A 64-Year-Old Woman With Vulvar Papule

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A 64-year-old woman presented with a 1-month history of asymptomatic vulvar papule. There were no adjacent skin changes, no inguinal lymphadenopathy, and no vaginal discharge. The past history was noncontributory. The patient underwent excisional biopsy.

An ellipse of skin 1.0 × 0.8 × 0.5 cm was removed. It contained a centrally raised tan lesion of 0.5 × 0.5 cm. Microscopic examination revealed a well-circumscribed cystic nodule seated in the dermis and connected to the epidermis. The lesion was delineated from the surrounding tissue by a layer of compressed connective tissue. There was no true capsule. The neoplasm had a complex pattern of papillae and tubular structures (Figure 1). The papillae were covered by double layers of epithelium. The superficial layer was composed of columnar cells with basally located nuclei and eosinophilic cytoplasm, and the deeper layer was composed of compressed cuboidal cells. The stroma of the papillae contained numerous plasma cells admixed with a few lymphocytes (Figure 2).

What is your diagnosis?
Pathologic Diagnosis: Vulvar Syringocystadenoma Papilliferum

Syringocystadenoma papilliferum (SP) is a benign cutaneous adnexal tumor with uncertain histogenesis. Investigations using special stains, immunohistochemistry, and electron microscopy have produced variable results; therefore, different authors have postulated differentiation toward eccrine, apocrine, and apoeccrine gland.1 Syringocystadenoma papilliferum is an uncommon skin adnexal tumor. The majority of these tumors occur in the head and neck region and are occasionally associated with nevus sebaceous and basal cell carcinoma. Helwig and Hackney2 suggested that 10% of SP cases develop into basal cell carcinoma. This association is attributed to similar allelic deletions of human homologue of the Drosophila patched gene (PTCH).3 Syringocystadenoma papilliferum has also been reported in other locations, including thigh, breast, and eyelid.1,4,5 However, it has rarely been reported in the female genitalia.6,7 Syringocystadenoma papilliferum sometimes occurs in association with other lesions, including condyloma acuminatum and verrucous carcinoma.8 It usually presents as an asymptomatic solitary nodule and rarely as multiple nodules with linear arrangements. Histologically, SP is characterized by papillary and tubular structures connected to the surface epithelium. The papillae are covered with 2 layers: inner epithelium with eosinophilic cytoplasm and outer myoepithelial cells. The stroma is typically infiltrated by plasma cells and lymphocytes. Complete excision is usually curative.

One differential diagnosis in the female genitalia is hidradenoma papilliferum, which is a benign tumor of apocrine sweat gland that usually presents as a dome-shaped tumor arising in the interlabial sulci and is usually asymptomatic. Histologically, hidradenoma papilliferum is characterized by a complex delicate fibrovascular branching stalk. Unlike SP, it is usually not connected to surface epithelium. Two cell linings cover the stalks: outer epithelial cells and inner myoepithelial cells. Another distinguishing feature is the absence of plasma cells in the papillary stalks.9,10

Syringocystadenoma papilliferum is a benign cutaneous adnexal tumor that rarely occurs in the female genitalia. Because of its association with malignant lesions, it should not be confused with hidradenoma papilliferum, which commonly occurs in this region.

References