Eccrine Porocarcinoma of the Auricle: A Case Report

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Eccrine porocarcinoma (EP) is a rare skin malignant lesion representing 0.005–0.01% of all cutaneous tumors. It is a tumor that most commonly present in elderly people aged over 60 years. Approximately 250 cases of EP have been reported since this disease was first described in 1963. However, only three cases occurring specifically on the ear (including the current case) have been documented in the literature to date. Based on the rarity of EP of the ear, we present this 78-year-old man with EP on the right ear lobule, which was diagnosed accidentally during the management of other unrelated problems. The etiology, diagnosis, treatment and prognosis of this disease are discussed, with a brief review of the literature in this report.

Key Words: auricle, eccrine porocarcinoma
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Eccrine porocarcinoma (EP) is a rare malignant lesion of skin. Fewer than 250 cases of EP have been reported worldwide since this disease was first described by Pinkus and Mehregan in 1963 [1–10]. Because of the rarity of EP, the etiology and nature of this disease warrants further investigation. Here, we present a case of EP occurring on the auricle and we discuss this disease in a brief review of the international literature.

Case Presentation

This case was a 78-year-old Asian male with diabetes mellitus for more than 30 years controlled with insulin, hypertension for more than 10 years, and end-stage renal disease with hemodialysis therapy for 7 years. He was brought to the otorhinolaryngology clinic of our hospital because of right ear fullness and hearing impairment that had lasted for several days. Under physical examination, the external ear canal was filled with cerumen, and there was a 1.5 × 0.9 × 0.3-cm pedunculate dark-brown tag originating from the right ear lobule. The appearance of this tag was similar to seborrheic keratosis. The patient remembered having had this lesion for about 2 years. No ulceration or any discomfort was noted with respect to the lesion. The neck was free of palpable lymph nodes. The cerumen was then removed and the symptoms completely subsided. The auricular lesion was excised from the base of the stalk under the request of the patient and his family. The specimen was sent for histopathological examination as a routine procedure.

Microscopically, the specimen showed proliferation of clear cells and squamoid cells, mainly confined to the epidermis (Figure 1). There were some small ducts that were lined by cuticles (Figure 2). Cellular atypia and brisk mitotic activity were noted. The immunohistochemical stain revealed focally positive for epithelial membrane antigen, but was negative for

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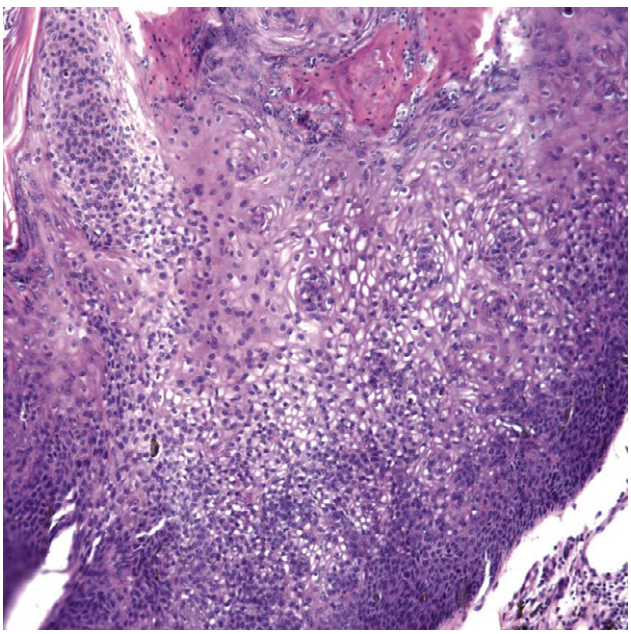

Figure 1. *Intraepidermal proliferation of clear cells and squamoid cells (hematoxylin & eosin; original magnification, 10×).*

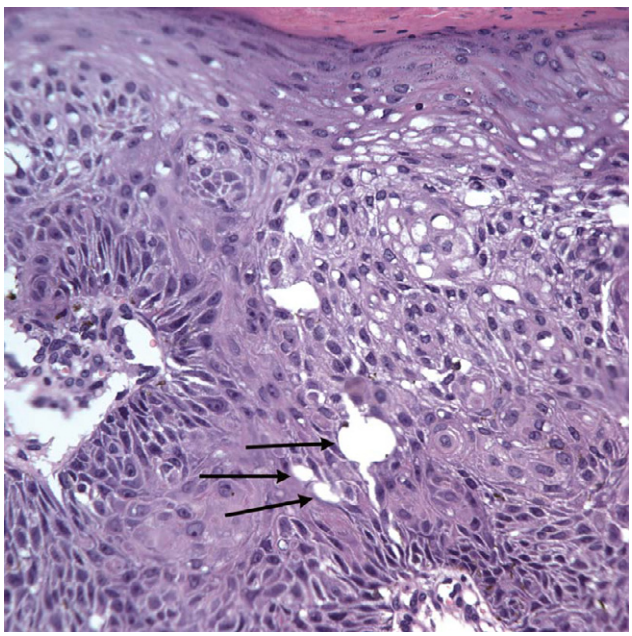

Figure 2. *The small ducts are lined by cuticles, a characteristic diagnostic feature of eccrine porocarcinoma (arrows; hematoxylin & eosin; original magnification, 40×).*

carcinoembryonic antigen. The features suggested eccrine porocarcinoma. After excision, scheduled follow-ups were arranged. To date, there has been no evidence of recurrence at 1 year after the operation.

