ODÜ Tıp Dergisi / *ODU Journal of Medicine*
http://otd.odu.edu.tr

Olgu Sunumu	***Case Report***
Odu Tıp Derg	*Odu J Med*
(2015) ek:44-47	*(2015) supplement:44-47*

Syringocystoadenoma Papilliferum: Two Cases Reports and Review of the Literature

Syringocystoadenoma Papilliferum*: İki Olgu Sunumu ve Literatürün Yeniden Gözden Geçirilmesi*

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Yazının geliş tarihi / Received: 14.01.2014 Düzeltme / Revised: 21.07.2014 Kabul tarihi / Accepted: 25.12.2014

Abstract

Syringocystadenoma papilliferum is an uncommon cutaneous adnexial tumor of uncertain histogenesis. Our patients, 58 and 48 years old, on the scalp and postauricüler located are male patients with nodular lesion. Prominent hyperkeratosis and multiple papillary projections were noted. The underlying dermal component was composed of numerous, closely spaced tubular structures; often with a wide dilation and these tubular structures. A dense inflammatory infiltrate composed of plasma cells was noted. Immunohistochemical examination of Kappa and Lambda viewed with biklonal immunoreactivity. Our cases are presented because of its rarity.
Keywords: Ecrine, Apokrin, Adnexial tumor

Özet

Syringocystadenoma papilliferum, histogenezi belirsiz derinin nadir görülen adneksiyel bir tümörüdür. Olgularımız, 58 yaşında skalpte ve 48 yaşında postauriküler yerleşimli nodüler lezyonu bulunan erkek hastalardır. Hiperkeratoz ve bazal epidermiste çok sayıda papiller uzanımlar vardır. Dermal komponentin altı, epitelyal kordlarla nadiren devam eden tübüler yapılar, genellikle geniş dilatasyon ile çok sayıda, yakın aralıklı tübüler yapılardan oluşmaktadır. Plazma hücrelerinden oluşan yoğun bir inflamatuar hücre infiltrasyonu vardır. İmmunohistokimyasal incelemede Kappa ve Lambda ile biklonal immünreaktivite izlenmiştir. Nadir görülen bir tümör olması nedeniyle olgularımız sunulmuştur.
Anahtar kelimeler: Ekrin, Apokrin, Adneksiyal tümör

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Introduction

Syringocystadenoma papilliferum is a rare skin adnexal tumor first described by Petersen in 1892 and most commonly occurs on the scalp and face. Clinically, tumor arises as a single nodule or several small nodules frequently in association with anorganoid nevus (1). Histopathological examination of the mass revealed an aggregation of neoplastic cells with cellular proliferation extending from the epidermis to the dermis. The remaining part of the tumor was composed of cystic invaginations with numerous projections oriented toward the lumen. There were two rows of cells on the projections; the cells on the luminal side were columnar and those at the apical aspect were small cuboidal cells. These histological changes were characteristic of syringocystadenoma papilliferum (SCAP). Syringocystadenoma papilliferum (SCAP) occurs singly or in association with other tumors, usually sebaceous nevus or basal cell carcinoma. Approximately one-third of recorded cases of SCAP arise in association with a pre-existing nevus sebaceous of Jadassohn. Less common lesions that are associated with SCAP include apocrine adenoma, condyloma accuminatum, hidrocystoma and hidradenoma papilliferum, poroma folliculare, apocrine acrosyringeal keratosis and giant comedo. Malignant degeneration within SCAP is rare and includes ductal sweat gland carcinoma, malignant SCAP (2). We report two cases because of the rarity of these tumors.

Case presentation

A 58-year-old man presented with a 10x10 mm ulcerative nodular lesion on the scalp. The other case was 48-year-old having a 7x5x2 mm gray colored tissue on the postauricular region. Clinical differential diagnosis included an epidermal cyst, basal cell carcinoma, plasmacytoma, keratoacanthoma or another benign adnexal tumor. One biopsy exhibited hyperplastic epidermis with keratoakantoma like appearance (Figure 1). The epidermis showed prominent hyperkeratosis. Multiple papillary projections were noted in basal epidermis (Figure 2). The underlying dermal component was composed of numerous, discrete, closely spaced tubular structures, often with a wide dilation, and these tubular structures were rarely continuous with epithelial cords. These were lined by a bilayered epithelium composed of a luminal row of columnar and cuboidal cells (Figure 3). A fair number of the columnar cells showed decapitation secretion. A dense inflammatory infiltrate composed of plasma cells was noted. Immunohistochemical staining for CD38 (Figure 4) and CD 138 positive, Kappa and Lambda viewed with biclonal immunoreactivity.

