—Case Reports—

A Case of Syringocystadenoma Papilliferum with Tubular Papillary Adenoma of the Chest

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We report a case of syringocystadenoma papilliferum (SCAP) combined with tubular papillary adenoma (TPA) arising on the chest of a 45-year-old Japanese woman. Histopathological examination revealed the characteristic findings of SCAP in the superficial part of the lesion and those of TPA in the deeper part. We reviewed the English literature about this combination. SCAP and TPA have the same cellular components, but show differences of the general structure. The combination of these two neoplasms is more frequent than expected by most dermatopathologists or pathologists. This combination is frequently seen in patients with nevus sebaceus (NS), but it is also found in patients without NS.
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Key words: syringocystadenoma papilliferum, tubular papillary adenoma, nevus sebaceus, tubular apocrine adenoma

Introduction

Syringocystadenoma papilliferum (SCAP) is a relatively rare neoplasm that has hamartomatous characteristics and was first reported by Pinkus in 1954[1]. It may occur de novo in children, but arises in patients with a history of nevus sebaceus (NS) in about one third of cases[2,3]. Only about 20% of SCAP lesions occur on the trunk, including the axilla, in Japanese patients[3]. SCAP is known to often be accompanied by other adnexal tumors such as tubular papillary adenoma (TPA), apocrine gland cyst (AGC), trichoblastoma, basal cell carcinoma, and sebaceoma[3]. Such combinations are frequently observed in the patients with NS, but are also seen in those without NS[3].

TPA is also called tubular apocrine adenoma (TAA) or papillary eccrine adenoma (PEA), and it is a rare benign neoplasm that features proliferation of the glandular epithelium and myoepithelial cells[4]. Ansai and colleagues reported a large case series of SCAP in Japanese patients; 19 out of 106 tumors were associated with TPA (17.9%), including nine out of 35 (25.7%) with NS and 10 out of 71 (14.1%) without NS[3]. Their report suggests that the association of these neoplasms may not be rare. However, there have only been l4 cases reported in the English literature before the present one[5–18].

Here we report an adult Japanese patient who developed de novo SCAP associated with TPA on the chest without pre-existing NS. We also discuss the reason why few cases of SCAP associated with TPA may have been reported to date.

Case Report

A 45-year-old Japanese woman presented with a right supraclavicular tumor that had existed since childhood. Her family history was unremarkable and her general health was good. Examination at the first visit revealed a 3 mm, non-tender red papule (**Fig. 1**). The lesion was excised under local anesthesia.

Histopathological examination showed that the tumor was slightly elevated and occupied the entire dermis. The epidermis near the lesion was hypertrophic, while the overlying epidermis was replaced by glandular epithelium (**Fig. 2**). The upper part of the tumor contained cysts and irregularly dilated tubules with deep invaginations, from which thick papillomatous projections lined

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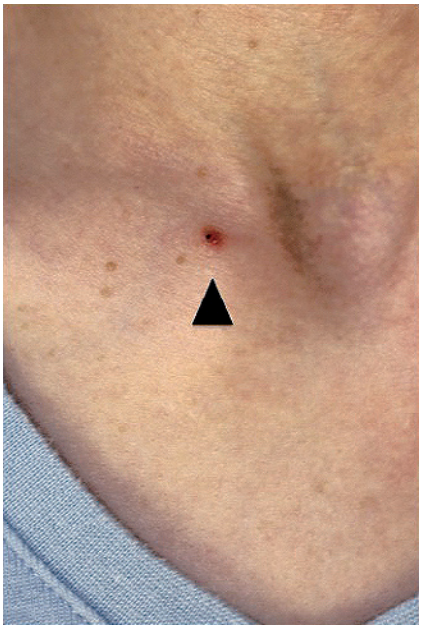

Fig. 1 Clinical findings of the patient at the first visit. A 3 mm non-tender red papule was observed (**arrowhead**).

