

Vulvar Syringoma: A Case Report

Vulvar Siringoma

Tülay OKMAN KILIÇ, MD,^a
Mustafa KÜÇÜK, MD,^a
Ömer YALÇIN, MD,^b
Yaşar ÇEĞİL, MD^a

Departments of
^aObstetrics and Gynecology,
^bPathology,
Trakya University Faculty of Medicine,
EDİRNE

Geliş Tarihi/Received: 02.06.2008
Kabul Tarihi/Accepted: 14.07.2008

*This case published as poster at 7th
International Congress of the Turkish-
German Gynecological Association,
2007 in Antalya.*

Yazışma Adresi/Correspondence:
Tülay OKMAN KILIÇ, MD
Trakya University Faculty of Medicine,
Department of Obstetrics and
Gynecology, EDİRNE
ajlankilic@hotmail.com

ABSTRACT Syringomas are benign tumors of the eccrine sweat glands that are most common on the periorbital region. In the English literature, syringoma of the vulva has rarely been reported. In etiology a hormonal influence has been suggested but, our immunohistochemical study revealed no estrogen and progesterone receptors in the tumor cells. We present the case of a 53-year-old woman, asymptomatic, no estrogen and progesterone receptors with vulvar syringoma, and review the literature on this unusual disorder. It is important to include syringoma in the differential diagnosis for papular lesions of the vulva.

Key Words: Vulva, syringoma

ÖZET Siringomalar ektrin ter bezlerinin iyi huylu tümörleridir ve çoğunlukla periorbital bölgede yaygındır. İngilizce literatürde nadir olarak bildirilmiştir. Etyolojide hormonal etki öne sürülmekte ise de, bizim immünohistokimyasal çalışmamızda tümör hücrelerinde östrojen ve progesteron reseptörleri gözlenmemiştir. Bu yazıda 53 yaşında, asemptomatik, östrojen ve progesteron reseptörü içermeyen vulvar siringomalı olgu sunup, bu nadir durum üzerine literatürü gözden geçirdik. Vulvanın papüler lezyonlarının ayırıcı tanısında siringomayı içermesi önemlidir.

Anahtar Kelimeler: Vulva, siyringoma

Türkiye Klinikleri J Gynecol Obst 2008;18:274-276

Syringomas are benign tumors of eccrine sweat glands. They are more common at puberty and characterized by multiple, yellowish-pink color and firm papular lesions especially on the lower eyelid. Syringoma of the vulva has rarely been reported. Most vulvar syringoma are asymptomatic and therefore overlooked. Uncommonly, vulvar syringomas may itchy.^{1,2}

We present the case of a 53-year-old woman, asymptomatic, no estrogen and progesterone receptors with vulvar syringoma, and we reviewed the literature on this unusual disorder.

CASE REPORT

The patient was a 53-year-old gravida 9, para 7 woman who has papular lesions of the vulva. She had complaint of genital lesion for one month. Physi-

cal examination revealed multiple, soft, yellowish-to-skin-colored, 1-2 mm diameter papules, on both sides of labia majora. No similar lesions were defined elsewhere on the body.

A biopsy of the lesion was performed. After overnight fixation in formalin, automatic tissue process was performed for all small polypoid tissues. Hematoxylin and eosin-stained (HE) sections were examined.

Microscopically, the surfaces of all tissues were covered by irregular acanthotic stratified squamous epithelium on the HE slides. In the dermis of each one, the benign tumor was observed next to skin appendages. The tumor was composed of multiple, small and dilated cystic sweat ducts, some of which were characterized by comma-like tails giving them a tadpole shape. Mitotic figures were absent (Figure 1). However, local calcifications and rare giant cell reactions were seen. Diagnose of syringoma was made with these histopathologic findings. Then a commercially available antibodies for estrogen and progesterone receptors were applied to sections using the streptavidin-biotin-peroxidase technique for immunohistochemical analysis.

DISCUSSION

The word syringoma is derived from the Greek syrx, meaning tube or pipe.