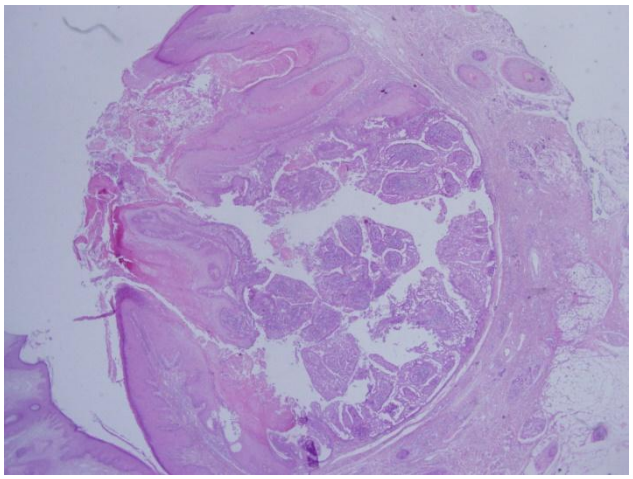

Figure 1. Hyperplastic epidermis with keratoakantoma like appearance, H-E, x40

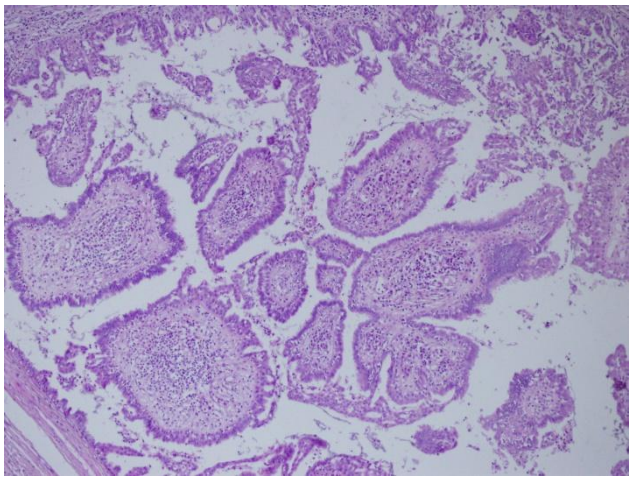

Figure 2. Multiple papillary projections in basal epidermis H-E, x100

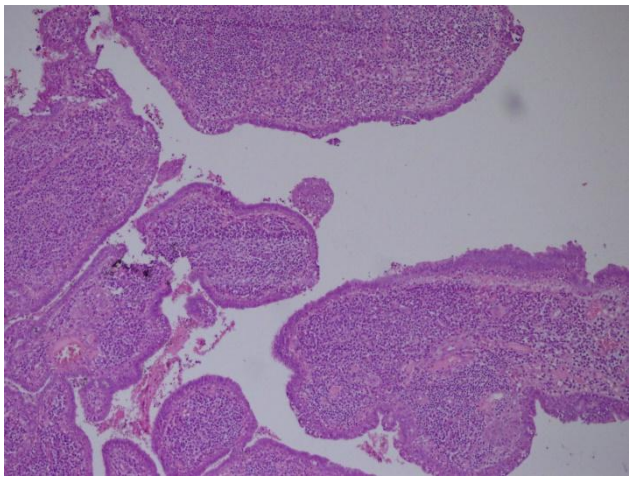

Figure 3. The underlying dermal component: discrete, closely spaced tubular structures with a wide dilation, rarely continuous with epithelial cords. H-E, X100

