

CASES FOR DIAGNOSIS

Nodular Lesion in the Vulvar Region

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Patient History

A 30-year-old woman consulted for a tumor in the vulvar region (Figure 1) that had appeared 4 months earlier. While it did not cause discomfort, she preferred to have removed.

Physical Examination

The examination revealed a firm, erythematous, well-delimited nodule, not adhered to the deep layers and painless to touch, in the posterior commissure of the labia majora of the vulva. No local or regional lymph-node enlargement was palpable.

Histopathology

The histological study showed a mass located in the dermis with no atypical features or inflammatory infiltrate (Figure 2). Tubular and cystic structures, as well as papillary folds projecting toward the cavity, were observed in the interior of the mass. These structures were composed of a single row of secretory cells and another of small cuboidal myoepithelial cells (Figure 2). The mass was surrounded by a fibrous capsule not connected to the epidermis.

What is your diagnosis?



Figure 1.

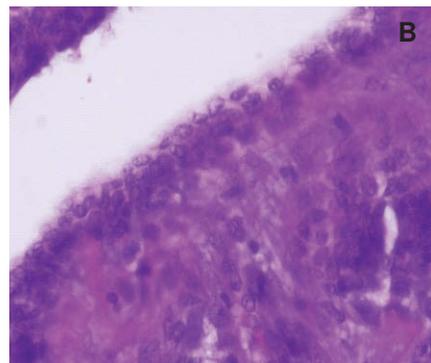
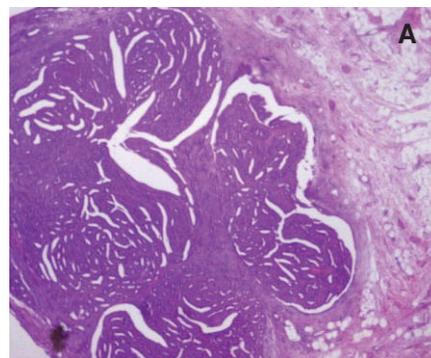


Figure 2.
(A) Hematoxylin-eosin, ×100.
(B) Hematoxylin-eosin, ×400

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Diagnosis

Hidradenoma papilliferum of the vulva

Course and Treatment

Four weeks after resection, the patient remained completely asymptomatic with no evidence of recurrence and was discharged.

Comment

Hidradenoma papilliferum of the vulva, also known as “tubular adenoma of the vulva,” is a rare, solitary, benign neoplasm with apocrine differentiation.¹ It is most commonly found in the vulva, although cases have also been reported in the perianal, nipple,² and eyelid region.³ The condition mainly affects women between 20 and 50 years of age.

The first case was described in 1878 by Werth,⁴ who defined it as a cystic tumor with a cylindrical epithelium that shares some pathological features with papillary adenoma of the breast. In 1941 McDonald⁵ suggested for the first time that this neoplasm could display apocrine secretion.

Clinically, this condition presents as a firm, reddish, well-delimited nodule of similar tone to the surrounding skin that may occasionally ulcerate and bleed. The nodule may also be exacerbated during menstruation, due to the presence of estrogen and progesterone receptors inside the tumor cells, thus suggesting hormonal control. Androgen receptors have also been found, similarly to ductal papilloma of the breast.⁶

In most cases, the lesion is benign, although transformation to adenocarcinoma has been described.⁷ Two reports have described the coexistence of hidradenoma papilliferum with extramammary Paget disease⁸ and vulvar melanoma,⁹ a situation considered by the authors to be incidental.

From the clinical standpoint, the differential diagnosis should consider the following: *a)* Bartholin gland cysts, which are more common in women of reproductive age and characterized by the formation of an abscess within the Bartholin glands, located in the posterior region of the vaginal opening. These cysts are extremely painful, which complicates therapeutic management. *b)* Angiokeratomas of the vulva or angiokeratomas of Fordyce. These are vascular tumors that can affect the vulva and scrotum; they are usually multiple in number and violaceous. *c)* Malignant melanomas, in which the clinical diagnosis is particularly hard to establish when amelanotic.

Histologically, the main differential diagnosis is syringocystadenoma papilliferum, characterized as a

hamartoma and most commonly found on the scalp and face. However, some cases have been described in the female genital region. It is usually present at birth or develops during early childhood, growing after puberty to later become stable. The lesion can ulcerate, bleed, and occasionally take on a verrucous appearance. It has also been described in association with nevus sebaceus of Jadassohn.¹⁰ The anatomical pathology is characterized by a cystic cavity with papillae lined by apocrine secretory epithelium; the papillae are composed of an inner layer of columnar epithelial cells exhibiting decapitation secretion and a second outer layer composed of cuboidal cells. The presence of lymphoplasmacytic infiltrate in the stroma of these papillae is characteristic.

Conflicts of Interest

The authors declare no conflicts of interest.

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