CASE REPORT

Infundibulocystic Squamous Cell Carcinoma

So Min Kim, Hyeree Kim, Hei Sung Kim, Sang Hyun Cho, Jeong Deuk Lee

Department of Dermatology, Incheon St. Mary's Hospital, School of Medicine, The Catholic University of Korea, Incheon, Korea

Infundibulocystic squamous cell carcinoma was first reported in 2008 as a subset of squamous cell carcinoma arising from the infundibulum of the hair follicle and exhibiting infundibular differentiation. It has well-differentiated, less-differentiated, and infiltrative forms. It was thoroughly analyzed in a series of cases in 2011 by Misago et al. and has been re-defined to include only the infiltrative form owing to its unique clinical and histological characteristics. Here, we report an interesting case of infundibulocystic squamous cell carcinoma in a 72-year-old man presenting with a mass on the left helix of the ear. (*Ann Dermatol* 27(3) 319~321, 2015)

-Keywords-

Squamous cell carcinoma

INTRODUCTION

Infundibulocystic squamous cell carcinoma (SCC) was coined in 2008 by Kossard et al.¹ to describe follicular SCC with infundibular differentiation. It has 3 forms: a well-differentiated form, less-differentiated form, and infiltrative variant. Follicular SCC was in a case series in 2004 by Diaz-Cascajo et al.² as SCC arising from the infundibulum of the hair follicle. In 2011, Misago et al.³ proposed that the term 'infundibulocystic SCC' should be restricted to the infiltrative variant. Here, we report a case compatible with infundibulocystic SCC as described by Misago et al.³.

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Corresponding author: Jeong Deuk Lee, Department of Dermatology, Incheon St. Mary's Hospital, School of Medicine, The Catholic University of Korea, 56 Dongsu-ro, Bupyeong-gu, Incheon 403-720, Korea. Tel: 82-32-280-5700, Fax: 82-32-506-9514, E-mail: leejd@olmh.cuk.ac.kr

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CASE REPORT

A 72-year-old man presented with a painful and at times pruritic, solitary, 1 × 2-cm, crusted, flesh-colored, and rubbery mass on the left helix of the ear that developed 1 year earlier (Fig. 1). The patient complained of bloody discharge. He had no other remarkable history besides hypertension and bronchiectasis. We performed punch biopsy of the crusted center of the mass. The histological findings showed eosinophilic irregularly shaped masses in the dermis infiltrated by inflammatory cells, atypical keratinocytes, and pleomorphic cells with hyperchromatic nuclei. The lesion was suggestive of SCC, so it was excised completely. Histologically, the excised lesion showed infiltrating infundibular canals and radiating cords characterized by atypical keratinocytes with pleomorphism, hyperchromatism, minimal mitosis, dyskeratosis, and keratin pearls in the dermis. There were also multiple cysts filled with laminated keratin (Fig. 2). One month after excision of the lesion, the patient received postoperative external beam radiotherapy for 2 months and has not exhibited recurrence for 3 and half years.

DISCUSSION

In 2004, Diaz-Cascajo et al.² proposed the term 'follicular SCC' to describe a distinct subset of SCC arising from the upper part of hair follicles, i.e., the infundibulum. They studied 16 cases of SCC; histologically, they exhibited atypical squamous epithelial cells arising from the upper part of hair follicles with almost no involvement of the overlying epidermis bordering the involved follicles^{2,4}. However, these cases did not exhibit follicular differentiation.

Kossard et al.¹ termed a subset of SCC with infundibular differentiation as 'infundibulocystic SCC' in order to define a group of tumors with loss of differentiation that might not clinically and/or histologically meet the criteria of keratoacanthoma. Histologically, infundibulocystic SCC is characterized by infundibular proliferation that is ductular, cyst-

Fig. 1. A solitary 2-cm, crusted, flesh-colored, and rubbery mass on the left helix.

