Annals of Clinical Case Reports

Case Report
Published: 24 Jan, 2022

Vulvar Syringomas Occurring in a 6-Year-Old Child

Fatima-Zahra Agharbi[1]*, *Fatima Zahra Chahboun*[1], *Marwa Faik*[1], *Ghita Basri*[1], *Abderrahmane Albouzidi*[2] *and Soumiya Chiheb*[1]

[1]Department of Dermatology, Sheikh Khalifa Hospital, Mohammed VI University of Health Sciences, Morocco

[2]Department of Pathology, Sheikh Khalifa Hospital, Mohammed VI University of Health Sciences, Morocco

Abstract

Syringomas or hidradenomas are benign adnexal tumors derived from the deep intraepidermal portion of eccrine sweat ducts. They appear at puberty or in the third to fourth decade. We report an original observation of vulvar syringomas in a 6-year-old girl with familial character.

Keywords: Syringomas; Vulvae; Hidradenomas

Introduction

Syringoma is a benign tumor derived from the excretory duct of the eccrine sweat gland clinically revealed by small papules of a few millimeters in diameter often asymptomatic and frequently localized on the lower eyelids. The vulvar localization that we report is rare.

Observation

A 6-year-old girl, with no notable pathological history, consulted for slightly pruritic vulvar lesions evolving since the age of 4 years (Figure 1). The history revealed the notion of similar lesions on the patient's mother and the older sister but in different location. Dermoscopic examination revealed a homogeneous yellowish area without structures, with linear vessels surrounding it without penetrating inside (Figure 2). A biopsy with histopathological examination of one of the lesions showed glandular structures embedded in a fibrous stroma, associated with cellular trabecula realizing a tadpole appearance. The wall of the ducts was lined by a double row of epithelial cells and containing a colloidal substance (Figure 3).

Discussion

Syringomas are benign cutaneous tumors developed at the expense of the excretory duct of the eccrine sweat glands. They are rarely isolated, often multiple, sometimes even eruptive, and appear in numbers of several dozen in a relatively short period of time [1]. They are more frequent in women and usually appear during puberty, although some lesions may appear later in the fifth or sixth decade [1,2]. They are most often located on the lower eyelid and Malar regions [1,3]. Other sites of predilection are the cheeks, trunk, thighs, armpits and abdomen [3]. Vulvar localization, as in our patient, is less frequent and often unrecognized [3-5]. Clinically, they appear as small papules of a few millimeters in diameter, flesh-colored, clear or pigmented, symmetrically distributed [6]. These papules are most often asymptomatic, but sometimes pruritic, especially on the vulva or during physical exertion. Their size may increase during physical exertion and hot baths, but also during the premenstrual period, during pregnancy and when using oral contraceptives [3,4]. This suggests that syringomas may be hormone-dependent [7]. The presence of familial cases supports

Figure 1: Slightly itchy lesions on the vulva.

OPEN ACCESS

***Correspondence:**
Fatima-Zahra Agharbi, Department of Dermatology, Sheikh Khalifa Hospital, Mohammed VI University of Health Sciences, Casablanca, Morocco, Tel: 0662740825;
E-mail: aghmarifz@gmail.com

Received Date: *01 Dec 2021*
Accepted Date*: 20 Jan 2022*
Published Date*: 24 Jan 2022*

Citation:
Agharbi F-Z, Chahboun FZ, Faik M, Basri G, Albouzidi A, Chiheb S. Vulvar Syringomas Occurring in a 6-Year-Old Child. Ann Clin Case Rep. 2022; 7: 2094.