Syringomas are common and the most frequently occur on the face. They appear the mostly

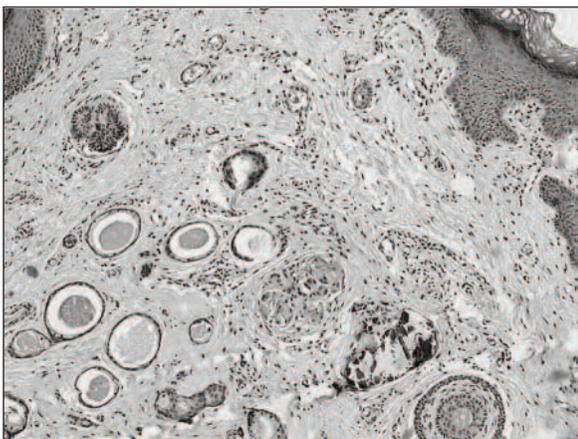


FIGURE 1: Note the dilated cystic sweat ducts, some of which were characterized by comma-like tails giving them a tadpole shape. HE,X50.

in Japanese women and patients with Down syndrome and Ehlers-Danlos syndromes.³⁻⁵

Friedman and Butler proposed a classification consists of four clinical variants of syringoma: a localized form, a familial form, a form associated with Down syndrome, and a generalized form that encompasses multiple and eruptive syringomas.⁶

Familial syringoma is rare and this form appears to be autosomal dominant.⁷ Syringoma associated with Down syndrome can either be localized or generalized.⁸⁻¹⁰ It was showed that the calcinosis also occurred in syringomas associated with particularly Down syndrome.^{11,12} However, Ozcelik et al reported a 34 year old case who has classical lesions associated with calcified vulvar syringoma without Down syndrome.¹³

Eruptive syringomas are a rare variant of syringomas and characterized by numerous lesions on the chest, neck, arms, upper portion of the abdomen, axillae, and periumbilical region at puberty or during childhood.^{4,14-17} Eruptive syringomas usually are bilateral and symmetric.¹⁴ The clear-cell variant of eruptive syringoma is associated with diabetes mellitus.^{8,14} The clear-cell variant is originating from glycogen deposits in the syringoma as a result of phosphorylase deficiency, which occurs in patients with diabetes.^{18,19}

Vulvar syringoma was first described in 1971 by Carneiro et al.³ It often develops just prior to or during puberty, with symptoms of pruritis sometimes correlating with pregnancy or menstruation. Thus a hormonal influence has been postulated²⁰ but, our immunohistochemical studies revealed no estrogen and progesterone receptors in the tumor cells. Huang et al. had reported cases having similar immunohistochemical features.²¹

Syringomas should be considered in the differential diagnosis of pruritic papular lesions of the vulva such as Fox-Fordyce disease, epidermal cyst, senile angioma, cherry hemangioma, lichen simplex chronicus, steatocystoma multiplex, lymphangioma circumscriptum, and condylomata accuminata.²²

Treatment of asymptomatic syringoma is usually not necessary unless they cause cosmetic issues. Up to now reported treatments for symptomatic patients include laser and excisional surgery, cryotherapy, electrodesiccation, some chemical peelings, topical atropine and tretinoin.^{1,2,21-25} All of them have the possibilities of frequent recurrences and postoperative complications such as scarring or pigmentary changes. Park et al. tried a new multiple-drilling method using CO2 laser for 11 patients having syringomas. All patients had good or excellent cosmetic results. No complications, such as scarring, erythema, and pigmentary changes, were observed.

They suggested that the multiple-drilling method by CO2 laser might be an alternative to gain good cosmetic results without complications if applied repeatedly.²⁶ Antihistaminics, topical corticosteroids and atropine may be used for pruritis.^{22,27,28}

Our case has neither Down syndrome nor diabetes mellitus. She has been observed for 6 months and all lesions have been spontaneously regressed. Thus no treatment was necessary.

In conclusion, it is always important to always include syringoma in the differential diagnosis for papular lesions of the vulva.

