Received: 31 January 2019 | Revised: 9 March 2019 | Accepted: 12 March 2019

DOI: 10.1002/ccr3.2129

CASE REPORT

Clinical Case Reports Open Access WILEY

Squamoid eccrine ductal carcinoma of the ear helix

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Key Clinical Message

Chronic cutaneous ulcer on the ear helix of 80-year-old male was diagnosed with squamoid eccrine ductal carcinoma (SEDC). SEDC is characterized by both atypical squamous and ductal differentiation. SEDC frequently appears on sun-exposed areas. It could show local recurrence despite complete resection. These characteristics demand cautious attention to SEDC.

KEYWORDS

cutaneous adnexal carcinoma

1 | INTRODUCTION

Squamoid eccrine ductal carcinoma (SEDC) is a rare skin tumor characterized by atypical squamous epithelial proliferation and eccrine ductal differentiation.[1,2] It often presents infiltrative patterns of squamous cell carcinoma in the epidermis and papillary dermis. In reticular dermis and subcutaneous layers, increased atypical ductal structures are found. SEDC typically occurs on sun-exposed body areas and presents as chronically developed nodular or ulcerative lesions. This tumor also shows local recurrence after complete resection.[3]

2 | CASE REPORT

We present a case of an 80-year-old male patient with a persistent, crusted ulcer on his left ear helix. The lesion first appeared several years ago without any physical injury. The size of the ulcer increased slowly over time and was measured at 1.5-cm in diameter at the first clinic visit (Figure 1A,B). The patients denied any pain or tenderness on the lesion during a physical examination. He previously tried a short-term treatment of oral antibiotics that were rendered ineffective. The patient's previous medical history was remarkable for hypertension and type 2 diabetes. We performed an initial punch biopsy for diagnosis. The biopsy revealed atypical epithelial proliferation with keratin pearls and irregular pseudoepitheliomatous lesion. Immunohistochemistry (IHC) noted that the specimen has positivity in P53 stain and increased Ki-67 index. In suspicion of squamous cell carcinoma, a wide excision of the remained tumor was planned. Preoperative axial CT scan revealed no evidence for metastasis or abnormal lymph node enlargement. We removed the skin lesion with a 2-mm resection margin and applied sterile dressing for secondary healing. Histopathology results showed ulcerative, infiltrative lesions extending from the epidermis to the upper dermis with atypical squamous cell proliferation (Figure 2A). Tubular and ductal structures with hyperchromatic nuclei were observed in the deeper layer of the dermis (Figure 2B,C). IHC showed positive staining for CK5/6, CK7, epithelial membrane antigen (EMA), P53, and P63 (Figure 2D). Additional IHC including carcinoembryonic antigen (CEA), S-100, gross cystic disease fluid protein 15(GCDFP-15), and periodic acid-Schiff (PAS) showed negative staining. Such findings were consistent with SEDC. During 3 months of postoperative short-term follow-up, the wound was re-epithelialized remaining partial atrophic scar. The patient kept

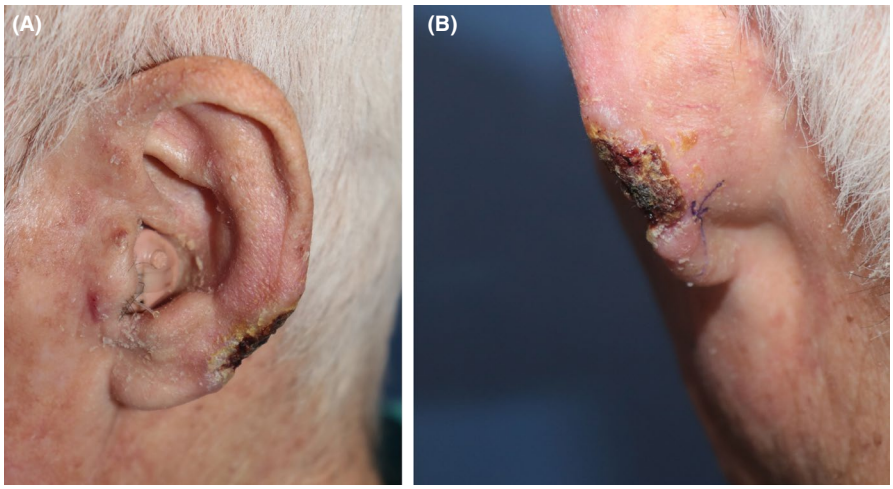

FIGURE 1 A, B, Well-defined, yellowish to black colored, crusted cutaneous ulcer on left ear helix

