Case series of rare nonvenereal vulvar dermatoses

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Abstract

Nonvenereal vulvar diseases such as syringoma and vestibular papillomatosis can be difficult to differentiate from genital warts. Misdiagnosis of these conditions can lead to improper treatment without significant improvement and prolonged suffering. Histopathology may differentiate these conditions from sexually transmitted diseases and help in appropriate treatment. Here, we present case series of four rare vulvar diseases.

Key words: Schwannoma, syringoma, vestibular papillomatosis, vulva

Introduction

Non venereal genital dermatosis may resemble sexually transmitted diseases. Diseases like lichen planus and psoriasis may have different clinical presentation when they occur on the genital area. Vulvul syringoma are associated with intense genital itching while on periorbital location syringoma are usually asymptomatic. Vulval vestibular papillomatosis resemble genital warts in the morphology but they are non-infectious and do not need treatment. Schwannoma is rarely seen in vulvar area may mimic other subcutaneous nodules like sebaceous cyst or lipoma. Role of histopathology is very important in early diagnosis and appropriate treatment of these diseases.

Case Reports

Case 1

A 22-year-old unmarried female presented with multiple raised lesions over genitals associated with severe itching for 1 year. Lesions gradually increased in size and number. Itching was severe, continuous, and was more intense during the menstrual cycle. The patient received multiple treatments in the past with topical steroids and oral antihistamines. There was temporary relief in itching, but there was no improvement in the lesions. Cutaneous examination revealed multiple grouped monomorphic skin-colored papules present over both labia majora with a mild reduction in hair density over the affected area [Figure 1a and b]. There was no evidence of similar lesions around the periorbital area or axillae. Clinical diagnosis of Fox–Fordyce disease or appendageal tumors was considered. Skin biopsy from the papular lesion revealed multiple ductal structures seen in the dermis. Cystic enlargement of two-layered tadpole-like ductal epithelial structures containing eosinophilic material was seen embedded in the fibrous stroma suggesting the diagnosis of syringoma [Figure 2a-c].

The patient was counseled about the benign nature of the disease, but she insisted on permanent treatment for the itching as it was very embarrassing. Lesions were ablated by carbon dioxide (CO_2) laser under topical anesthesia. Considering the risk of scarring, ablation was kept superficial and deeper removal of the lesion was avoided.

After the first session, the patient reported significant improvement in the itching resulting in discontinuation of antihistamines. After complete healing of the ablated lesions, itching subsided completely [Figure 3a and b].

The second session was done 1 month after the first session and the remaining persistent lesions were also ablated similarly. The patient was unavailable for follow-up after the second ablation, but she verbally confirmed complete clearance of lesions without any scarring, dyspigmentation, or recurrence even after 2 years of treatment.

Cases 2 and 3

Two young unmarried sexually inactive females aged 29 years and 19 years presented for evaluation of asymptomatic rough lesions on the vulva for the duration of 1 year and 4 months, respectively. The lesions were gradually increasing in number. There was no history of similar lesions affecting other mucosae. Both the patients were worried about the possibility of infectious and malignant nature of the disease.

Cutaneous examination revealed multiple soft verrucous papillae seen on the vestibular area in the first patient [Figure 4a] and on the labia minora and vestibular area in the second patient [Figure 4b]. The lesions were symmetrically distributed. Skin biopsy from the lesions in both the patient revealed digitate papillomatosis with hyperkeratosis. There was the presence of finger-like projections of loosely arranged connective tissue covered with normal mucosal epithelium, containing multiple blood vessels. There is no evidence of koilocytes or epithelial dysplasia [Figure 5a and b].

Final diagnosis of vulvar vestibular papillomatosis (VVP) was made. The patient was counseled about the benign nature of the disease and that no treatment was required

Case 4

A-32-year old female presented with a minimally painful vulvar swelling of 7 years' duration that was gradually increasing in size. Cutaneous examination revealed a firm mobile subcutaneous nodule over labia majora [Figure 6]. The lesion was excised under local anesthesia [Figure 7]. Excision biopsy revealed a well-encapsulated tumor with thin, wavy nuclei, fibrillary cytoplasm, and nuclear palisading suggesting the diagnosis of schwannoma [Figure 8a and b].

