Case Report

Rare sweat gland tumors of vulva: Report of two cases

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Abstract

Syringomas and Fox–Fordyce disease are appendageal skin disorders. While syringomas represent an adenoma of the intraepidermal eccrine duct, Fox Fordyce disease occurs due to blockage of the apocrine sweat duct. In both conditions, extragenital sites are more frequently involved than the genitalia. We herein report two young females, one with syringomas on the face and vulva and the other with Fox Fordyce disease involving axilla, areola and vulva, thereby citing the importance of examination of genitalia in these disorders.

Key words: Fox-Fordyce disease, non-venereal, syringoma

INTRODUCTION

Syringomas are appendageal tumors of the intraepidermal eccrine sweat gland ducts. Their occurrence is more common in women than men, with adolescence being the most common time of onset. However, further lesions can develop later in life and reported cases range between the first and sixth decades of life. 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. Vulvar syringoma is a relatively rare occurrence, with only few cases reported in the literature to date.[1]

Fox–Fordyce disease, also known as apocrine milia, is a rare skin disorder that primarily affects women. The disorder is characterized by intense itching especially in the underarm area, the pubic area and around the nipples. In Fox–Fordyce disease, abnormalities affecting the apocrine sweat glands cause inflammation and enlargement of the glands with characteristic intense itching. Skin near an affected area may become darkened and dry and multiple, small, raised bumps (papules) may develop. Hair follicles in the affected area can become secondarily damaged, resulting in hair loss. The exact cause of Fox–Fordyce disease is unknown.

CASE REPORTS

Case 1

A 25 year old female patient presented with asymptomatic skin colored papules over the face and genitals [Figure 1] for the past 10 years. The lesions commenced over the face and gradually appeared over the genitalia. Patient had history of multiple topical applications without any significant response. Clinical examination revealed multiple discrete skin-colored papules just above the upper lip, malar area and both labia majora. Biopsy from a papule over genitalia showed numerous small ducts lined by two rows of flat epithelial cells. Some ducts possessed small comma like tails of epithelial cells giving a tadpole-like appearance. In addition, there were solid strands of basophilic epithelial cells. Few cystic ductal lumina filled with keratin were also seen near the epidermis [Figure 2]. The above clinical and histopathological picture clinched the diagnosis of syringoma.

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Case 2
A 17 year old female patient presented with itching and skin lesions in both axillae [Figure 3] for the past one year. On examination, axillae showed multiple confluent erythematous papules. Further examination of labia majora [Figure 4] and areola [Figure 5] revealed similar lesions. Biopsy revealed hyperkeratosis with follicular plugging, spongiosis, mild acanthosis, dilated apocrine ducts with periductal lymphocytic infiltration [Figure 6]. Hence a diagnosis of Fox–Fordyce disease was made on clinical grounds based on age, sex, duration, typical sites of lesions and histopathology.

DISCUSSION

Syringoma
Syringoma was first described in 1872 by Kaposi and Biesiadeki as lymphangioma tuberosum multiplex. The clinical presentation is in the form of multiple or solitary, localized or generalized, small, skin colored to yellowish papules up to 1-5 mm in diameter.[2] Vulvar syringomas have been described in association with extragenital lesions. Examination of the rest of the body, especially the eyelids and malar areas, is essential when a suspected syringoma is found in the vulvar region. Likewise, examination of the vulvar skin is mandatory when this lesion is found outside the genital area. Clinically patients with vulvar syringomas may present with increasing discomfort and itching, especially during warmer months.[3] It has been suggested that syringomas are hormonally responsive based on observations that they increase in size during pregnancy,[4] premenstrual period, with the use of oral contraceptives and are known to occur in women during puberty. In a case report by Yorganci et al., immunohistochemistry of vulvar syringoma was shown to display progesterone positivity.[5] Vulvar syringomas with vulvar pruritus has been reported in a 9 year old child.[6] Venerophobia may sometimes be associated with genital lesions.[7] The differential diagnosis for vulvar syringoma is broad, and biopsy with histologic examination may exclude diseases such as epidermal...
cyst, steatocystoma multiplex, lymphangioma circumstriptum, lichen simplex chronicus, Fox–Fordyce disease, angio keratomas, senile angioma, condyloma acuminatum, candidiasis, scabies, pediculosis, allergic and irritant contact dermatitis, psoriasis, and lichen sclerosus et atrophicus.