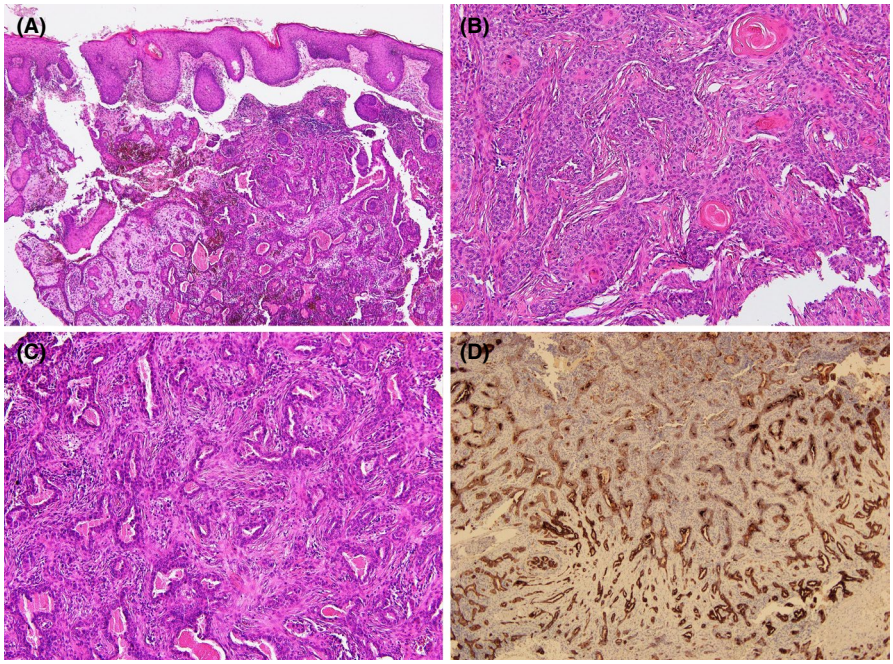

FIGURE 2 A, Irregular pseudoepitheliomatous change and infiltrative ductal growth pattern in the deep dermis are observed (Hematoxylin & eosin, original magnification: ×40). B, In mid-dermis, proliferative squamoid cells and nests coalesce into the desmoplastic stroma. (Hematoxylin & eosin, original magnification: ×200). C, In the deep dermis, ductal proliferation with cytologic atypia and pleomorphism are marked (Hematoxylin & eosin, original magnification: ×200). D, Immunohistochemistry shows diffuse positive stains presenting ductal differentiation (Epithelial membrane antigen [EMA], original magnification: ×100)

follow-up visits at 2-month intervals for another 6 months, showing no signs of recurrence.

3 | DISCUSSION

Squamoid eccrine ductal carcinoma is a seldom reported variant of adnexal carcinoma. Wong et al[2] first described three cases of SEDC in 1997, and only ten case reports and two original articles have been published since.[2-11] Of all cases, involving 37 patients analyzed in the two articles, only four patients were noted for SEDC occurring in the ear.[2,3] This report is the first to present both clinical photos and histopathological findings for a case of SEDC of the ear.

Clinically, SEDC predominantly appears on the head and neck area, presenting as a nodular or ulcerative skin lesion. It often occurs in the middle- and old-aged and tends to more affect male than female. van der Horst et al[3] reported that in 23 of 30 patients, the lesion appeared on sun-exposed areas above the neck. Other five patients had affected areas on the extremities.

Histopathologic features of SEDC show the atypical squamous cell infiltration and eccrine ductal proliferation. The tumors are poorly demarcated and deeply seated with infiltrating stands of atypical epithelial cells. Duct differentiation is often observed in the deeper layers of the tumor, distinct from the atypical squamoid lesion. EMA, CEA immunohistochemistry help highlighting ductal differentiation in most SEDC cases.[3] CK5/6, CK903, Cam 5.2, and CK116 also can show positive staining in a few cases.[5]

Nonmelanoma skin tumors, as well as malignant adnexal skin tumors, should be considered for the differential

diagnosis of SEDC. SEDC is commonly misdiagnosed as squamous cell carcinoma (SCC). In our case, first histopathologic assessment of the initial punch biopsy was also SCC. Shallow shave or punch biopsy specimens would not be able to find both atypical squamous and duct proliferation at the same time because duct-like structures in SEDC are mostly located in the deep dermis. Identifying deep, infiltrating lumina could help to differentiate SEDC from SCC.

Furthermore, SEDC needs to be differentiated from other malignant adnexal skin tumors. Microcystic adnexal carcinoma, syringoid eccrine carcinoma, and cutaneous mucoepidermoid carcinoma could show similar features of SEDC. Microcystic adnexal carcinoma and syringoid eccrine carcinoma are classified as sclerosing sweat duct carcinoma. They have propensities to appear on the face and high rates of local recurrence are reported like SEDC.[4] However, microcystic adnexal carcinoma presents with multiple prominent keratinous cysts and a lack of significant cytologic atypia. Syringoid eccrine carcinoma is comprised of numerous cords, strands, and ducts without keratinous cysts or squamous differentiation.

Cutaneous mucoepidermoid carcinoma is a rare presentation of mucoepidermoid carcinoma. Mucoepidermoid carcinoma mostly presents with primary carcinoma of the major and minor salivary glands. Cutaneous involvement is scarcely reported. It usually appears as asymptomatic nodules or ulcers on the head and neck, as well. Histologically, it shows glandular and squamoid differentiation with mucin-secreting cells. Special staining, such as PAS, can help differential diagnosis by detecting mucin deposition.[12] Such clinical and histopathologic features distinguish cutaneous mucoepidermoid carcinoma from SEDC and other malignant skin tumors.

The choice of treatment for SECD patients is a wide excision of the tumor. Nonetheless, long-term outcomes of SEDC treatment are underreported due to its rarity. It is once regarded as a low-grade malignant potential tumor. Recent studies revealed that SEDC could progress with lymph node metastasis, locoregional, or systemic metastasis.[7,8,13] Frouin et al[4] described that SEDC shows a higher recurrence rate compared to the microcystic adnexal carcinoma and syringomatous carcinoma. A local recurrence and regional lymph node metastasis rate of up to 38% have been reported.[3] Median time to local recurrence was 14 months. Therefore, physicians have to inform the patients of a possibility of regional relapse and maintain a long-term follow-up plan. In conclusion, such defining characteristics of SECD demand close attention for diagnosis and treatment.

CONFLICT OF INTEREST

None declared.

AUTHOR CONTRIBUTION

SY and YL: reviewed medical records and drafted the manuscript. SC: provided pathological analysis. WK: supervised the study.

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How to cite this article: Yim S, Lee YH, Chae SW, Kim W-S. Squamoid eccrine ductal carcinoma of the ear helix. *Clin Case Rep*. 2019;7:1409–1411. https://doi.org/10.1002/ccr3.2129