paget's disease usually presents as a slowly expanding, sharply demarcated erythematous plaque that may be eczematous, crusting, scaling, or occasionally ulcerated. Pruritis is a common presenting symptom, but pain, burning, irritation, and bleeding can also occur. Establishing an early diagnosis is often difficult and delayed because of nonspecific clinical appearances which may cause a delay in performing a biopsy [5]. Histologically porocarcinomas demonstrate invasive sheets and cords of obviously anaplastic and pleomorphic cells, high mitotic activity and tumor necrosis. The hallmark histopathological finding in extramammary Paget's Disease is the presence of Paget's cells in the epidermis - large cells with an abundant, clear, finely granular cytoplasm, large centrally located nucleus often containing a prominent nucleolus which causes the malignant cells to stand out in contrast to neighboring small bland benign epithelial cells [6]. The above features of porocarcinomas are often misinterpreted as poorly differentiated squamous cell carcinoma or melanoma and IHC is required [7]. In this case IHC ruled out poorly differentiated squamous cell and melanoma. Early diagnosis and resection are paramount to reducing patient mortality and potential metastasis. Nodal involvement is the main and most frequent form of metastatic presentation of porocarcinoma, with cutaneous metastasis and zosteriform presentation being an unusual type [8]. The highly metastatic potential of this disease can pose as a negative prognostic indicator if diagnosis and treatment are delayed and metastasis develops. The average age of diagnosis ranges from 13 to 84 years, with a mean age of 57 years [9]. Literature review yields limited information on the site of origin of these two tumors. Here we report the coexistence of extramammary Paget's disease and porocarcinoma supporting the hypothesis of a potential common origin. The advancement of molecular and genetic testing can potentially assist in delineating the origin of these two tumors. To date, no studies have discovered any shared biomarkers between extramammary Paget's disease and its mammary counterpart; however, further molecular profiling is indicated to gain a more comprehensive understanding of porocarcinomas [10]. This may be helpful in the treatment of the disease and beneficial for optimal patient care if indeed they share similar molecular profiles.

Conclusion

The coexistence of porocarcinoma and extramammary Paget's

disease is a rare neoplastic phenomenon with only a handful of cases reported in the literature. The highly metastatic potential of porocarcinoma can severely impact the morbidity and mortality for individuals if diagnosis and treatment are not defined early. Additional clinical studies including molecular and genetic testing can potentially bring further context to understanding the origins of these two neoplasms, its relationship, (if any) to breast carcinomas, and possible therapeutic treatment regimes.

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