

What Is Your Diagnosis?



A 61-year-old man presented with a 1.4×1.4-cm soft, pink, papillomatous, ulcerated nodule on his left perianal area of 3 years' duration. The lesion would bleed when irritated.

PLEASE TURN TO PAGE 133 FOR DISCUSSION

Christine C. Tam, MD, private practice, Clinton, Maryland.

Allan C. Harrington, MD, Department of Dermatology, Johns Hopkins University, Baltimore, Maryland.

The authors report no conflict of interest.

The Diagnosis: Syringocystadenoma Papilliferum

Syringocystadenoma papilliferum (SCAP) is an uncommon tumor of sweat gland origin.^{1,2} Although approximately half of these tumors are present at birth, they can appear at any time later in life.^{2,3} Syringocystadenoma papilliferum can arise de novo, but in 40% of cases, it is associated with nevus sebaceus.³

The clinical presentation of SCAP is not distinctive. It can present as a solitary patch, papule, plaque, or nodule (Figure 1),² or as several papules often arranged linearly.⁴⁻⁶ Its color varies and may be flesh colored, pink, red, brown, or gray.^{2,4} Its surface may be smooth, hyperkeratotic, verrucous, papillary, or moist and fleshy. The tumors tend to grow slowly and usually are less than 4 cm in size.² Occasionally, they are pedunculated or umbilicated and even exude fluid or keratinous debris from a central pore.^{2,7} Syringocystadenoma papilliferum may be pruritic or ulcerate and bleed with irritation.² It is located on the head and neck 75% of the time but also has been reported on the extremities and trunk, including the groin, buttocks, and gluteal fold.^{2,3,8}

Although its clinical presentation is variable, SCAP can be distinguished histologically by its ductlike structures with irregular papillary

projections, invaginating from the surface epithelium into the underlying dermis, sometimes forming cystic cavities (Figure 2). The ductlike structures are lined by stratified squamous epithelium near the epidermal surface, transitioning to a double-layered epithelium below.² The 2 layers consist of an inner luminal layer of columnar cells with some evidence of decapitation secretion, resting on an outer basal layer of cuboidal cells.^{2,7} The surrounding stroma contains an inflammatory infiltrate composed predominantly of plasma cells (Figure 3). The surface of the lesion may show hyperkeratosis, parakeratosis, acanthosis, and papillomatous changes. Sweat glands usually are present beneath the lesion.² The histogenesis of SCAP is controversial, with evidence supporting apocrine, eccrine, apoeccrine, and pluripotent appendageal cell origin.^{1,2,6}

The differential diagnosis of SCAP is broad. It includes nevus sebaceus, hidradenoma papilliferum, molluscum contagiosum, verruca vulgaris, squamous cell carcinoma, keratoacanthoma, and basal cell carcinoma.

Syringocystadenoma papilliferum is treated by excision.² Rapid growth, pain, ulceration, or bleeding in a long-standing lesion may indicate malignant



Figure 1. Soft, papillomatous, ulcerated nodule on the left perianal area.

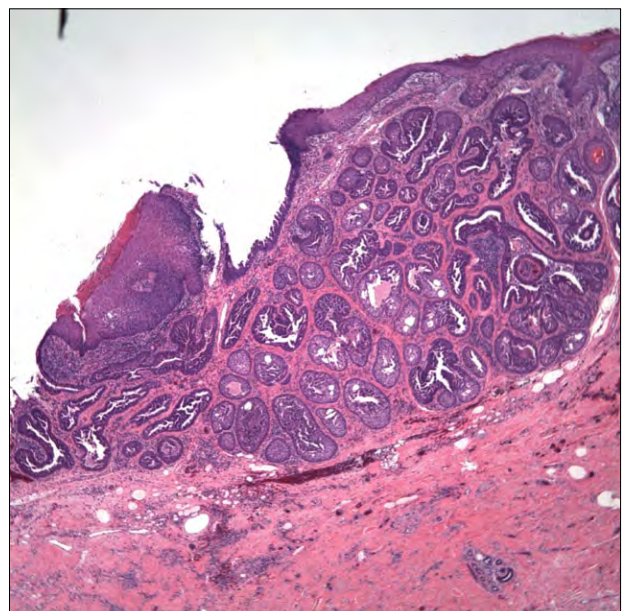


Figure 2. Ductlike structures and cysts (H&E, original magnification $\times 40$).