Biopsy with microscopic examination is the key to establishing the diagnosis. Histologically the epidermis is normal; upper dermis and middermis reveals a plethora of small colloid material-containing cystic ducts and solid epithelial strands contained within the surrounding fibrous stroma of these two layers. At the end of the ducts are often found comma-shaped “tadpole” tails comprising of bulgings formed by weakly organized ductal structures.[8]

Treatment for syringomas is usually not necessary, and is often performed at visible areas of the body for cosmetic reasons, or if it is symptomatic. Treatment with oral tranilast, topical atropine; partial removal by excision, electrodessication, and carbon dioxide laser treatment can be performed with satisfactory results. Although pruritis may resolve, tumor recurrence and scar formation following treatment are common.[1,9]

Fox–Fordyce disease

Fox–Fordyce disease is characterized by firm itchy follicular papules in anatomical sites where apocrine glands occur, namely the axillae, areolae and genitalia. The exact etiology of Fox–Fordyce disease is unknown, but hormonal factors are thought to have a role in its pathogenesis. Emotional and physical stimuli may also play a role in the disease course. The disease occurs mainly in women soon after puberty, but can be postmenopausal. Intense itching occurs in the axillae; sometimes in anogenital region and around the breasts, usually provoked by emotional stimuli. Skin colored or slightly hyperpigmented dome shaped follicular papules develop at a later stage.[10]

The earliest visible change in histopathology is a small vesicle in the apocrine duct. Early changes may be seen most easily in transverse sections.[11] This progresses to an inflammatory lesion followed by rupture and plugging of the duct leading to apocrine sweat retention. Bormate et al contend that perifollicular xanthomatosis (foam cells) is a specific, relatively consistent, and distinct histologic feature in 7 cases.[12] Apocrine acini dilation may be another helpful nonspecific histologic finding.[13] FFD needs to be differentiated from lichen amyloidosis, Darier’s disease, syringoma, lichen simplex chronicus and spongiotic dermatitis clinically or pathologically. The findings of focal spongiosis in upper infundibulum associated with a perifollicular lymphohistiocytic infiltrate can facilitate the diagnosis of FFD.[14]

Response to treatment is often unsatisfactory. Treatment modalities include topical and intralesional steroids (use limited by atrophy), topical clindamycin, UV rays, topical retinoic acid, pimecrolimus, oral contraceptives and oral retinoids. Surgical options include electrocautery, surgical excision of the affected skin and subcutaneous removal of apocrine glands.[10,15]

CONCLUSION

Syringomas are neoplasms demonstrating sweat duct differentiation. Genital syringomas may cause genital pruritus and may be aggravated prementrually and during pregnancy. Fox–Fordyce disease is an apocrine gland milia, occurring mostly in women during adolescence or soon afterward. The axillae and areolae are the primary sites of involvement, but the labia majora, pubes, umbilicus and perineum may be affected. Genital
involvement is rare in both cases, and hence we report these cases.

REFERENCES

1. Miranda JJ, Shahabi S, Salih S, Bahtiyar OM. Vulvar syringoma, report of a case and review of the literature. Yale J Biol Med 2002;75:207-10.
2. Mendiratta V, Harjai B, Gupta T. Vulvar syringomas in an Indian female. Indian J Dermatol 2007;52:158-9.
3. Gerdsen R, Wenzel J, Verlich M, Bieber T, Petrov W. Periodic genital pruritus caused by syringoma of the vulva. Acta Obstet Gynecol Scand 2002;81:369-70.
4. Bal N, Aslan E, Kayaselcuk F, Tarim E, Tunçer I. Vulvar syringoma aggravated by pregnancy. Pathol Oncol Res 2003;9:196-7.
5. Yorganci A, Kale A, Dunder I, Ensar A, Sertcelik A. Vulvar syringoma showing progesterone receptor positivity. Br J Obstet Gynaecol 2000;107:292-4.
6. Garman M, Metry D. Vulvar syringomas in a 9-year-old child with review of the literature. Pediatr Dermatol 2006;23:369-72.
7. Agrawal S, Kulshrestha R, Rajal A, Sidhu S. Localized vulvar syringoma causing vulval pruritus and venerophobia. Australas J Dermatol 2004;45:236-7.
8. 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.
9. Iwao F, Onozuka T, Kawashima T. Vulval syringoma successfully treated with tranilast. Br J Dermatol 2005;153:1228-30.
10. Bologna J, Jorizzo J, Rapini R. Diseases of eccrine and apocrine sweat glands. In: Callen J, Horn T, Mancini A, editors. Dermatology. 2nd ed. Mosby: Elsevier; 2008. p. 547.
11. Shastory ME, Krivda SJ, Tziakiansky GW. Fox Fordyce disease: Diagnosis with transverse histologic sections. J Am Acad Dermatol 2000;42(1 pt):89-91.
12. Bormate AB Jr, Leboit PE, McCalmont TH. Perifollicular xanthomatosis as the hallmark of axillary Fox-Fordyce disease: An evaluation of histopathologic features of 7 cases. Arch Dermatol 2008;144:1020-4.
13. Macareno RS, Garces SJ. Dilatation of apocrine glands. A forgotten but helpful histopathological clue to the diagnosis of axillary Fox-Fordyce disease. Am J Dermatopathol 2009;31:393-7.
14. Kao PH, Hsu CK, Lee JY. Clinicopathological study of Fox-Fordyce disease. J Dermatol 2009;36:485-90.
15. Pock L, Svrckova M, Machackova R, Hercogova J. Pimecrolimus is effective in Fox Fordyce Disease. Int J Dermatol 2006;45:1134-5.

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