

## CASE REPORT

### Syringocystadenoma papilliferum arising in a naevus sebaceus

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#### Abstract

Naevus sebaceus is a cutaneous hamartoma with the potential of developing into benign or malignant neoplasms. Syringocystadenoma papilliferum (SCAP) have been reported to originate from naevus sebaceus. SCAP is a rare, benign adnexal skin tumour of apocrine or eccrine type of differentiation which typically presents as a nodule or a plaque on the scalp or face. We report a case of syringocystadenoma papilliferum arising in an undiagnosed pre-existing naevus sebaceus in a 56-year-old female.

*Keywords:* Dermatology, hamartoma, benign neoplasms

#### INTRODUCTION

Naevus sebaceus is a cutaneous hamartoma with hyperplasia of the epidermis, sebaceous glands and apocrine glands. It usually occurs in the head and neck region and is almost always present since birth or childhood. Naevus sebaceus has been well-documented to have the potential of developing into both benign and malignant neoplasms. 18.9% of patients with naevus sebaceus have secondary benign neoplasms while another 2.5% have malignant neoplasms.<sup>1</sup> We report a case of syringocystadenoma papilliferum (SCAP) arising from a naevus sebaceus. Due to its relative rarity, SCAP is often underdiagnosed. Although a benign tumour, it has the ability to mimic a malignancy.

#### CASE REPORT

A 56-year-old Malay lady with underlying type 2 diabetes mellitus, hypertension and gout was referred to our Dermatology Clinic by her local government clinic with a swelling over her right temporal scalp. This lesion has been present since birth with associated alopecia over the lesion. However, over the last 2 months prior to presentation, it has been increasing in size with intermittent bleeding. She was initially treated with oral antibiotics and dressings without improvement. On physical examination, there

was a linear, fleshy papillomatous exophytic mass measuring 3 cm x 2 cm over her right temporal scalp with surrounding erythema, maceration and pus discharge (Fig. 1). There was no palpable lymphadenopathy. An excisional biopsy was performed under local anaesthesia. The lesion was excised completely with margins of 0.6 cm and with a depth up to the subcutaneous plane.

#### Pathology findings

We obtained a scalp biopsy sample measuring 1.1 x 1 x 2 cm which showed a pedunculated pinkish tumour arising from the epidermis. Histopathological examination showed papillomatosis with cystic invaginations extending downwards from the epidermis. Numerous papillary projections were extending into the lumina of these invaginations (Fig. 2). The invaginations and papillary projections were lined by glandular epithelium consisting of an outer layer of small cuboidal cells and inner layer of tall columnar cells. Underlying apocrine metaplasia was also seen, suggesting the presence of a naevus sebaceus (Fig. 3). There were no sebaceous glands seen in the sample. No atypical or malignant cells were identified. There was no lymphatic/vascular permeation noted. Surgical margins were clear.

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FIG. 1: Preoperative picture shows a papillomatous exophytic mass over the right temporal scalp.

*Clinical course*

Follow-up at 2 months showed a well-healed scar with no recurrence of the tumour. The patient continues to have regular visits in the Dermatology Clinic.

**DISCUSSION**

SCAP is the second commonest benign neoplasm

arising from a pre-existing naevus sebaceus after trichoblastoma. However, in Taiwan, SCAP is more common in patients with naevus sebaceus compared to trichoblastoma.<sup>2</sup> The histological findings of SCAP arising from a naevus sebaceus is characterised by papillomatous hyperplasia of the epidermis as well as the irregular duct like structures and cystic spaces lined with a double layered epithelium.<sup>3</sup>

SCAP can be derived from both eccrine and apocrine glands. A positive immunoreactivity for gross cystic disease fluid proteins 15 and 24 and zinc-2 glycoprotein demonstrates evidence of apocrine differentiation.<sup>4</sup> However, immunohistochemical analysis of cytokeratins in syringocystadenoma papilliferum demonstrates similarities to eccrine poromas and the ductal component of eccrine glands.<sup>5</sup> It is likely that SCAP arises from undifferentiated cells with the potential to exhibit both apocrine and eccrine characters.