Discussion

Syringoma is a benign adnexal tumor that was first described by Kaposi and Biesiadeki in 1872.[1] Based on histochemical and electron microscopic findings, the cell of origin of this tumor is intraepidermal eccrine sweat glands. It is characterized by multiple asymptomatic skin-colored or slightly yellowish papules. Vulvar syringoma without extragenital involvement is an extremely rare variant of syringoma.[2] It can have varied presentations. Most commonly, they appear symmetrically on the labia majora as multiple flesh-colored or brownish papules. The other presentations include cystic lesions and lichenoid plaques.

Dermoscopy reveals multiple bright yellowish cystic enlargements.[3] Vulvar syringoma should be considered in the differential diagnosis of pruritus vulvae and vulvar papular lesions, such as Fox–Fordyce disease, epidermal cysts, milia, senile angiomas, condyloma acuminata, steatocystoma multiplex, vulvar idiopathic calcinosis, lymphangioma circumscriptum, and lichen simplex chronicus.

Patients with vulvar syringoma may complain of severe itching. Pruritus may become more severe during menstruation, pregnancy, and in summer.[4] Increased

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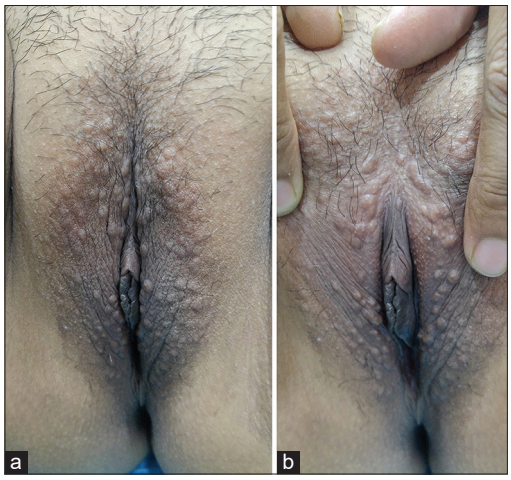

Figure 1 (a and b): Multiple grouped monomorphic skin colored papules present over both labia majora with a mild reduction in hair density over the affected area

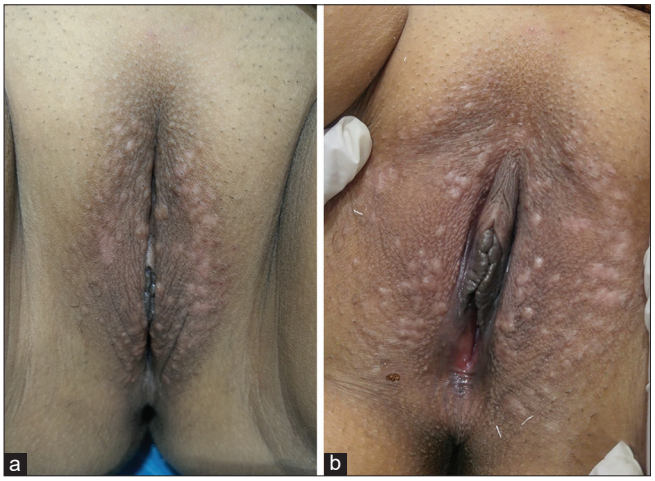

Figure 3: After 1 month of fractional carbon dioxide laser ablation

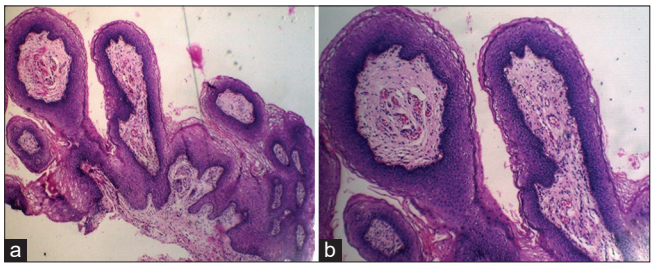

Figure 5: (a) Digitate papillomatosis with hyperkeratosis, finger-like projections of loosely arranged connective tissue covered with normal mucosal epithelium (×40, H and E). (b) Multiple blood vessels with no evidence of koilocytes or epithelial dysplasia (×100, H and E)

pruritus during menstruation and detection of estrogen receptors and progesterone receptors in some patients favored hormonal etiology. The persistence of the lesions may occasionally cause venereophobia and carcinophobia.