Fig. 2. Histological findings of the excised mass. (A) Infiltrating infundibular canals and radiating cords with multiple cysts in the dermis (H&E, $\times 40$). (B) Multiple cysts filled with laminated keratin in the dermis (H&E, $\times 40$). (C) Neoplasm cords or nests composed of atypical keratinocytes with hyperchromatism, pleomorphism, dyskeratosis, and minimal mitosis (H&E, $\times 200$).

ic, or both. The authors classified infundibulocystic SCC into well-differentiated, less-differentiated, and infiltrative forms according to the histologic findings. The less-differentiated form exhibits progressive loss of infundibular differentiation, and the infiltrative variants exhibit numerous irregular infundibulocystic lobules extending into the deep dermis and subcutis without features of keratoacanthoma. The authors conclude the well-differentiated form could be applied to keratoacanthoma. They also emphasize that the presence of proliferative canals or cysts represents true differentiation of the infundibulum and not just the replacement of hair follicles by SCC.

Misago et al.³ studied the clinicopathological features of 8 cases of SCC with infundibular differentiation that included follicular SCC and the less-differentiated and infiltrate variants of infundibulocystic SCC. The follicular SCCs were clinically characterized by nodules with a central keratotic

area, and the less-differentiated forms of infundibulocystic SCC were characterized by nodules with an ulcerative crater. However, the infiltrative variant exhibited the unique clinical feature of an erythematous keratotic plaque.

Histologically, follicular SCCs exhibit radiating neoplastic cords or nests as cytological features, while the less-differentiated infundibulocystic SCCs exhibit a central crater filled with a low to moderate amount of keratin, with neoplastic cells invading the deep dermis. Both forms exhibit 2 to 3 contiguous infundibular structures merging to form a dilated infundibular cystic structure. Because of their similarities, Misago et al.³ propose combining follicular and less-differentiated infundibulocystic SCCs under a single term: either infundibular or follicular SCC.

Interestingly, the infiltrative variants of infundibulocystic SCC exhibit an erythematous keratotic plaque as a unique clinical feature. They have distinct histopathological findings

characterized by numerous micro- or dilated infundibular cysts of various shapes that penetrated into the deep dermis and subcutaneous tissue but do not have the architectural feature of a central keratin-filled crater. The cysts contain laminated or compacted keratins. These histological features are distinct from those of the well- and less-differentiated forms of infundibulocystic SCC. Therefore, Misago et al.³ propose restricting the term 'infundibulocystic SCC' to this infiltrating variant. These histological features may be similar to those of microcystic adnexal carcinoma, which is characterized by numerous keratinous cysts in the superficial dermis as well as small basaloid or squamous nests and infiltrating cords and ductular structures in the deeper dermis. However, unlike infundibulocystic SCC, microcystic adnexal carcinoma has a paucicellular desmoplastic stroma, the cell nuclei are uniform with minimal pleomorphism or atypia, and mitosis is rare^{5,6}.

The present case exhibited a clinical finding of a crusted, flesh-colored, rubbery mass that differed from keratoacanthoma or the infiltrative variant of infundibulocystic SCC mentioned by Misago et al.³. However, the histological findings of the present case included infundibular cords and nests of neoplastic cells, which are suggestive of SCC, as well as laminated keratin-filled cysts which are compatible with infundibulocystic SCC. Therefore, we made a diagnosis of not merely SCC, but as infundibulocystic SCC as redefined by Misago et al.³.

In 2012, Klingman and Chen⁷ raised the issue of whether the infundibulum is epidermal or follicular; they comment that there is no evidence the infundibular epithelium is different from the epidermis. In response, Misago⁸ emphasized the importance of recognizing this infundibular SCC both clinically and histopathologically in order to distinguish it from keratoacanthoma. As mentioned by Kossard et al.¹, the presence of infundibular and cystic changes reflects true infundibular differentiation of a follicular tumor.

In summary, the infundibulocystic SCC is a novel concept introduced in 2008 and mentioned in very few reports. We report an unusual case that meets the histopathological criteria of infundibulocystic SCC as redefined by Misago et al.³.

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