Katoulis and Bozi have described 3 types of SCAP. Our patient had the first type, namely the plaque type, which presents as a hairless plaque on the scalp and is often associated with naevus sebaceus. It is generally apparent at birth and gradually enlarges after puberty. The second type is the linear type, which usually presents as multiple papules with umbilication and may therefore be misdiagnosed as molluscum contagiosum. Although our patient presented with a linear lesion, it lacked a verrucous

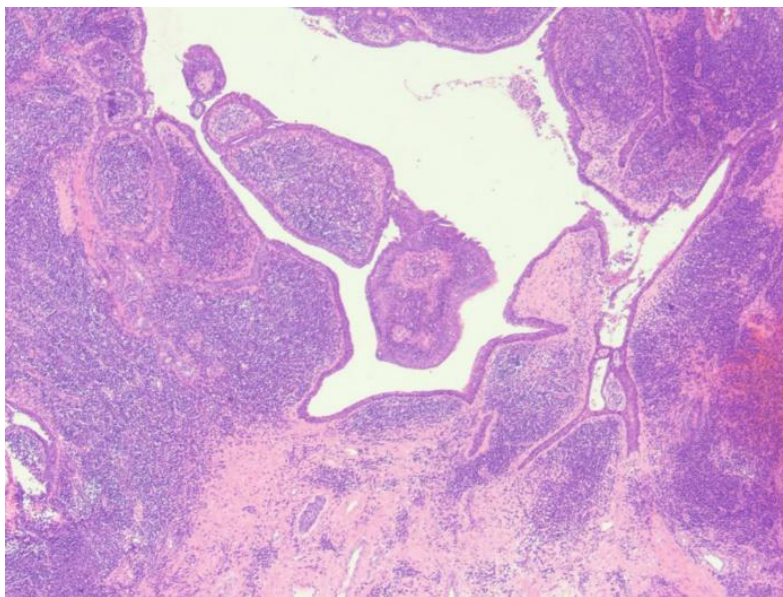


FIG. 2: Cystic invaginations with numerous epithelial papillomatosis which was lined by an outer layer of small cuboidal cells and inner layer of tall columnar cells (H&E, x4).

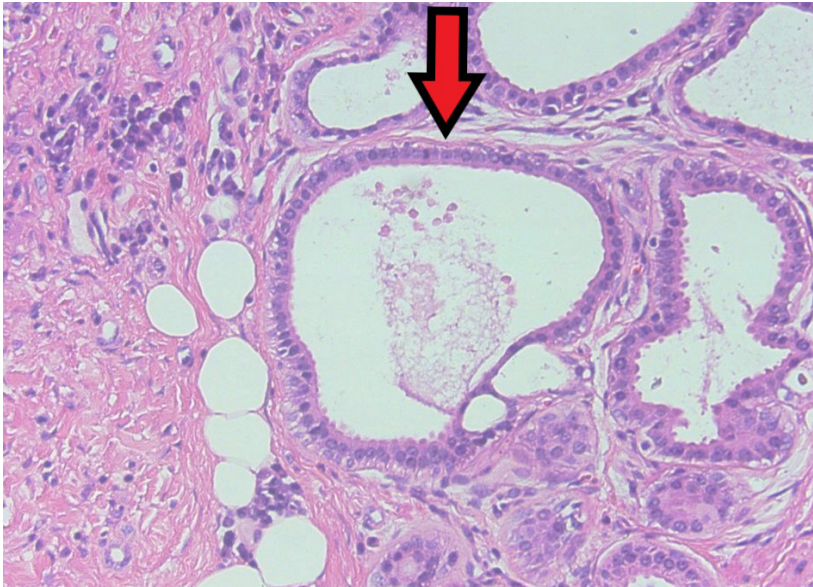


FIG. 3: Sweat glands with apocrine metaplasia (red arrow), a feature of nevus sebaceus (H&E, x10).

surface with umbilication. The last type is a solitary nodule which generally presents on the trunk. Malignant transformation of SCAP to basal cell carcinoma (BCC), squamous cell carcinoma (SCC), or syringocystadenocarcinoma papilliferum (SCACP) may occur. BCC transformation has been reported in up to 10% of cases, whereas transformation to SCC and SCACP occurs less frequently.<sup>6</sup> Ulceration and rapid enlargement of the lesion could indicate malignant transformation. Due to the risk of malignant transformation, as well as its varied presentation, the diagnostic and treatment modality for SCAP is excision with histopathological confirmation. Excision can be done by surgical excision, Mohs micrographic surgery or carbon dioxide laser excision.<sup>7</sup> For naevus sebaceus, surgical excision may be delayed till after adolescence as risk of malignancy is low.

## CONCLUSION

SCAP is a rare tumour that usually occurs in the head and neck region. Clinically it mimics squamous cell carcinoma. In our case, the lesion was histologically confirmed as SCAP with the presence of apocrine metaplasia, suggesting the presence of a previously undiagnosed naevus sebaceus. As it is a challenging entity to diagnose clinically, high awareness and histological confirmation is required. Treatment is by complete surgical excision.

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