

# Intradermal Syringocystadenoma Papilliferum on the Popliteal Fossa: A Rare Dermal Variant in an Atypical Location

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## ABSTRACT

Syringocystadenoma papilliferum (SCAP) is a benign adnexal tumor commonly found on the scalp and face, and often associated with nevus sebaceous, with about half of cases appearing in early childhood. SCAP exhibits cystic invaginations with papillary structures and a double-layered glandular epithelium linked to the epidermal surface and stromal plasma cells. We are reporting a rare instance of intradermal SCAP in a 55-year-old male. He sought evaluation for a long-standing asymptomatic dark-pink papule in his left popliteal fossa, measuring 0.7 x 0.5 x 0.4 cm. A shave biopsy revealed papillary dermal fibrosis, glandular epithelium with apocrine secretion, and papillary projections without an epidermal connection. Infundibulofollicular keratinization was observed, along with stromal plasma cells. The patient chose local excision as the treatment option. This case highlights the rarity of intradermal SCAP, especially in the left popliteal fossa, with only one other reported case in the literature.

**KEYWORDS:** Papillomatous lesions; intradermal; adnexal tumor; syringocystadenoma papilliferum

## INTRODUCTION

SCAP is a benign adnexal tumor with endophytic and exophytic growth patterns. SCAP is often associated with benign adnexal lesions, such as nevus sebaceous and apocrine nevi. It is commonly located on the scalp, followed by the face. Typically, SCAP presents as a solitary gray to dark brown papillomatous plaque ranging from 0.5 to 16 cm. It occurs equally in both sexes between 5 and 65 years old, with half of the cases present at birth or early childhood.<sup>1</sup> Classical histopathological features include cystic invaginations with papillary architecture communicating with the epidermal surface and lined by a double layer of glandular epithelium associated with stromal plasma cells.<sup>2</sup>

Herein, we present a solitary isolated intradermal SCAP in a 55-year-old male at the popliteal fossa.

## CASE REPORT

A 55-year-old male with a medical history of a right eyelid basal cell carcinoma and metastatic lung adenocarcinoma

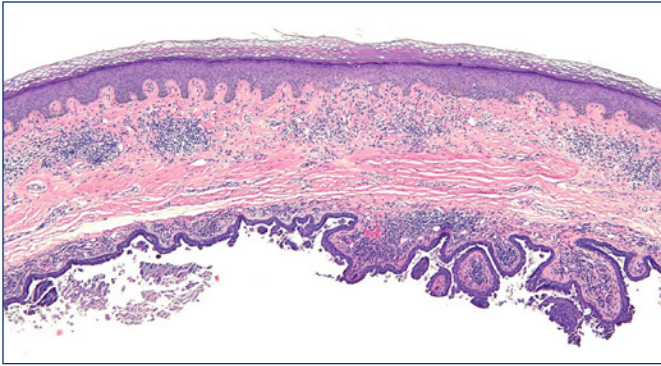
presented with a dark pink papule on his left popliteal fossa. The papule had been asymptotically present for several years and has decreased in size following chemotherapy. He denied any congenital, acquired lesions or trauma in the lesion's area.

Physical examination revealed a 0.7 x 0.5 x 0.4 cm single tan-red to tan-brown papule on the left popliteal fossa (Figures 1,2). A shave biopsy demonstrated papillary dermal fibrosis with a cystic invagination in the mid-dermis lined by glandular epithelium with apocrine secretion and papillary projections. A portion of the cyst lining demonstrated infundibulofollicular keratinization. The stroma contained intervening plasma cells (Figures 3,4). Considering the low potential of malignant transformation of SCAP, the patient agreed to local excision.

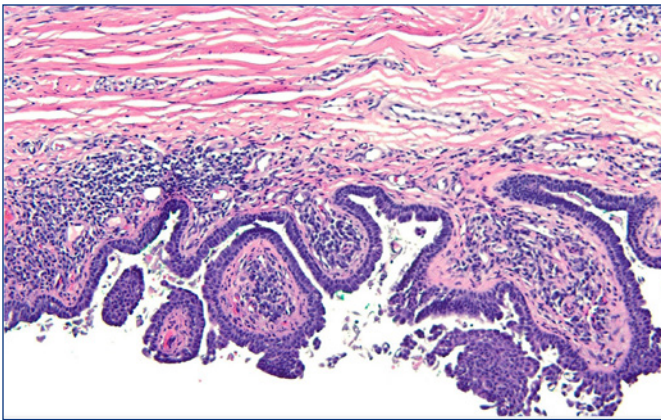
Figures 1,2. Tan-red to tan-brown papule at the left popliteal fossa.



