

CASES FOR DIAGNOSIS

Multiple Vulvar Papules

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Case Description

A 46-year-old woman with no personal or family history of interest consulted for asymptomatic vulvar lesions that appeared 4 months earlier. She had not undergone any treatment.

Physical Examination

Both labia majora had multiple whitish, monomorphous papules of cystic appearance and with a diameter of 3 mm (Figure 1). No lesions were observed in the rest of the vulva. A biopsy was taken of one of the lesions (Figure 2).

Histopathology

A histopathological study of one of the lesions showed that the dermis contained a cystic cavity lined by a stratified squamous epithelium with a granular layer (Figure 2).



Figure 1.



Figure 2. Hematoxylin-eosin, ×200.

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What is your diagnosis?

Diagnosis

Multiple vulvar epidermoid cysts

Course

Due to the absence of symptoms, the lack of a request for treatment from the patient, and the benign nature of the lesions, a decision was made to take a conservative approach and refrain from treatment.

Comment

Cysts are extremely common benign cutaneous lesions. Diagnosis is obtained histologically and the lesions are divided into 3 main groups based on the presence or absence of epithelial lining and its type (stratified squamous, stratified nonsquamous, and absence of epithelium). Epidermoid cysts are the most common type and can appear at any site, but most often on the face and trunk. The lesions originate in the follicular infundibulum and, therefore, are also known as infundibular cysts. Microscopic examination shows a cystic cavity filled with keratin layers, lined with stratified squamous epithelium with a granular layer. Milia are small epidermoid cysts, of 1 to 2 mm in size and histologically identical.¹

A literature review showed few published cases of multiple vulvar cysts such as the one we describe. To date, we have only found one case of milia-type multiple epidermoid cysts in the labia majora that appeared spontaneously in a 64-year-old woman.² There have been reports of other types of multiple cysts in the vulva, such as pigmented follicular cysts, steatocystomas, and proliferating trichilemmal cysts.

Pigmented follicular cysts were described in 1982, and only a few cases have been published. They usually present as a single pigmented nodule in the head or neck of adult males. The cysts are lined with stratified squamous epithelium with a granular layer that connects to the epidermis similar to a pore, with hairs observed in the interior. At the time of writing, only one case of multiple follicular cysts in the vulva has been reported, in which a 62-year-old woman had 8 lesions on the labia majora that were treated with CO₂ laser.³

Steatocystoma multiplex is a rare hamartomatous, autosomal dominant disorder of the pilosebaceous unit. It appears during puberty as multiple papules and nodules, mainly on the trunk and upper limbs. The microscopic study shows a cyst surrounded by a fine squamous epithelium with a granular layer that is in turn lined with a fine, irregular, wavy cuticle of eosinophilic appearance. Small sebaceous lobules are found in the cyst wall or

adjacent to it. There have been reports of 2 cases (in patients of age 81 and 60 years) of occasional multiple vulvar steatocystomas that required no treatment.⁴

Proliferating trichilemmal cysts typically appear as nodules in the scalp of elderly women; they are usually benign but may be focally invasive and therefore surgical excision with wide margins is recommended. Histologically, they consist of thick anastomosed bands and nodules of squamous epithelium. This epithelium shows a proliferation of cells with abundant eosinophilic cytoplasm that keratinize abruptly and lead to dense, homogeneous keratin that fills the cystic space. Only one case has been described of a 60-year-old woman with multiple proliferating trichilemmal cysts in the labia majora and minora of the vulva, suprapubic skin, and thigh.⁵ The lesions were flesh-colored papules and nodules between 3 mm and 1 cm that were fast-growing in terms of number and size. Some of them had ulcerated. Treatment was a vulvectomy.

The differential diagnosis for multiple vulvar papules is broad and other entities should be taken into account, in addition to the cystic lesions mentioned above. Although syringomas and Fox-Fordyce disease affect other sites, several cases have been described in which they appear as multiple papules in the vulva.^{6,7} Another 2 conditions, acantholytic dyskeratosis of the vulva and vulvar idiopathic calcinosis, may present as multiple whitish papules, but only a few cases have been published.

Multiple epidermoid cysts in the vulvar area are uncommon. Because the lesions are benign, the treatment will depend on the patient's request. The more uncomfortable lesions can be surgically removed, with CO₂ laser therapy also an option.

Conflicts of Interest

The authors declare no conflicts of interest.

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