

## Isolated Vulval Syringoma Presenting as Hyperpigmented Papules: Report of a Rare Presentation

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### Abstract

**Observation:** Syringomas are benign tumours of sweat glands that commonly present over face. Syringomas appear as small, multiple, skin-coloured-to-yellowish papules occurring commonly over upper and lower eyelids, malar areas; infrequently over the axillae, neck, chest, upper arms, and abdomen. The incidence of vulvar syringoma is relatively rare. Syringomas are commonly seen in adolescence. We report a case of isolated vulval syringoma in a middle aged lady presenting with an altered morphology.

### Introduction

Syringomas are benign adnexal tumors arising from the eccrine duct. They mainly affect women during puberty and middle age. Clinically they present as multiple or solitary, localized or generalized, small, skin colored to yellowish papules up to 1-5 mm in diameter [1]. They are characterized by small, firm, rounded, flesh-colored or yellowish papules located on the eyelids, anterior region of the neck, trunk, or abdomen. Vulva is an unusual site for syringoma but may present in association with extragenital sites.

Histology is diagnostic. dermal proliferation composed of cells arranged in nests and ducts within a fibrous stroma are seen. Some ducts present characteristic small, comma-shaped epithelial cell tails that resemble a tadpole. Normally, the ducts are lined with 2 rows of epithelial cells and may be filled with eosinophilic material [2,3]. Response to treatment is usually unsatisfactory. Treatment modalities include topical and intrale-

sional steroids, UV rays, topical retinoic acid, picmrolimus, oral contraceptives and oral retinoids. Electrocautery and surgical excision are other options [4,5].

### Case Report

A 40 year old lady was referred to us by gynaecologist with history of some rashes over her genitalia for last 10 months. She complained of mild pruritus and rapid increase in number of this lesions over this period. On examination multiple hyperpigmented non-tender papules of 2-3 mm size were present over vulva (Figure 1). No similar lesions were found elsewhere in the body. There was no history of premenstrual flare up of the papules. There was no history of bleeding from the lesions. We advised a skin biopsy and it revealed proliferation composed of nests and ducts of epithelial cells embedded in a stroma of collagen bundles. The ducts were lined with 2 layers of cuboid cells and some showed tadpole-like extensions.



**Figure 1 .** Multiple hyperpigmented papules over vulva

**(Figure 2).** A diagnosis of syringoma of vulva was made.

### Discussion

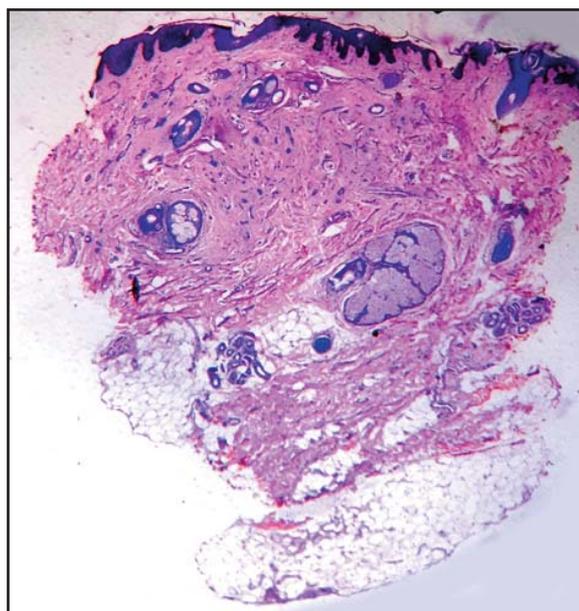
Syringoma was first described in 1872 by *Kaposi* and *Biesiadeki* as lymphangioma tuberosum multiplex [6,7]. Females are commonly affected with adolescence being the most common time of onset. However, lesions can develop later in life ranging between the first and sixth decades of life. The lesions usually are bilateral and symmetrically distributed. .

Vulvar syringoma is a relatively rare occurrence, with only few cases reported in the literature to date [8,9].

Vulvar syringomas are usually associated with extragenital lesions. Hence, its mandatory for the clinician to the rest of the body when syringoma is found in the vulvar region and vice versa. Patients with vulvar lesions commonly present with increasing discomfort and itching, especially in the summer [10]. Based on the observation that there is an increase in size of lesions during puberty, pregnancy, [11] premenstrual period and with the use of oral contraceptives it is suggested that syringomas are hormone responsive.

Vulvar syringomas may be a cause for venophobia [12].

Vulvar syringomas need to be differentiated from epidermal cyst, steatocystoma multiplex, Fox–Fordyce disease, angiokeratomas, senile angioma, condyloma acuminatum and scabies. A biopsy from the lesion with charac-



**Figure 2.** H/P/E in in H/E stain 100x shows the classical tadpole bodies

teristic tadpole bodies confirms the diagnosis. Thus it can be concluded that isolated vulvar syringomas are a rare incidence and is often difficult to diagnose clinically.

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