**Figure 3.** Hematoxylin and eosin (H&E) stained shave biopsy of the left popliteal fossa, (40x), showing papillary architectures of the adnexal tumor without epidermal connection.



**Figure 4.** H&E stained shave biopsy of the left popliteal fossa, (100x), showing a polypoid-appearing proliferation of bland-appearing glandular epithelium overlying dermal papillae with an inflammatory infiltrate composed of lymphocytes and numerous plasma cells.



**DISCUSSION**

SCAP is an uncommon skin adnexal tumor of sweat gland origin that mostly requires biopsy for diagnosis. The tumor usually presents as a skin-colored, gray, pink to dark brown, hairless, firm nodule, or papule; however, other clinical morphologies such as verrucous, papillary, hyperkeratotic, or fleshy excrescences have also been described.<sup>1</sup> Occasionally, SCAP shows central umbilication with a small fistula draining serous fluid.<sup>1</sup>

Though the origin of SCAP is uncertain, some studies have suggested a hamartomatous origin that could arise from apocrine glands, eccrine glands, or both undifferentiated pluripotent stem cells.<sup>3-4</sup> A study demonstrated that one-third of SCAP cases develop within a nevus sebaceus, a congenital hamartoma histologically characterized by a complex and abnormal proliferation of epidermal and adnexal structures.<sup>5</sup>

This case is unique for several reasons. Most SCAP cases occur during childhood,<sup>1</sup> yet our patient is an adult. Most of the reported cases in the literature, save for one,<sup>9</sup> have

an observed epidermal connection. While SCAP affects the scalp, followed by the face, our case was on the left popliteal fossa. Of the few reported lower extremities cases (Table 1),<sup>3,4,6,9</sup> only one has been reported intradermal and located on the thigh.<sup>9</sup>

**Table 1.** Reported cases of focal epithelial hyperplasia in individuals of Caribbean descent

| Report                             | Age and Sex | Past Medical History | Location  | Variants                        | Connected to the Epidermis or Intradermal |
|------------------------------------|-------------|----------------------|-----------|---------------------------------|---|
| Malhotra et al, <sup>3</sup> 2009  | 28M         | Not significant      | Thigh     | Liner and segmental             | Connected with the epidermis              |
| Agrawal R et al, <sup>4</sup> 2013 | 24M         | Not significant      | Thigh     | Papules and nodules             | Connected with the epidermis              |
| Vinod K et al, <sup>6</sup> 2013   | 18M         | Not significant      | Lower leg | Multiple papulo-nodular lesions | Connected with the epidermis              |
| Noriko Y et al, <sup>7</sup> 2004  | 26M         | Not significant      | Lower leg | Papule                          | Connected with the epidermis              |
| Kouki C et al, <sup>8</sup> 2022   | 56W         | Not significant      | Lower leg | Warty nodule                    | Connected with the epidermis              |
| Yamamoto et al, <sup>9</sup> 2008  | 46F         | Not significant      | Thigh     | Nodule                          | Intradermal                               |

Abbreviations: F, female; M, male.

Histopathology, SCAP typically reveals cystic invaginations of the infundibular epithelium projecting into the dermis that communicates with the epidermal surface with or without epidermal erosions and serum crust. The superficial aspects are lined by squamous epithelium transitioning to glandular epithelium. These invaginations have papillary architecture lined by glandular epithelium composed of flattened to cuboidal myoepithelial basal layer underneath the luminal layer of columnar cell that shows apocrine morphology. The stroma consists of fibrovascular connecting tissue with numerous plasma cells admixed with some lymphocytes.<sup>10</sup> However, this case demonstrated mid-dermal cystic invagination lined by glandular epithelium with apocrine secretion and papillary projections. Infundibulofollicular keratinization was observed on a portion of the cyst lining and numerous intervening plasma cells within the stroma.

Differential diagnoses for intradermal SCAP are Hidradenoma Papilliferum, which presents at the perineum or vulva of women and lacks plasma cells, and Tubular Apocrine Adenoma, which also lacks plasma cells.

Genetically, a study noted that a subset shows loss of heterozygosity for *PTCH1* (*PTCH*) and *CDKN2A* (p16),<sup>11</sup> suggesting a role for loss of suppressor genes in some cases.

*HRAS* and *BRAF V600* mutations have also been reported in many cases.<sup>12</sup> SCAP is usually curable with a complete surgical excision with a meager rate of recurrence or malignant transformation.

In conclusion, we present a case of intradermal SCAP at the left popliteal fossa in an adult male treated with surgical excision. Cases of intradermal SCAP exceedingly rare, with only one other case reported in the literature.

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