Verrucous and papillomatous plaque on the vulva

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Case study
A previously well Caucasian woman, 59 years of age, presented with a single lesion on her vulva that was noted to be slowly enlarging over several months. The patient was not on any regular medication, had no sexual history of unprotected sex and no family history of similar dermatological conditions.

Physical examination of the vulva revealed a well-delineated, 2 x 1.5 cm whitish-pink verrucous plaque, with a papillomatous and velvety surface of soft consistency and without infiltration (Figure 1).

Full blood count and standard biochemistry tests were normal. An initial diagnosis of condyloma acuminatum (anogenital wart) was suspected given the typical clinical presentation. However, there was no response to podophyllotoxin treatment. A skin punch biopsy was performed and confirmed a histological diagnosis of syringoma (Figure 2). No specific treatment was prescribed and the patient continued periodic clinical review.

Question 1
What are vulval syringomas?

Question 2
What are the clinical signs and symptoms?

Question 3
What are the main differential diagnoses?

Question 4
What is the most appropriate management?

Answer 1
Syringoma is a benign adnexal neoplasm, of apocrine or eccrine origin, with mainly ductal differentiation. Syringomas can appear on any site of the skin surface, although they more frequently involve the periorbital area, particularly the eyelids. Vulval syringomas are an under-diagnosed cutaneous condition in clinical practice as most cases do not present with symptoms and may go unnoticed. Vulval syringomas tend to present during puberty and are more frequent between the third and fourth decade of life. However, they can appear at any age, and rare cases have been described in children and the elderly.

Answer 2
Vulval syringomas may exclusively affect the genital area, or they may be associated with other concurrent syringomas in extragenital sites, mainly the periorbital area. Three different clinical forms of presentation in the vulva have been described:

Figure 1. Whitish-pink plaque with a papillomatous surface on the vulva

Figure 2. Ductal epithelial structures, some of which appeared as tadpole, embedded in fibrous stroma (haematoxylin and eosin x10)
• multiple bilateral and symmetrical small-sized papules (between 1 and 5 mm in diameter), skin tone or brown in colour
• myeloid variant with cyst-like, whitish or yellowish bilateral and symmetrical papules
• bilateral and symmetrical lichenoid plaques, similar in appearance to lichen simplex chronicus.

Answer 3
Clinical diagnosis of vulval syringomas is usually difficult because of the non-specific appearance of the lesions. When dealing with vulval papules or plaques, several differential diagnoses should be considered:
• epidermal cysts
• milia
• idiopathic vulvar calcinosis
• steatocystoma multiplex
• condylomata acuminata (anogenital wart)
• lymphangioma circumscripta
• Fox-Fordyce disease
• angiokeratomas
• soft fibromas
• desmoplastic trichoepithelioma
• eccrine epithelioma
• lichen planus
• lichen simplex chronicus. Hence, biopsy and histological examination are mandatory to confirm the definitive diagnosis. Vulval syringomas must be considered in the differential diagnosis of chronic vulval pruritus.1–5

Key points
• Vulval syringomas have variable clinical presentations.
• They should be suspected if a verrucous or papillomatous plaque does not respond to standard treatments for condyloma acuminatum (anogenital wart).
• Skin biopsy and histological study is necessary to confirm the diagnosis.
• No treatment is necessary, but it may be performed in symptomatic cases or for cosmetic reasons.
• Therapeutic options have variable outcomes and a high relapse rate.

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Competing interests: None
Provenance and peer review: Not commissioned, externally peer reviewed.

Acknowledgements
We thank José Luis Agud and Fernando Burgos for their reviews and editing of the manuscript.

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