The treatment for vulvar syringoma is not standardized. Only a minority of patients achieve adequate control of pruritus with topical corticosteroids, with or without oral antihistamines, tranilast, topical atropine, curettage, cryotherapy, electrosurgery, and resection are some of the

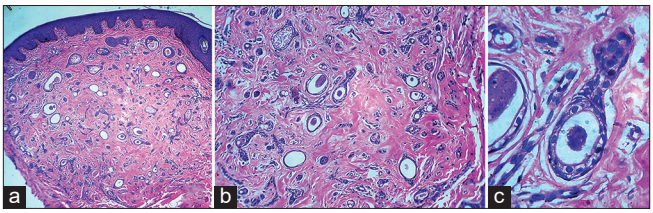

Figure 2: (a) Skin biopsy from the papule revealed multiple ductal structures embedded in fibrous stroma in the dermis (×40, H and E). (b) Ductal structures and cystic enlargement of two-layered (tadpole-like) ductal epithelial structures containing eosinophilic material (×100, H and E) (c) Tadpole-like ductal epithelial structures (×400, H and E)

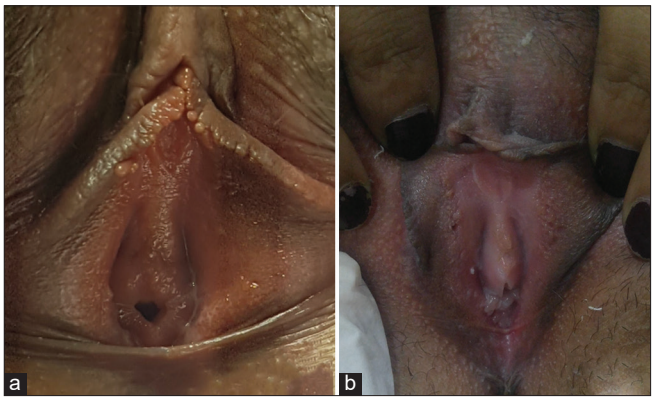

Figure 4: (a) Soft verrucous papillae on vestibular area. (b) Papillae on labia minora and vestibular area

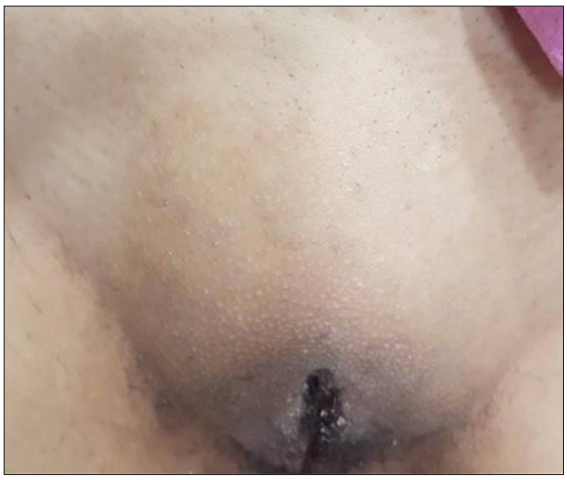

Figure 6: Firm mobile, deep subcutaneous nodule over labia majora, 1.5–2 cm in size

treatments that have been used with variable results.[5] One of the best therapeutic options is carbon dioxide laser treatment, which has proven to be highly effective for the relief of pruritus and for resolving lesions safely and easily.