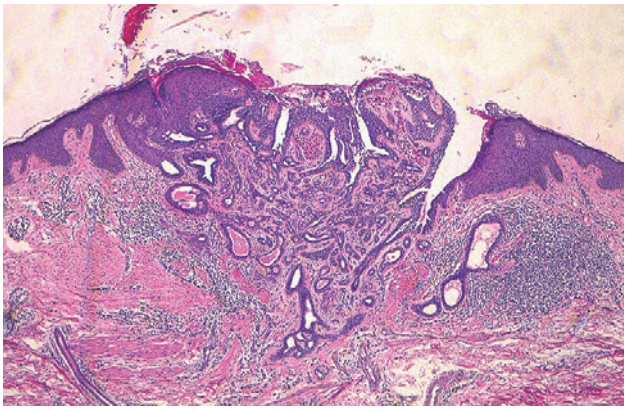

Fig. 2 Histopathological findings at low power. The tumor was slightly elevated and occupied the entire dermis. The epidermis was hypertrophic near the lesion, and the overlying epidermis was replaced by glandular epithelium. (HE stain, original magnification×20)

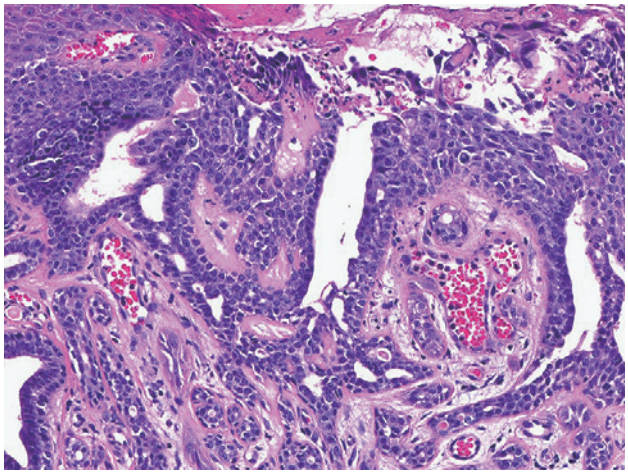

Fig. 3 The superficial part of the lesion contained cysts and irregularly dilated tubules with deep invaginations, from which emerged thick papillomatous projections lined by 2 layers of epithelial cells. Diffuse plasma cell infiltration was observed in the stroma. (HE stain, original magnification×100)

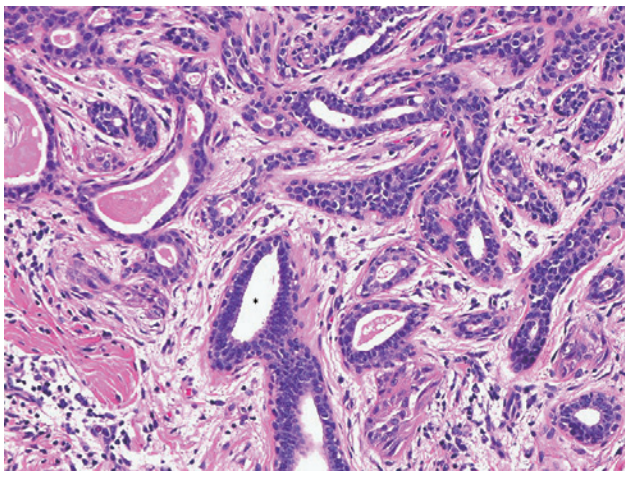

Fig. 4 The deep part of the tumor was composed of relatively small oval tubules, some of which were branching and/or irregular. Some tubules contained intraductal papillary projections. The tubules were composed of 2 layers of epithelial cells. The peripheral layer consisted of cuboidal or flattened cells, while the luminal layer was formed by columnar cells showing decapitation secretion (**asterisk**). (HE stain, original magnification×100)

with 2 layers of epithelial cells emerged (**Fig. 3**). The deep part of the tumor was composed of relatively small oval tubules, some of which were branching and/or irregular in shape. Some of these tubules contained intraductal papillary projections. The tubules were composed of 2 layers of epithelial cells. The peripheral layer consisted of cuboidal or flattened cells, while the luminal layer was columnar cells showing decapitation secretion (**Fig. 4**).