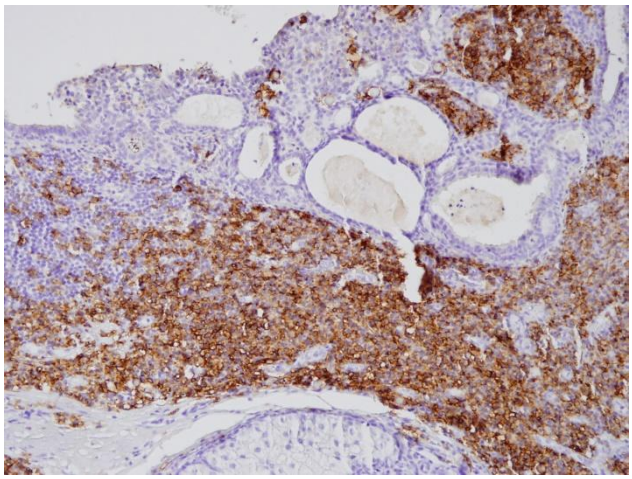

Figure 4. Immunohistochemical staining for CD38 İHK, CD38, X200

Discussion

Syringocystadenoma papilliferum has often been reported to arise in association with other tumors. Helwig and Hackney reported the association of SCAP with a sebaceous nevus in 34 of 100 SCAP cases and the association of SCAP with a basal cell carcinoma in nine of 100 SCAP cases. Fujita et al. reported that SCAP was, most frequently associated with a sebaceous nevus (40 cases) in a total of 126 SCAP cases, followed by basal cell carsinoma (13 cases), sebaceous epithelioma (4 cases), apocrine hydrocystoma (4 cases), trichoepithelioma (2 cases), and ecrine spiroadenoma (1 case), in descending order (3). The coexistence of SCAP with other lesions has been reported, including apocrine acrosyringeal keratosis, cutaneous horn, papillary ecrine adenoma, condyloma accuminatum, tubular apocrine adenoma, verrucous carcinoma, giant comedo, poroma folliculare, congenital papillated apocrine cystadenoma, and hydradenoma (4). Our cases were not associated with any lesion. In conclusion, the tumor epithelium lining the cystic invagination or lumen in Syringocystadenoma papilliferum was composed of several cell types demonstrating various developmental stages. The clear cells were considered to have a functional significance as stem or progenitor cell populations and to be involved in tumor growth and evolution. The basal tumor cells demonstrated a tendency to differentiate toward basal cells in the transitional portion or myoepithelial lineage, and luminal cells toward ductal or secretory epithelium. When compared with the adult sweat glands, however, the degree of differentiation in most tumor cells was less mature. The results obtained there support the classical concept that syringocystadenoma papilliferum is a hamartomatous tumor that arises from pluripotent cells (5). Syringocystadenocarcinoma papilliferum resembles syringocystadenoma papilliferum and is marked by funnel-shaped epidermal invaginations demonstrating a gradual transition from keratinizing squamous epithelium at the surface to variable layers of glandular epithelium within subjacent cystic spaces. This epithelial transition mirrors the physiologic epithelial transition of the apocrine gland to the follicular infundibulum. In the healthy apocrine gland are two layers of bland epithelial cells, an inner luminal layer and an outer basal layer, which also line the papillary projections of the benign syringocystadenoma papilliferum. In the malignant counterpart, the epithelial cells vary in thickness and show malignant cytological features, such as high nuclear to cytoplasmic ratios, nuclear irregularity, and coarse chromatin. Secondly, previous reports described long-standing lesions with new onset of bleeding or ulceration accompanying the malignant progression (6-8). Most of the cases were aged 60 and old. Scalp is the predilection site. The lesion size varied from 1.5 to 13 cm. The sizes of our cases were 1 cm and 0.7 cm. Most of the cases had long duration before admission. Histopathologically, SCACP and SCAP demonstrate irregular papillomatous invaginations lined by cells neighboring the stroma. Plasma cells and lymphocyte infiltration of the stroma, especially in the papillary projections, is another characteristic view. On the other hand, nuclear and cellular atypia, loss of polarity, and full-thickness proliferations of neoplastic cells disrupting the two-layered organization indicate SCACP. Squamous cell carcinoma, basal cell carcinoma and malignant melanoma should be considered. Clinically for the differential diagnosis and the histological differences between these tumors are well-defined (2). Although the histological appearance of SCACP is characteristic, primary or metastatic adenocarcinoma of other origins should be excluded. Some malignant tumors of the gastrointestinal tract, breast, and thyroid; renal cell carcinomas; and other tumors may metastasize to the skin in advanced stages. These lesions usually mimic the original tumor, although not always. SCAP is usually a benign tumor and surgical excision is curative. No recurrence was determined at 1-year follow-up of our patients.

References

1. Shindo M, Yamada N, Yoshıda Y, Yamamoto O, Morino S. V. Syringocystadenoma papilliferum on the male nipple, Journal of Dermatology 2011; 38: 593–6.
2. Monticciolo N. L, Schmidt J. D, sand Morgan M. B. Case Report: Verrucous Carcinoma Arising Within Syringocystadenoma Papilliferum. Annals of Clinical & Laboratory Science, 2002; 32 (4): 434-7.
3. Suzuki T, Ikeda H, Hamasaki Y, Hatamochı A, Yamazaki S. Case Report:Syringocystadenoma papilliferum associated with apocrine poroma, Journal of Dermatology 2006;33: 249–51.
4. Yamato O, Doı Y, Hamada T, Hısaoka M and Sasaguri Y, An immunohistochemical and ultrastructural study of syringocystadenoma papilliferum, British Journal of Dermatology 2002; 147: 936–45.
5. Leeborg N, Thompson M, Rossmiller S, Gross N, White C, Gatter K. Diagnostic Pitfalls in Syringocystadenocarcinoma Papilliferum: Case Report and Review of the Literature, Arch Pathol Lab Med, 2010;134(8), 1205-9.
6. Elfatoiki Fz, Khadir K, Ouakadi A, Azzouzi S, Bahechar N, Benchikhi H.Syringocystadenoma papilliferum: Unusual location, Dermatol Online Journal2011; 17 (6), 7.
7. Misago N,M.D. Letter To The Editor, Syringocystadenoma papilliferum with extensive apocrine nevus, Journal of Dermatology 2006; 33: 303–5.
8. Aydın O. E, S. Barıs, Selman Ozkan H, And Gore O. A Rare Tumor: Syringocystadenocarcinoma Papilliferum Dermatol Surg 2011;37:271–4.