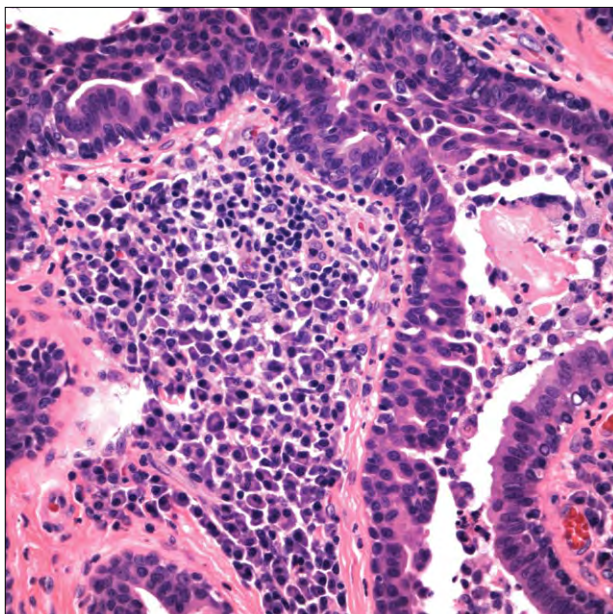


Figure 3. Double-layered epithelial lining of ductlike structures with a dense plasma cell infiltrate in the stroma (H&E, original magnification $\times 400$).

transformation to the rare syringocystadenocarcinoma papilliferum.^{9,10} Cases of SCAP associated with basal cell carcinoma have been reported, but they all coexisted with nevus sebaceus.^{2,3}

REFERENCES

1. Pinkus H. Life history of naevus syringadenomatosus papilliferus. *AMA Arch Derm Syphilol.* 1954;63:305-322.
2. Helwig EB, Hackney VC. Syringadenoma papilliferum; lesions with and without naevus sebaceous and basal cell carcinoma. *AMA Arch Derm.* 1955;71:361-372.
3. Mammino JJ, Vidmar DA. Syringocystadenoma papilliferum. *Int J Dermatol.* 1991;30:763-766.
4. Narang T, De D, Dogra S, et al. Linear papules and nodules on the neck. syringocystadenoma papilliferum (SP). *Arch Dermatol.* 2008;144:1509-1514.
5. Gönül M, Soylu S, Gül U, et al. Linear syringocystadenoma papilliferum of the arm: a rare localization of an uncommon tumour. *Acta Derm Venereol.* 2008;88:528-529.
6. Patterson JW, Straka BF, Wick MR. Linear syringocystadenoma papilliferum of the thigh. *J Am Acad Dermatol.* 2001;45:139-141.
7. Mammino JJ, Vidmar DA. Syringocystadenoma papilliferum: report of an unusual case [letter]. *Int J Dermatol.* 1991;30:828.
8. Brown FS. Pathology quiz case 2: syringocystadenoma papilliferum. *Arch Dermatol.* 1982;118:202-204.
9. Park SH, Shin YM, Shin DH, et al. Syringocystadenocarcinoma papilliferum: a case report. *J Korean Med Sci.* 2007;22:762-765.
10. Chi CC, Tsai RY, Wang SH. Syringocystadenocarcinoma papilliferum: successfully treated with Mohs micrographic surgery. *Dermatol Surg.* 2004;30:468-471.



NEED MORE INFORMATION?

Access this related article in our online archives at www.cutis.com

Microcystic Adnexal Carcinoma Arising Within a Nevus Sebaceus

Use our Advanced Search to find this article and more online!