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CASE REPORT

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Vulvar syringomas — an underrecognized condition

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ABSTRACT

Syringoma is a benign neoplasm originating from the eccrine ducts of the sweat gland. Lesions are most commonly located on the face. Rarely, do syringomas develop in the vulvar region, where they may be accompanied by persistent pruritus. The article presents the case of a 34-year-old female patient, in whom small nodules and pruritus of the vulva had persisted for approximately two years. The paper includes a description of the dermoscopic features of the skin lesions. The final diagnosis was made based on the histopathological examination. The paper also discusses the most used therapeutic methods in this rare entity.

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Keywords: vulvar syringoma, dermoscopy, dermatoscopy, pruritus vulvae

INTRODUCTION

Syringomas are benign neoplasms of the adnexa, most commonly presenting as multiple skin-coloured nodules located on the face, especially on the lower eyelids, neck, and trunk. Uncommonly, the occurrence of syringomas is limited to the vulva, without involvement of extra-genital locations [1].

CASE REPORT

A 34-year-old woman presented to the Department of Dermatology with a two-year history of periodically itchy lesions in the form of several millimetre nodules located on the labia majora. In addition, the patient reported an occasional, peri-menstrual swelling in the affected area. A dermatological examination revealed the presence of confluent flesh-coloured nodules extending from the posterior conjunctiva of the labia majora to the urethral region (Fig. 1A). Videodermoscopic examination (Canfield D200^{EVO}) showed a reticulated network of thin telangiectatic blood vessels accompanied by multifocal areas of hypopigmentation (Fig. 1B, black arrows) and numerous shiny white clods located in clusters on a cobblestone-like background (Fig. 1B, 1C, blue arrows).

The clinical presentation did not allow for a definite diagnosis. Syringomas, steatocystoma multiplex and amyloidosis were considered in the differential diagnosis. A biopsy was taken for histopathological examination, which showed double-layered ductules, nests and small cysts of epithelial cells without atypia in fibrosing stroma. The final diagnosis of genital syringomas was unequivocally made (Fig. 2). The patient was informed about the benign nature of the condition.

DISCUSSION

Syringomas are benign adnexal tumours originating from the intraepidermal sweat duct of the eccrine gland. They are estimated to affect 1% of the population, with a predilection for women during puberty and the second or third decade of life [2]. Most cases are sporadic, but approximately 11.5% of patients may have a familial variant associated with an autosomal dominant mutation of 16q22 [3]. A higher incidence has also been observed in Down syndrome, Marfan syndrome, Ehlers–Danlos syndrome and in patients with diabetes [2].

Syringomas are generally not accompanied by additional symptoms. However, lesions located in the vulvar region may be susceptible to oestrogens. During the premenstrual phase, during pregnancy, in women using oral contraception, and during the summer months, nodules may tend to be swollen, giving a sensation of change in size. Pruritus may occur, as well [4].

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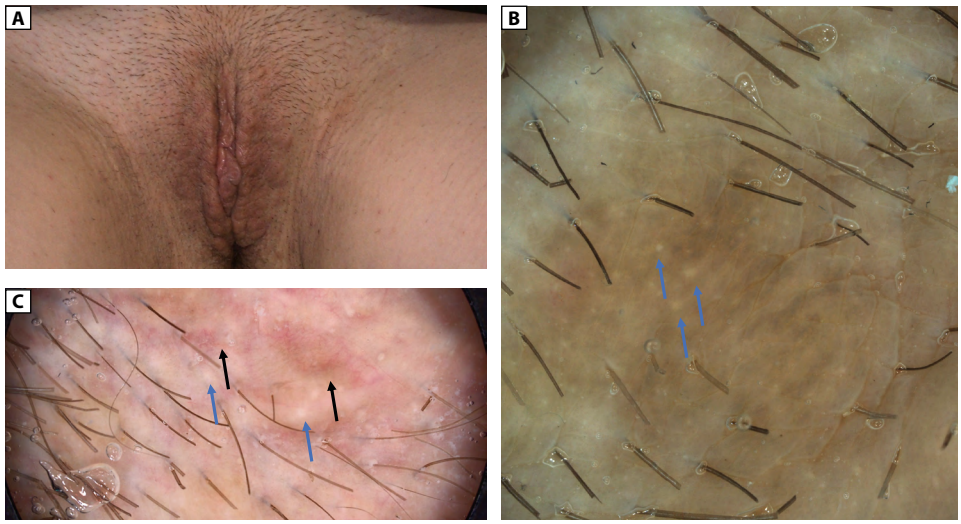

Figure 1. Clinical presentation (**A**); reticulated network of thin telangiectatic blood vessels (black arrows) and shiny white clods (blue arrows) (**B**); shiny white clods (blue arrows) (**C**)