VVP was first recognized in 1981. Initially, it was named "pseudocondylomata," but other names include vestibular papillae, vestibular microwarts, hirsuties papillaris vulvae, hirsuotid papillomas of vulvae, vulvar squamous papillomatosis, and micropapillomatosis labialis. VVP is a normal variant in the morphology of vulvar architecture and is not related to HPV infection.[6] It is like female counterpart of male pearly penile papules. Clinically, clusters of pink, soft, symmetrically arranged 1–2 mm

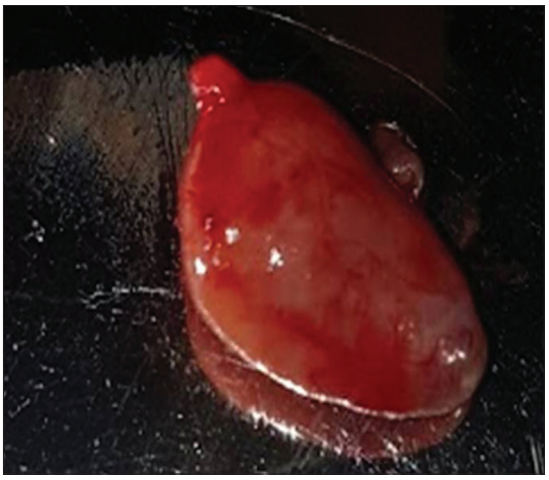

Figure 7: Excised specimen

diameter tubular papillae on inner labia, hymen, or peri urethral area with round tips, separate bases, and lack of circumscribed whitening on 5% acetic acid application are diagnostic of VVP.[7] VVP is mostly asymptomatic; however, vulvar pruritus, pain, and tenderness on vestibular touch, burning, and dyspareunia have been noted in some patients.

Dermoscopy shows profuse and irregular vascular channels in multiple cylindrical filiform projections. The bases of the individual projections remain separate.[8] The histology of this condition is characterized by finger-like protrusions of a loose connective tissue covered by normal vulvar epithelium.

Treatment is mainly counseling regarding benign nature of the condition. However, cryotherapy can be tried for patients with cosmetic concern or those with symptomatic lesions.[9]

Schwannoma (also known as neurilemmoma) is a benign, solitary, encapsulated, and nodular slow-growing neoplasm of the peripheral nerve sheath. They rarely occur in the external genitalia with only few cases of vulvar schwannoma reported. Differential diagnoses for patients with vulvar mass include neurofibroma, epidermoid cyst, labial cyst, lipoma, liposarcoma, fibrosarcoma, angiosarcoma, and Bartholin cyst. Histopathological examination is required for accurate diagnosis and treatment. The histological hallmarks of schwannoma are two types of Antoni areas. Strong immunostaining for S-100 protein distinguishes benign from malignant schwannoma. The conventional variant is the most common type of vulvar schwannoma, but plexiform and ancient variant types also have been reported.[10] Simple surgical excision and follow-up is the treatment of choice and prognosis is generally excellent following the operation.

To summarize, all four cases presented here highlight the importance of histopathology for the early diagnosis and effective management of vulvar dermatoses.

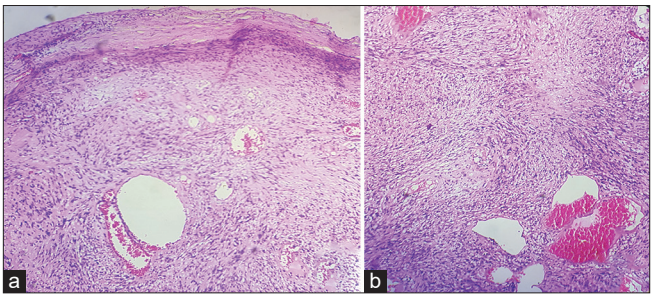

Figure 8: (a) Well encapsulated tumor with thin, wavy nuclei, fibrillary cytoplasm, and nuclear palisading (×40, H and E). (b) Spindle cells arranged in palisading pattern (Antoni A) with Verocay body formation in the deep dermis (×400, H and E)

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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