Photo Quiz

Pruritic papules on the vulva

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CLINICAL FINDINGS

A 49-year-old woman was referred from the gynaecology clinic for the evaluation of vulval pruritus of 6-month duration. She observed periodic exacerbation of itching during menstrual cycles. Previous treatment with topical steroids and antifungal were ineffective in alleviating her symptoms. Genital examination revealed discrete, small (1–3 mm), firm, non-tender, yellowish brown papules on the labia majora bilaterally [Figure 1]. There was no evidence of similar lesions in other body parts.

HISTOPATHOLOGICAL FINDINGS

A punch biopsy from the lesion showed numerous tubular structures with central lumina and a thin epithelial lining. Some ducts possessed small comma-shaped epithelial tails giving a tadpole-like appearance [Figure 2].

WHAT IS YOUR DIAGNOSIS?

Figure 1: Yellowish-brown papules in the labia majora

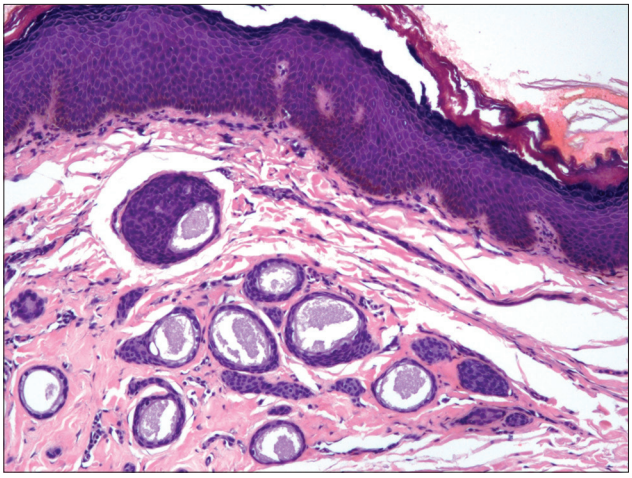

Figure 2: Cystically dilated eccrine glands with interspersed small cell nests and cords (H and E, original magnification ×4)

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Answer: Syringoma

DISCUSSION

Syringomas are common, benign neoplasm that arises from the intraepidermal part of eccrine sweat duct. They are characterized clinically by multiple, small, firm, skin coloured to yellowish papules with a rounded or flat topped surface. They are, usually, localized to the face and are frequently seen in the infraorbital region and cheeks; sometime they may occur in a generalized distribution.[1] Syringomas usually appear during the adolescence period and are more common in women. Their occurrence in the genital region is very rare; syringomas localized to the vulvar region can pose a diagnostic challenge to the clinician.[1]

Vulvar syringoma (VS) are frequently seen in association with extragenital lesions thus making it mandatory to examine the genital region in patients presenting with syringoma over the face or vice versa. Most of the patients affected with VS may not seek medical attention as they are generally asymptomatic.[2] Occasionally, they may cause considerable distress to the patient in the form of pruritus and may affect the quality of life adversely. Huang *et al*. in their series of 18 patients with VS have recorded itching in 72% of patients.[3] The exact cause for itching is not known; itching may be more severe in summer months or during pregnancy or menstruation. It has been suggested that VS are hormone responsive as they may increase in size during pregnancy, premenstrual period or with the use of oral contraceptive pills. Immunohistochemistry studies have yielded conflicting results; few reports have shown the presence of estrogen and progesterone receptors while other reports have failed to confirm this finding.[4,5]

Classically VS present as multiple, flesh coloured to brownish papules on the labia majora. Other clinical variants include white cystic papules ("milium-like") and lichenoid plaques variants.[3] Rarely VS may present with a solitary lesion.[5] Chronic scratching may cause of considerable lichenification of the vulva; hence, VS should be considered in patients who present with lichen simplex chronicus of the vulva.[3]

Vulvar syringoma must be considered in the differential diagnosis of all nodular or papular genital lesions. This includes epidermal cyst, steatocystoma multiplex, lichen simplex chronicus, lymphangioma circumscriptum, senile angioma, early condyloma acuminatum, Fox-Fordyce disease, angiokeratoma, nodular scabies and lichen sclerosus et atrophicus.[1] Histopathological examination will help to arrive at a diagnosis and should be done in all cases.

There is no effective modality of treatment for VS. Topical atropine, tretinoin and oral tranilast have been tried in few patients with success.[6] Physical modalities of treatment like cryotherapy or electro-cautery and surgical excision and full-thickness grafting have been recommended in few reports. Carbon dioxide laser has been found to be very useful in few patients.[3] Tumour recurrence and scar formation following treatment are common; persistence of the lesion may occasionally cause venereophobia or carcinophobia in the affected patients.[6]

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