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Unusual Complex Apocrine Tumor Consisting of Syringocystadenoma Papilliferum and Apocrine Hidrocystoma within a Single Lesion: A Case Report

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Syringocystadenoma papilliferum (SCAP) and apocrine hidrocystoma (AH) are benign apocrine neoplasms that usually occur separately. SCAP arises predominantly in head and neck, while AH typically develop in periorbital area. We report a case of a 68-year-old male with an asymptomatic erythematous papulonodule that occurred on his back 3 years ago. Histologic examination showed cystic invagination extending from the epidermis into the dermis with some papillary projections. The invaginated portion was lined by epithelial bilayer composed of cuboidal and columnar cells, and decapitation secretion was observed in the inner epithelial layer. In the deep dermis, multiple cystic spaces with variable sizes were observed, and these cysts also presented double layers of the epithelium and decapitation secretion. According to such histologic features, the coexistence of SCAP and AH within a single lesion was demonstrated. The patient was recommended to completely remove the remaining lesion after punch biopsy, but he refused further surgical management. Herein, we report an unusual case of complex apocrine tumor with a rare composition in an atypical site.

Keywords: Apocrine neoplasms, Hidrocystoma, Syringocystadenoma papilliferum

INTRODUCTION

Syringocystadenoma papilliferum (SCAP) is a relatively rare benign cutaneous appendage tumor that typically occurs in children and young adults, and usually forms as a solitary dark-brown plaque or papules[1]. Approximately 75% of SCAP are located in the head and neck region, most frequently in the scalp[2]. Another benign cutaneous appendage tumor, apocrine hidrocystoma (AH), mostly arises in the face of middle-aged adults, especially along the eyelid. It appears as translucent papules of various colors, ranging from yellowish-brown to dark blue[1,3]. Histologically, the cystic spaces of both SCAP and AH are lined by the epithelial bilayer made up of small cuboidal or columnar cells in the inner layer and myoepithelial cells in the outer layer. In both tumors, decapitation secretion is observed in the inner layer, indicating the apocrine secretion[1-3]. SCAP shows a cystic epidermal invagination with papillary projection[2], whereas AH appears as unilocular or multilocular cysts located within the dermal layer, away from the epidermis[1,3]. Although two or more cutaneous appendage tumors often coexist within a single lesion, the coexistence of SCAP and AH without any other additional tumor is very rare[4]. Herein, we report a case of a complex apocrine tumor in which SCAP and AH coexisted in a single lesion on the back of an elderly male patient. In addition to its rare combination of SCAP and AH, the age of onset and the location of the lesion seemed to be also uncommon considering the typical clinical characteristics of each component tumor of this complex lesion. Therefore, we thought it worthwhile to report our patient's case, focusing on the unusual combination and the discordance with the preferred age and frequent site of each disease.

CASE REPORT

A 68-year-old male patient presented with an asymptomatic erythematous papulonodule approximately 1 cm×1 cm in size on the back that appeared 3 years prior to his visit to the hospital (Fig. 1). The patient had a medical history of hyperlipidemia, but he had no family history of note. A skin biopsy was performed in the papulonodular area of the lesion and revealed two distinct types of cystic lesions in the upper and lower dermis. In the upper dermis, a cystic invagination extending from the epidermis toward the dermis was observed with connection to the epidermal opening. In the lower dermis, multiple cystic structures of various sizes were present (Fig. 2A). The upper portion of the invaginated area was composed of keratinized squamous epithelium, and the lower part of it contained papillary projections extending toward the cystic cavity, that were lined by a two-layered epithelium with inner columnar cells and outer simple cuboidal cells (Fig. 2B).

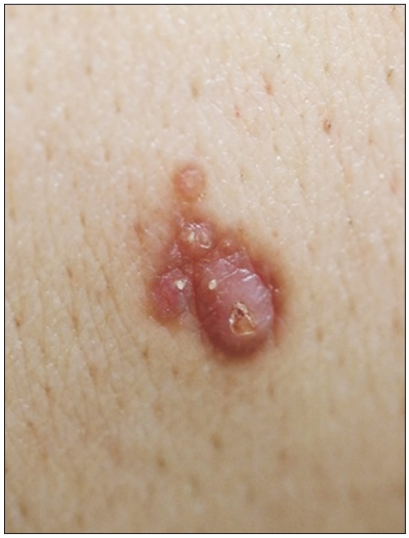

Fig. 1. Erythematous papulonodule approximately 1 cmX1 cm in size on the back.