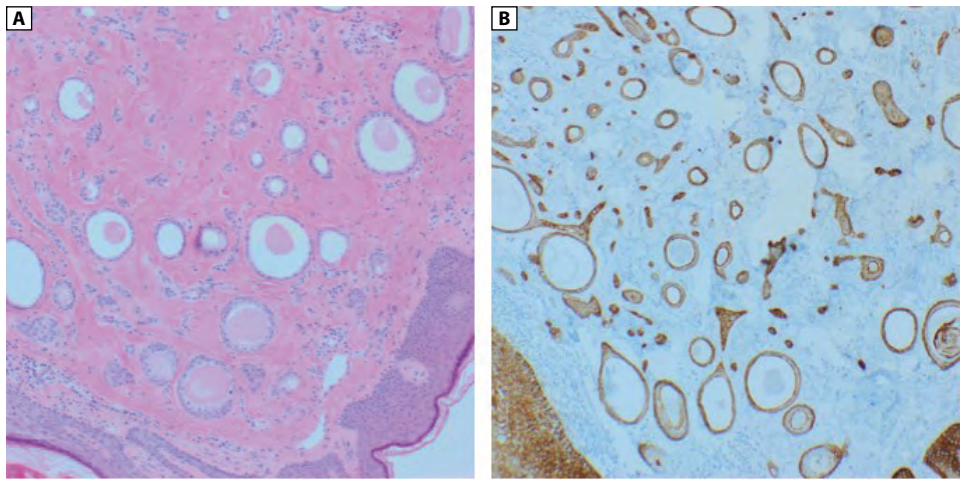

Figure 2. Syringomas — histopathological examination. Haematoxylin and eosin staining, magnification × 100 (**A**); immunohistochemical staining of cytokeratin (CK) 5.6, magnification × 100 (**B**)

Three clinical presentations of vulvar syringoma skin lesions have been identified. The most common presentation is that of multiple flesh-coloured or brownish papules located symmetrically on the labia majora. Cystic lesions or lichen planus-like plaques may also be present [5]. Cutaneous mastocytosis, fibrofolliculomas, velus hair cysts, steatocystoma multiplex, Fox–Fordyce disease, lichen simplex and lichen planus should be considered in the differential diagnosis [6].

Undoubtedly, the unspecific clinical presentation can be confusing both for gynaecologists and dermatologists. An unequivocal diagnosis is made on histopathological examination, which shows a dermal proliferation composed of cells arranged in nests and channels with fibrous stroma. Some of these channels have characteristic small tails of comma-shaped epithelioid cells, resembling tadpoles [1]. An auxiliary non-invasive method is being sought to make a preliminary diagnosis. For this purpose, dermatoscopy has been used, which showed the presence of shiny whitish-yellow oval or round structures on a pink background, histologically corresponding to small colloid deposits in cystic ducts. In addition, a reticulated vascular network consisting of short telangiectatic vessels accompanied by areas of hypopigmentation was reported in the literature [2, 7, 8].

Syringomas are benign tumours, usually asymptomatic. Their treatment is mainly for aesthetic reasons. There are various therapeutic options: cryosurgery, dermabrasion, chemical peels, electrocoagulation, surgical excision and lasers [9]. High-energy devices, such as carbon dioxide

laser, argon laser, and erbium-YAG laser are the most common choices. However, it is important to note that these methods may lead to the development of blisters or leave post-inflammatory hyperpigmentation, scarring, and delayed wound healing [10]. Aksoy Sarc and Onder [11] have also successfully used a yellow laser with a wavelength of 577 nm. Topical therapies reported so far include tretinoin and atropine [12, 13]. For syringomas located in the periorbital region, successful treatment attempts have been made using botulinum toxin type A in monotherapy or combination therapy with carbon dioxide laser or Erb-YAG laser [14, 15]. However, reports of the use of botulinum toxin for vulvar syringomas are lacking in the English-language literature.

CONCLUSIONS

Syringomas should be considered in the differential diagnosis of lesions occurring in the vulvar region. Dermoscopy can be used as an auxiliary method, but a definite diagnosis is made based on histopathological examination. In the authors' opinion, both dermatologists and gynaecologists must be aware of this entity to properly evaluate and treat patients with nodular skin-coloured lesions accompanied by vulvar pruritus.

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Author contributions

All named authors meet the International Committee of Medical Journal Editors (ICMJE) criteria for authorship for this article, take responsibility for the integrity of the work as a whole, and have given their approval for this version to be published.

Conflict of interest

KK, MŻ, and AK declare that they have no conflict of interest. AR has worked as a consultant or speaker for AbbVie, Bioderma, Celgene, Chema Elektromet, Eli Lilly, Galderma, Janssen, Leo Pharma, Medac, Menlo Therapeutics, Novartis, Pierre-Fabre, Sandoz, and Trevi, and participated as Principal Investigator or Subinvestigator in clinical trials sponsored by AbbVie, Drug Delivery Solutions Ltd, Galderma, Genentech, Janssen, Kymab Limited, Leo Pharma, Menlo Therapeutics, MetrioPharm, MSD, Novartis, Pfizer, and Trevi. Adam Reich is a member of the journal's Editorial Board.

Ethics statement

Informed consent was obtained from the patient for participation in the study and publication of the article, including publication of clinical photographs.

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Supplementary material

None.

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