# Syringocystadenoma Papilliferum

*SİRİNGOKİSTADENOMA PAPILLIFERUM*

**Ayşe BOYVAT, Rana (Yavuzer) ANADOLU, Cengizhan ERDEM**

Ankara University Medical School Department of Dermatology

**SUMMARY**

*A rare case of a syringocystadenoma papilliferum with a red solitary on the posterior neck is presented.*

*Syringocystadenoma papilliferum is an uncommon adnexal tumor that commonly occurs as a congenital lesion with 75% of cases being located on the head and neck. Ft has distinct dermatopalhologic features, and although most of the lesions are apocrine derived the origin of syringocystadenoma papilliferum is still being debated. Due to the infrequent development of secondary basal cell carcinoma surgical excision is the treatment choice.*

ÖZET

*Boynun arka kısıntında lokalize kınımı solitcr plak ile karekterize bir siringokistadenoma papillifenım olgusu sunulmuştur. Siringokistadenoma papillifenım %75 olguda baş ve boyunda yerleşen, sıklıkla konjenital, nadir bir adneksiyel tümördür. Lezyonların çoğu apokrin kökenli olmakla birlikle siringokistadenoma papilliferumıın nereden geliştiği hala tartışmalıdır. Sık olmayarak sekonder bazal hücreli karsinom gelişebildiği için cerrahi esktasvon seçkin tedavidir.*

Keywords: Syringocystadenoma papilliferum

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## INTRODUCTION

Syringocystadenoma papilliferum is a rare benign tumor of sweat gland origin. The majority of the lesions are found in the region of the head and neck (1-4). The lesion is often present at birth as a single, hairless, smooth, raised area that grows very slowly to become an elevated plaque and then a verrucous lesion (2,5,6). In about one-third of the cases syringocystadenoma papilliferum is associated with nevus sebaceus; and in about one-third of the cases basal cell carcinoma develops within the lesion (4,5,7).

We report a case of a syringocystadenoma papilliferum in a 19 year old girl.

## CASE REPORT

A 19 year old girl attended the Dermatology Department of Ankara University Medical School in November 1989 with a lesion on her nape. She satated that it had been present since birth and an incerase in the size of the asymptomatic lesion had been noted when she was 15 years old. History of an occasional mucopurulent discharge from the lesion was present.

On examination she had a 4x2 cm red solitary plaque composed of 2-5 mm shiny papules of firm

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Yazışma Adresi: Dr.Ayşe BOYVAT
Ankara University Deparment of
Dermatology, ANKARA

consistency on her nape (figl). Her general condition was good and systemic examination revealed no abnormality. Results of routine laboratory investigations were normal.

A biopsy was performed from the lesion which showed typical dermalopathologic features of syringocystadenoma papilliferum (Ankaia Univeisity Medical School Dermatopalholugy Lab. 909/89). Routine hematoxylin and cosin staining revealed villous projections within cystic invaginations of a papillomatous epidermis lined by two layers of cells-tall columnar apocrine secretory cells and small cuboidal cells. A dense inflammatory infiltrate consisting mainly of plasma cells v-erc present in the dermis (Fig 2).

## DISCUSSION

Syringocystadenoma papilliferum is usually first noted at birth or in early childhood, and approximately one third of syringocystadenomas develop in conjunction with an already existing nevus scbaceus on the scalp (4,6-8). It consists of either one papule or several papules in linear arrangement or a solitary plaque. Close inspection reveals thet the plaques comprises clusters of papules 2-10 mm in diameter, while the plaques themselves attain a diameter as large as 4 cm. Some of those papules maybe umbli cated and simulate molluscum coniagiosum (2,4,^).

Approximately 75% of the lesions are located on the head and neck, but they have been found on the shoulder girdle, axillae, chest, thighs, and in the genital area as well (1-4). In our case the localization of the lesion and its clinical features are typical for syringocystadenoma papilliferum.