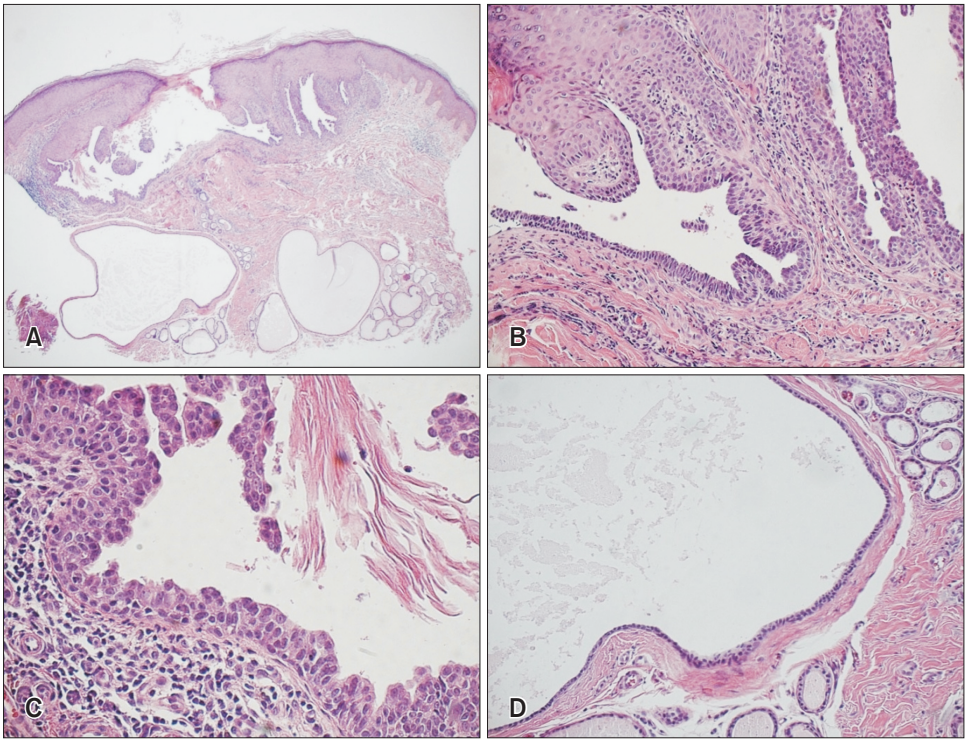

Fig. 2. (A) Two distinct types of cystic lesions coexisting in the upper and lower dermis. A cystic invagination extending from epidermis toward the upper dermis with connection to the epidermal opening. Several cystic cavities of various sizes in the lower dermis (H&E, ×40). (B) The invaginated cyst lined by epithelial bilayer with papillary projections. Columnar epithelial cells in the inner, or luminal, layer and small cuboidal epithelial cells in the outer layer (H&E, ×200). (C) Decapitation secretion in the inner layer of the invaginated cyst and some cellular debris in the cystic cavity. Plasma cell infiltrations in the papillary core (H&E, ×400). (D) A large cyst in the lower dermis, also lined by epithelial bilayer consisting of small cuboidal epithelial cells in the inner layer and myoepithelial cells in the outer layer. Focal decapitation secretion in the inner layer of the deep dermal cystic lesion and some amorphous materials in the cystic cavity (H&E, ×200).

Decapitation secretion was observed in the inner columnar epithelial cells, and cellular debris was found in the invaginated cavity. Plasma cell infiltrations were also observed in the papillary core of the invaginated cavity (Fig. 2C). Apart from this invaginated structure, several cystic cavities of various sizes in the lower dermis were lined by double layers of epithelial cells. The luminal layers of these dermal cysts were composed of cuboidal epithelial cells and had focal decapitation secretion. The outer layers of the dermal cystic lesions were composed of myoepithelial cells whose long axes were parallel to the wall of the cysts. In the cystic cavities of lower dermis, unstained materials without nuclei were found (Fig. 2D). Immunohistochemical (IHC) staining revealed the same pattern of reaction in both types of cystic structures, showing carcinoembryonic antigen (CEA) and epithelial membrane antigen (EMA) expression in the inner layer of epithelium and alpha-smooth muscle antibody (α-SMA) expression in the outer layer. Based on the above clinical and histopathological findings, the patient was diagnosed with a complex apocrine tumor where SCAP and AH coexisted. After a punch biopsy, the patient was recommended to surgically remove the remnant lesion, but he refused further management. We received the patient's consent form about publishing all photographic materials.

DISCUSSION

Tumors of the cutaneous appendages encompass both benign and malignant tumors and can be classified as eccrine, apocrine, follicular, or sebaceous gland tumors, according to their origins and differentiation patterns. Since cutaneous appendage tumors are typically located within the dermis, alterations in the epidermis are relatively mild. Therefore, it is difficult to correctly diagnose these tumors via visual clinical features alone, and skin biopsy is essential to confirm these tumors[1]. Typically, only one type of cutaneous appendage tumor exists within a given lesion but, occasionally, two or more types can coexist within a single lesion[4].