REFERENCES

1. Belardi MG, Maglione MA, Vighi S, di Paola GR. Syringoma of the vulva. A case report. *J Reprod Med* 1994;39:957-9.
2. Tay YK, Tham SN, Teo R. Localized vulvar syringomas--an unusual cause of pruritus vulvae. *Dermatology* 1996;192:62-3.
3. Carneiro SJ, Gardner HL, Knox JM. Syringoma of the vulva. *Arch Dermatol* 1971;103:494-6.
4. Weiss E, Paez E, Greenberg AS, San Juan E, Fundaminsky M, Helfman TA. Eruptive syringomas associated with milia. *Int J Dermatol* 1995;34:193-5.
5. Soler-Carrillo J, Estrach T, Mascaró JM. Eruptive syringoma: 27 new cases and review of the literature. *J Eur Acad Dermatol Venereol* 2001;15:242-6.
6. Friedman SJ, Butler DF. Syringoma presenting as milia. *J Am Acad Dermatol* 1987;16:310-4.
7. Met Metze D, Wigbels B, Hildebrand A. Familial syringoma: a rare clinical variant. *Hautarzt* 2001;52:1045-8.
8. Patrizi A, Neri I, Marzaduri S, Varotti E, Passarini B. Syringoma: a review of twenty-nine cases. *Acta Derm Venereol* 1998;78:460-2.
9. Schepis C, Siragusa M, Palazzo R, Ragusa RM, Massi G, Fabrizi G. Palpebral syringomas and Down's syndrome. *Dermatology* 1994;189:248-50.
10. Rhodes LE, Verbov JL. Widespread syringomas in Down's syndrome. *Clin Exp Dermatol* 1993;18:333-4.
11. Schepis C, Torre V, Siragusa M, Albiero F, Ciccarello R, Gagliardi ME, et al. Eruptive syringomas with calcium deposits in a young woman with Down's syndrome. *Dermatology* 2001;203:345-7.
12. Maroon M, Tyler W, Marks VJ. Calcinosis cutis associated with syringomas: a transepidermal elimination disorder in a patient with Down syndrome. *J Am Acad Dermatol* 1990;23:372-5.
13. Özçelik S, Altıoğlu Çiğ F, Akyol M, Göze F. Vulvar siringomalı bir olgu. *C. Ü. Tıp Fakültesi Dergisi* 2004;26:91-4.
14. Janniger CK, Brodtkin RH. Eruptive syringomas. *Cutis* 1990;46:247-9.
15. Parker RK, Raque CJ. Eruptive syringoma. *Pediatr Dermatol* 1993;10:91.
16. Teixeira M, Ferreira M, Machado S, Alves R, Selores M. Eruptive syringomas. *Dermatol Online J* 2005;11:34.
17. Seirafi HH, Akhyani M, Naraghi ZS, Mansoori P, Dehkordi HS, Taheri A, et al. Eruptive syringomas. *Dermatol Online J* 2005;11:13.
18. Kitamura K, Muraki R, Tamura N. Clear cell syringoma. *Cutis* 1983;32:169-72.
19. Kudo H, Yonezawa I, Ieki A, Miyachi Y. Generalized eruptive clear-cell syringoma. *Arch Dermatol* 1989;125:1716-7.
20. Wallace ML, Smoller BR. Progesterone receptor positivity supports hormonal control of syringomas. *J J Cutan Pathol* 1995;22:442-5.
21. Huang YH, Chuang YH, Kuo TT, Yang LC, Hong HS. Vulvar syringoma: a clinicopathologic and immunohistologic study of 18 patients and results of treatment. *J Am Acad Dermatol* 2003;48:735-9.
22. Gerdson R, Wenzel J, Uerlich M, Bieber T, Petrow W. Periodic genital pruritus caused by syringoma of the vulva. *Acta Obstet Gynecol Scand* 2002;81:369-70.
23. Sánchez TS, Daudén E, Casas AP, García-Díez A. Eruptive pruritic syringomas: treatment with topical atropine. *J Am Acad Dermatol* 2001;44:148-9.
24. Gómez MI, Pérez B, Azaña JM, Núñez M, Ledo A. Eruptive syringoma: treatment with topical tretinoin. *Dermatology* 1994;189:105-6.
25. Stevenson TR, Swanson NA. Syringoma: removal by electrodesiccation and curettage. *Ann Plast Surg* 1985;15:151-4.
26. Park HJ, Lee DY, Lee JH, Yang JM, Lee ES, Kim WS. The treatment of syringomas by CO(2) laser using a multiple-drilling method. *Dermatol Surg* 2007;33:310-3.
27. Karaoğlu S, Eken A, Kantaş O. Siringoma. *Türkiye Klinikleri J Dermatol* 1994;4:32-4.
28. Solak Tekin N, Altınyazar HC, Doğan Gün B, Koca R, Çınar S. Vulvanın siringoması. *Türkiye Klinikleri J Gynecol Obst* 2003;13:66-8.