Hidradenoma Papilliferum of the Vulva – A Case Report
Smita Pathak, Shruti Desale, Sneha Joshi
Department of Pathology, Maharashtra Institute of Medical Education and Research, Pune, Maharashtra, India

ABSTRACT
Hidradenoma papilliferum is a rare benign cystic and papillary neoplasm arising from apocrine glands. It occurs commonly on the vulval or perianal regions of middle-aged women. The present case is a 36-year-old female patient who presented with an asymptomatic nodule of 2 months duration in the vulval region. Hidradenoma papilliferum characteristically occurs in the anogenital region of women between the ages of 30 and 49 years. Clinically, the neoplasm is a unilateral asymptomatic, skin colored papule or nodule approximately 0.5 cm in diameter. Treatment of choice is local excision. Simple excision is usually curative, however, because malignant transformation has been reported, long-term clinical follow-up is suggested.

Key words: Apocrine, hidradenoma papilliferum, vulva

INTRODUCTION
Hidradenoma papilliferum is a rare benign adnexal tumor considered to be of apocrine origin that occurs almost exclusively in females in the anogenital region.[1] It usually presents as an asymptomatic, flesh-colored nodule, most commonly located on the vulva.[2]

We report a case of 36-year-old female patient who presented with an asymptomatic nodule of 2 months duration in the vulval region.

The case is reported for its rarity.

CASE REPORT
A 36-year-old female patient presented with a nodule on the vulva since 2 months. There was no history of pain, itching, bleeding, or fever. Her general and systemic examination showed no abnormal findings. Local examination showed well circumscribed reddish nodule of size 2.5 cm in diameter on the labia majora. The nodule was firm and non-tender. The differential diagnoses considered were sebaceous cyst or granulomatous lesion.

The hematological, biochemical and serological investigations were within normal limits.

Surgical excision was done, and the specimen was received in the Department of Pathology.

On gross examination, it was a cystic nodule 1.5 cm in diameter. On cut section, it was cystic and showed the presence of papillary projections in the cystic cavity.

Microscopic examination showed a benign cystic lesion with papillary folds, tubules, and cystically dilated spaces. The lumen was lined by inner columnar cells with faintly eosinophilic cytoplasm and outer flattened myoepithelial cell. Decapitation secretion was seen in the lumen. Based on these findings diagnosis of hidradenoma papilliferum was made.

DISCUSSION
Hidradenoma papilliferum was first described by Werth in 1878 for a cystic tumor on the vulva characterized by...
Histopathologically by a cylindrical epithelium that protruded into a cystic space; however at that time; he thought it was an aberration of gland formation and gave no specific name to it. Despite similar lesions had been reported since then, the term Hidradenoma papilliferum was not used until 1943.\(^6\)

Hidradenoma papilliferum is an uncommon benign cystic neoplasm arising from apocrine glands, seen in middle-aged women between the ages of 30 and 49 years.\(^6\)

It is located almost exclusively in the vulvar region, particularly the labium majus. Less frequently hidradenoma papilliferum involves perianal and anal area. Three lesions that are not located in the anogenital region are referred to as ectopic. These ectopic tumors are quite rare and have been reported to occur on the eyelid, face, scalp, external auditory canal, axilla, and back.\(^3,4\)

Clinically, the lesion is usually unilateral and asymptomatic but may be revealed by itching, pain, bleeding, or discharge especially if it ulcerates.\(^9\) It appears as a slow growing, firm dermal papule or nodule, typically smaller than 2 cm.\(^3,6-9\) The lesion is generally well circumscribed and freely mobile under palpation.\(^9\)

Histopathologic examination shows a well circumscribed, cystic lesion in the mid dermis surrounded by a fibrous capsule with no connection to overlying epidermis [Figure 1]. Within the tumor is seen many tubular epithelial structures which are double layered and consist of a basal layer of cuboidal to flat basophilic myoepithelial cells and a luminal layer of columnar cells with oval, pale staining nucleus near the base and evidence of active decapacitation secretion like that seen in apocrine glands.\(^3,5\) [Figures 2-4]. The apocrine nature of the secretion in hidradenoma papilliferum has been established by histochemical, enzyme-histochemical, and electron microscopic examination.\(^4\)

Histochemically the luminal cells contain many large PAS positive diastase resistant granules as seen in the secretory cells of apocrine glands. In addition, luminal cells show positivity for Non-specific esterase and acid phosphatase, the so-called apocrine enzymes. The outer cells stain positively for Alkaline phosphatase as myoepithelial cells do.\(^3,4\)

Immunohistochemical studies demonstrate that epithelial cells lining the papillae express low molecular weight cytokeratin, CEA, EMA, and gross

**Figure 1:** Photomicrograph showing well circumscribed cystic papillary tumor mass in the dermis with no connection to overlying epidermis (H and E ×5)

**Figure 2:** Photomicrograph showing tumor mass composed of tumor cells arranged in papillary and tubular pattern (H and E ×10)

**Figure 3:** Photomicrograph showing tumor mass composed of double layered tubular epithelial structures (H and E 10×)
cystic disease fluid protein 15. The epithelial cells also express strong immunoreactivity for androgen and estrogen receptors.[6]

Electron microscopic examination demonstrates features of luminal cells that are regarded as characteristic for apocrine cells. First numerous membrane limited, secretory granules of varying size and density containing lipid droplets are present in the apical portion of cells. Second is evidence of decapacitation secretions, portions of apical cytoplasm containing large secretory granules that are released into the lumen.[4]

Both the histopathologic and ultrastructural characteristics of hidradenoma papilliferum support an apocrine line of differentiation although some authors have postulated the possibility of origin from Wolffian duct or accessory mammary glands.[5,7]

Hidradenoma papilliferum is a benign neoplasm cured by simple excision. Malignant transformation to adenocarcinoma or adenosquamous carcinoma has been reported very rarely. Hence, long-term clinical follow-up is suggested.[5,6,8]

Our patient was a 36-year-old female who presented with an asymptomatic nodule over vulva since 2 months. The diagnosis was made after surgical excision followed by a histopathologic examination. The patient is on follow-up and has not reported any recurrence till now.

CONCLUSION

When an adult female presents with a nodular lesion in the anogenital region, hidradenoma papilliferum should be kept in mind along with other conditions such as sexually transmitted diseases and other benign and malignant tumors. Diagnosis is based on histopathology. Surgical excision is, therefore, required for identification and definitive cure.

REFERENCES


Source of support: Nil. Conflict of Interest: None