SCAP is a benign tumor originating from apocrine glands. The most frequently involved site of SCAP is the head and neck that accounts for 75% of reported cases, while 20% appear on the trunk, and 5% on the extremities. About 51% of SCAP cases exist since birth, and acquired forms usually arise in early childhood or puberty. However, it is uncommon for SCAP to newly arise at an old age. AH is also a benign cutaneous appendage tumor, known to occur mainly in middle-aged individuals[1,2]. The common sites for AH are the face or scalp, which accounts for 73.7% of all cases. Specifically, 33% of AH tumors are found around the orbital area. 13.7% of these tumors are found on the trunk, and 12% on the extremities[3].

In this report, the patient presented with a complex apocrine tumor consisting of both SCAP and AH. It is atypical for a complex lesion of SCAP and AH to appear on the back, considering the fact that the frequently involved regions of both tumors are the head and neck. It is also unusual for SCAP to occur in a 68-year-old patient.

The most common cutaneous appendage tumor that coexists with SCAP is a nevus sebaceous. About 40% of SCAP cases are associated with it[2]. In this case, however, no evidence of nevus sebaceous was found, but instead, SCAP was associated with AH. According to the study of Ansai et al.[4] that reviewed 308 patients with one or more of the three apocrine tumors (SCAP, AH, and tubular papillary adenoma), 106 patients had a mixture of more than two types of tumors, but only 2 patients showed a pure combination of SCAP and AH. Based on the above results, the composition of the complex cutaneous appendage tumor in this case is considered to be rare. Several previous studies reported cases in which SCAP and AH coexisted in one lesion (Table 1)[5-9]. However, in most of these cases, the combination of SCAP and AH existed together with other additional types of cutaneous diseases[5-7,9]. One case indicating the pure combination of SCAP and AH around the eyebrow was previously reported with a focus on its scarcity[8]. However, in this case, a complex lesion occurred on the back which was not a frequent location for both of the component tumors, making this case more uncommon than previously reported one[8] mentioned above.

Cutaneous appendage tumors are generally diagnosed through a skin biopsy, and sometimes expression of some biomarkers that are detected via IHC staining could yield meaningful results helping the diagnostic process. CEA is an antigen expressed in epithelial cells showing endodermal derivation and differentiation. It is present in the epithelial cell membrane of the secretory ducts of apocrine and eccrine glands. EMA is an antigen expressed in glandular epithelial cells and is present on the cuticles of apocrine and eccrine secretory coils and ducts. α-SMA is present in the cytoplasm of myoepithelial cells[10]. In SCAP and AH, IHC staining for CEA and EMA is positive in the apical surface of the epithelial cells forming the

Table 1. Summary of previously reported cases containing both syringocystadenoma papilliferum and apocrine hidrocystoma

Report	Sex/age (yr)	Site	Composition
Schewach-Millet et al.[5] (1984)	Male/8	Thigh	Syringocystadenoma papilliferum Apocrine hidrocystoma Hidradenoma papilliferum
Lin et al.[6] (2007)	Male/56	Cheek	Syringocystadenoma papilliferum Apocrine hidrocystoma Clear cell syringoma
Arias-Santiago et al.[7] (2009)	Male/19	Pre-sternal area	Syringocystadenoma papilliferum Apocrine hidrocystoma Verruca
Keyal et al.[8] (2019)	Male/60	Eyebrow	Syringocystadenoma papilliferum Apocrine hidrocystoma
Basu et al.[9] (2020)	Female/67	Forehead	Syringocystadenoma papilliferum Apocrine cystadenoma Prurigo nodularis Basaloid follicular proliferation Sebaceoma Nevus sebaceous
Present case	Male/68	Back	Syringocystadenoma papilliferum Apocrine hidrocystoma

luminal layer, with varying degrees of staining[10,11]. Also, the basal cells of SCAP lesion may show positive reaction in IHC staining for α-SMA, but could be negative depending on the degree of tumor maturation and basal cell differentiation[11]. In AH, the outer layer of the epithelium showed positive reaction in IHC staining for α-SMA[10,12]. In this case, IHC staining for CEA, EMA and α-SMA exhibited the typical pattern of reaction for SCAP and AH described in the previous studies[10-12].

AH is a benign tumor, so its treatment is usually focused on cosmesis. In the case of single lesions, surgical removal may be performed, and in multiple lesions, procedures such as laser treatment, electrodesiccation and curettage, or topical atropine application may be performed[1,13]. On the other hand, total resection is recommended for SCAP due to its possibility of transforming into syringocystadenocarcinoma papilliferum, which is a malignant form of SCAP[1]. However, a few cases of SCAP were reported to be successfully treated with CO_2 laser or Mohs micrographic surgery in an area where complete resection was difficult[14,15].

Our present case is unique, as SCAP and AH coexisted in a single lesion without other tumors. Furthermore, the patient's age and the site of the lesion are quite unusual considering the clinical characteristics of each disease.

CONFLICTS OF INTEREST

The authors have nothing to disclose.

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