In a fully developed lesion the epidermis shows varying degress of papillomatosis. One or several cystic invaginations extend downwards from the epidermis. In the lower portion of these invaginations numerous papillary projections extend into the lumina lined by glandular epitclium consisting of two iow~ of **i** ells. The luminal row consists of high columnar icll.s and occasionally some of these cells show active decapitation secretion The outer row consists of small cuboidal cells. A highly diagnostic feature is the almost invariable presence of a dense cellular infiltrate composed almost entirely of plasma cells in the stroma of the tumor, especially in the p.»pil|jiy projections (5). In out case the dermatopiihologii examination of the lesion revealed characteristic features of syringocystadenoma papilliferum.

There is no unanimity about the direction of differentiation in syringocystadenoma papilliferum. **Results** of electron microscopic studies and hi'-tochcmical analyses have been contradictory (5,8,10-12). Pinkus, Lever and Sehaumberg Lever agree that although most of the lesions are apocrine derived, occasional ecrine derived tumors also occur. Accoding to these authors the tumor originates from undifferantiatcd pluripotcntial cells (2,5,6,8,10). This might also explain why a tumor with apocrine differentiation is located 90% to areas normally devoid of apocrine glands, and why it occurs with such frequency at an age when these glands are anatomically and functionally inconspicuous.

In about one-third of the cases syringocystadenoma papilliferum is associated with a nevus sebaeus. Other associated tumors are trichoepithelioma, scbaceus epithelioma, apocrine hydocystoma and ecrine siproadenoma (4,7,8,10,13). In about 10% of the cases a basal cell cercinoma develops but this is noted only in lesions that also exhibit a nevus sebaccus. Very few instances of transition into an adenocarcinoma with regional lymph node metastases have been reported (4,5,7). In our patient no other coexisting lesion like nevus scbaceus or trichoepithelioma has been found. Due to the infrequent development of secondary basal cell carcinoma surgical excision is the treatment of choice. Radiotherapy is ineffective (7,8). In the present case total surgical excision was carried out.

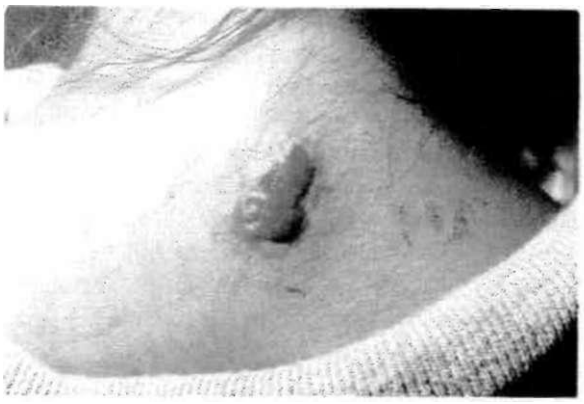

Fig 1. Clinical appearance of the case.

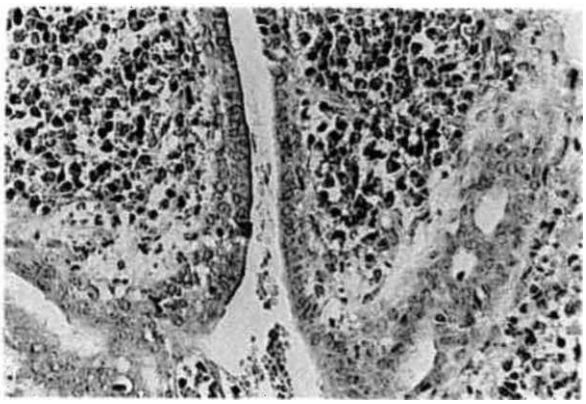

Fig 2. Histopathologic picture of syringocystadenoma papilliferum is characteristic with the glandular epitelium that consists of double rows of cells which shows decapitation secretion in the luminal part (IIEx40).

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