ISSN: 2474-1655

***Copyright** © 2022 Fatima-Zahra Agharbi. This is an open access article distributed under the Creative Commons Attribution License, which permits unrestricted use, distribution, and reproduction in any medium, provided the original work is properly cited.*

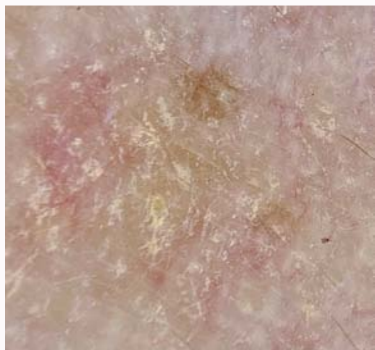

Figure 2: Yellowish lesion without structures, with vessels surrounding it without penetrating to inside.

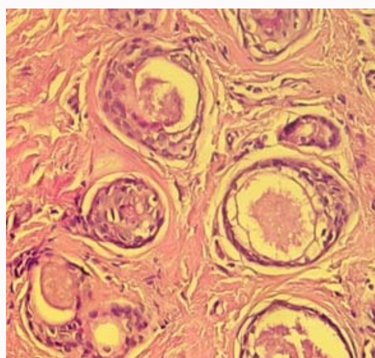

Figure 3: Presence of luminous structures bordered by two cellular foundations associated with small epithelial cords.

a genetic theory, but the pathogenesis of syringomas is poorly understood [1]. The early age of onset in our patient before puberty can be explained by the familial character. Histology shows solid cell cords and small cystic cavities in the dermis, bordered by a double cell base, which sometimes have a small epithelial extension giving the characteristic racket or tadpole appearance [8]. Immunostaining is not helpful. Syringomas express progesterone receptors, which may explain the occurrence of eruptive forms during pregnancy. They specifically express keratin 77 but not keratins 8 and 18 found in the secretory portion of the gland [9]. These benign lesions usually remain stable or may sometimes disappear spontaneously. Treatment of syringomas is usually not necessary and therapeutic abstention is possible. However, because of the aesthetic demand, treatment options include dermabrasion, electrodessication, Carbon Dioxide laser (CO_2 laser) alone or combined with trichloroacetic acid, cryotherapy, and topical retinoids with varying degrees of success. Dermatoscopy of syringomas has been rarely reported in the literature and the most frequently reported dermoscopic sign is a homogeneous, structureless, ivory-white or yellowish area with irregular and ill-defined margins, surrounded in some cases by linear vessels that are poorly branched at the periphery that surround it without penetrating inward as a crown [10].

Conclusion

Our observation is original by two points: the early age of occurrence and the vulvar localization which is rare.

References

1. Lau J, Haber RM. Familial eruptive syringomas: Case report and review of the literature. J Cutan Med Surg 2013;17:84-8.
2. Scherbenske JM, Lupton GP, James WD, Kirkle DB. Vulvar syringomas occurring in a 9-year-old child. J Am Acad Dermatol. 1988;19:575-7.
3. Nibhoria S, Tiwana KK, Yadav A. Vulvar syringoma: A rare case report. J Clin Diag Res. 2014;8:FD06.
4. Dereli T, Turk BG, Kazandi AC. Syringomas of the vulva. Int J Gynaecol Obst. 2007;99:65-6.
5. El Makrini NI, Zaouri H, Benzekri L. Syringomes vulvaires. Annales de dermatologie et de vénéréologie. 2016;143:579-80.
6. 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.
7. Huang Y, Chuang Y, Kuo T. Vulvar syringoma: A clinic pathologic and immunohistologic study of 18 patients and results of treatment. J Am Acad Dermatol. 2003;48:735-9.
8. Bouyahyaoui Y, Meziane M, Bennani A. Syringome éruptif généralisé. Annales de dermatologie et de vénéréologie. 2012;139;508-9.
9. Langbein L, Cribier B, Schirmacher P. New concepts on the histogenesis of eccrine neoplasia from keratin expression in the normal eccrine gland, syringoma and poroma. Br J Dermatol. 2008;159:633-45.
10. Saa SR, Ciancio RM, Cabo H. Estudio dermatoscópico de los siringomas periorbitarios. Med Cutan Iber Lat Am. 2015;43:132-5.