In the superficial part of the tumor, diffuse plasma cell infiltration was observed in the stroma, but the deeper stroma was thicker and condensed around the tubular structures without plasma cell infiltration (**Fig. 3, 4**). We diagnosed the deep part of the lesion as TPA and the superficial part as SCAP.

Discussion

TAA was first reported by Landry and Winkelman[20], but Falck and Jordaan[19] considered it to be identical to PEA that was first described by Rulon & Helwig[21] and they proposed the term tubulopapillary hidradenoma. Other authors disagreed with their opinion, because PEA does not show decapitation secretion or any connection with the epidermis[22]. Though TAA is frequently located on the scalp, PEA is primarily located on the extremities. TPA contains a spectrum of lesions from PEA to TAA. Recently, the term TPA has been used for such lesions in the World Health Organization classification of skin tumors[4], because it is sometimes difficult to precisely distinguish between these tumors.

The characteristic feature of TPA is numerous irregular tubular structures usually lined by 2 or more layers of epithelial cells that are scattered in the dermis and sometimes the subcutaneous tissue. The outer layer of cells is generally cuboidal and exhibits myoepithelial differentiation, while the inner layer (luminal cells) is usually columnar and the cells often show decapitation secretion.

On the other hands, SCAP has a cystic and papillary structure composed of epithelial cells projecting downward into the dermis and it opens onto the skin surface via one or more orifices. Like TPA, this tumor is composed of two layers of epithelial cells. The outer layer is formed by cuboidal cells that show myoepithelial differentiation and the inner layer is columnar cells that demonstrate decapitation secretion. Dense plasma cell infiltration is often observed.

TAA was first reported in 1972[20], but Fisher[23] thought that it was a variant of SCAP. Umbert and Winkelmann[24] reported that TAA could be differentiated from SCAP because TAA lacked cystic dilated apocrine invaginations extending downward from the epidermis and also did not show stromal infiltration of plasma cells. Other authors have agreed with their opinion[5,22]. Ishiko et al.[8] reviewed 19 cases of TAA reported in the literature. They described several points for differentiating between TAA and SCAP: 1) TAA does not have cystic dilated apocrine invaginations extending down from the epidermis; 2) TAA has no papillary projections; and 3) infiltration of plasma cells is rare or absent in TAA. The histopathological features of our case are similar to those of other reports[5–18], with SCAP in the superficial part and TAA in the deeper part.

As mentioned above, both TPA and SCAP have the same components, which are apocrine epithelium and myoepithelial cells, so differentiation between these two neoplasms is mainly based on the general structure of the lesion. Ansai and colleagues reported[3] that TPA and SCAP sometimes exhibit histopathologic overlap. However, only 14 previous cases have been reported in the English literature, as stated above[5–18]. Kazakov et al. perfomed a study involving histopathological reappraisal of tubular adenoma (TA) and SCAP by four dermatopathologists[25]. They reported that TA and SCAP have similar histopathological findings and that there are no universally accepted diagnostic criteria for classifying these lesions, even among experienced dermatopathologists and pathologists. SCAP and TPA have the same cellular components despite differences of the general structure, which might be the reason Fisher[23] thought that TAA is a variant of SCAP. This might also explain why the combination of these two neoplasms is more frequent than expected by most dermatopathologists and pathologists. Furthermore, this combination is frequently seen in NS patients (7 out of 15 reported cases including our case[5–18] and 19 out of 29 cases reported by Ansai and coworkers[3]), but there are also such cases in patients without NS.

We consider that there are also similarities between AGC and these two neoplasms. For example, lesions exhibiting both AGC and TPA may have been designated as apocrine cystadenoma. Perhaps we should accept the concept of unifying SCAP, TPA, and AGC.

Conflict of Interest: None declared.

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