Images in Clinical Practice

Syringoma masquerading as steatocystoma multiplex

A 35-year-old married female presented with the history of vulvar skin lesions since 20 years. The lesions gradually appeared on vulva when she was 15 years of age. The lesions were asymptomatic in nature. There was no history of similar lesions elsewhere or in any other family member. Clinical examination revealed multiple, discrete, asymptomatic, firm, dark-brown-colored papules and cysts which were present on the labia majora [Figure 1]. The size of the lesions varied from 1 mm to 1 cm. Per speculum examination and the bacteriological swabs from both the vulva and the vagina were normal. Systemic examination was also normal. Complete hemogram and chest radiograph was normal. A clinical differential diagnosis of steatocystoma multiplex, Fox-Fordyce's disease and lymphangioma circumscriptum/lymphangiectasia were considered and one of the lesions was excised for histopathological examination. Histopathology revealed numerous ductules lined by flattened cuboidal epithelium and containing amorphous debris surrounded by hyalinized stroma with the presence of comma shaped appendages [Figure 2]. All these observations were consistent with the clinical diagnosis of syringoma. Immunohistochemistry for estrogen and progesterone receptors could not be performed due to its nonavailability.

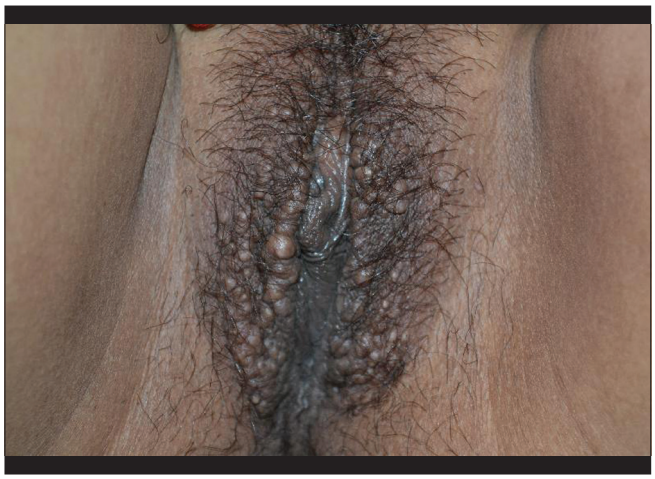

Figure 1: Multiple, discrete, asymptomatic, firm, dark-brown-colored papules and cyst over the labia majora

Syringomas are benign adnexal tumors that occur more commonly in the middle-aged females over the periorbital location. Clinically, syringomas appear as small, multiple, firm, skin colored-to-yellowish papules, 1 to 3 mm in diameter, localized most commonly to the lower eyelids and malar areas, but can also occur in the axillae, neck, chest, upper arms, and abdomen. The lesions usually are bilateral and symmetrically distributed. Syringomas affecting the genital area are rare and may affect the genital areas as part of their generalized occurrence. Atypical syringoma include those which are unilateral, linear, or bathing trunk type in distribution. A large series comprising of 18 patients suffering from vulvar syringoma describes the clinicopathologic and immunohistologic features; a total of six patients also had coexisting periorbital syringomas years before the occurrence of vulval lesions. Our patient had only vulval lesions similar to the earlier cases. The lesions were asymptomaic unlike other cases. We cannot comment on the estrogen or progesterone receptor sensitivity of the lesions in our case. Previous studies have shown the variable expressions of progesterone and estrogen receptors with the positive staining of progesterone receptor marker in eight out of nine extragenital syringoma cases. Vulvar syringoma is

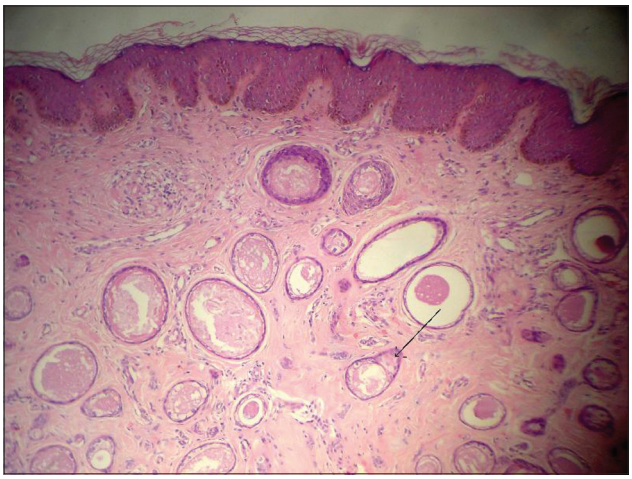

Figure 2: Histopathology showing numerous ductules lined by flattened cuboidal epithelium and containing amorphous debris surrounded by hyalinized stroma with the presence of comma shaped appendages (H and E, ×10)

How to cite this article: Naveen KN, Pai VV, Sori T. Syringoma masquerading as steatocystoma multiplex. Indian J Dermatol Venereol Leprol 2012;78:365-6.

Received: June, 2011. **Accepted:** July, 2011. **Source of Support:** Nil. **Conflict of Interest:** None declared.

not been reported from south India and only one case has been reported from India. In spite of our extensive literature search, nowhere syringoma measuring more than 1 cm in diameter has been mentioned.

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Access this article online	
Quick Response Code:	Website: www.ijdvl.com
	DOI: 10.4103/0378-6323.95458