Discussion

The incidence of EPs accounts for 0.005–0.01% of all cutaneous tumors [1,2]. Most EPs arise on the lower limbs, followed by the trunk and head [2]. Only three cases of EP (including the present case) have been reported to specifically occur on the ear. McMichael and Gay [4] reported a 71-year-old African-American woman with EP on her left ear, who received Mohs surgery and transposition flap repair. She remained tumor-free at her 1-year post-surgery follow-up. Klenzner et al [6] reported a 60-year-old woman with EP on the pinna of her left ear, with acute facial palsy as the initial presenting symptom. She underwent ablation of the left ear, a subtotal left parotidectomy and a selective ipsilateral neck dissection. Although the cause of the facial palsy remained idiopathic, it subsided within 8 weeks of the operation. EP is a tumor that predominantly presents in elderly people aged over 60 years [2,4,6,7,10]. Female predominance was reported for EPs in several studies [2,7], while male predominance was noted in reviews of head and neck EPs [5,10].

The etiology of EP is still unclear. However, the prediagnostic duration of these lesions is usually counted in years, even decades, suggesting that the lesion undergoes malignant changes from the pre-existing benign eccrine poroma [2]. An increased risk of EP has been reported in immunosuppressed patients, such as patients with HIV infection, organ transplantation or diabetes mellitus [10,11].

Because of the rarity and nonspecific appearances of EP, the tentative clinical diagnosis would never be correct, and might be misdiagnosed as squamous cell carcinoma, squamous cell carcinoma *in situ* (Bowen's disease), pyogenic granuloma, basal cell carcinoma, seborrheic keratosis, amelanotic melanoma, verruca vulgaris or metastatic adenocarcinoma [2,6]. Shiohara et al [7] simply described the clinical appearance of EP as a reddish nodule, a reddish lesion, a brown nodule or a brown lesion, and the reddish nodule was the most common feature of EP. The diagnosis of EP is rendered on histopathologic features of intraepidermal ductal differentiation and as either an invasive architectural pattern and/or significant cytologic pleomorphism [2]. Immunohistochemical or specific staining such as for carcinoembryonic antigen, epithelial membrane antigen and periodic acid Schiff may help

diagnose EP but are not exclusive. The immunohistochemical and staining features of EP include highlighted luminal/ductal structures [2,7].

Wide excision of the primary tumor is the therapy of choice, and the curative rate was reported to be 70–80% [12]. It was reported that EP was associated with 17–30% local recurrent rates, 19–50% lymph node spreading rates and 9–33% distant metastasis rates [2,7,10,12]. Robson et al [2] reported that several factors, including mitoses, the presence of lymphovascular invasion and tumor depth >7 mm, were associated with worse prognosis. After reviewing previous literature, Asians seemed to have worse prognoses than Europeans [2,7,10,12]. It is difficult to determine the efficiency of other therapeutic modalities compared with wide excision alone, because they have not been systemically evaluated. However, adjuvant postoperative radiotherapy is considered for EP cases with very poor prognostic factors [5]. In the study by Shiohara et al [7], chemotherapies were given to cases with local recurrence, regional lymph node or distant metastasis; unfortunately, the results were disappointing.

In summary, EP is a rare cutaneous malignancy of elderly people that is predominantly found on the lower limbs and trunk areas, and extremely rarely on the ear. The etiology of EP is unknown, but may originate from malignant changes of long lasting preexisting benign lesions. The diagnosis of EP is by primary microscope and could be assisted by immunocytochemical stains. Wide excision of the primary tumor is the therapy of choice. Histopathologically, mitoses, tumor depth and lymphovascular invasions are predictors of outcome. Clinically, local recurrence, regional lymph node and distant metastases yield poor prognoses. Because the rarity of EP limits large-scale systemic evaluation of its nature and therapeutic efficacy, longer follow-up and a meta-analysis of published studies are warranted.

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耳廓汗腺癌 — 病例報告

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汗腺癌是一種罕見的皮膚惡性腫瘤，約佔所有皮膚腫瘤的 0.005% to 0.01%，是一種主要生長在 60 歲以上成人的疾患。自西元 1963 年此病第一次發表在文獻上，至今共僅約 250 例左右被文獻記載；截至目前為止，僅有三例 (包含本報告病例) 發生在耳部。由於耳部汗腺癌如此罕見，本報告提出一 78 歲男性意外於處理其他病症時發現之右耳垂部汗腺癌病例以供討論；並於本報告中將汗腺癌的病因學、診斷、治療與預後做一簡短的文獻回顧。

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