

# Plaque-type vulval syringomas presenting with vulval pruritis

Dr. David Wright<sup>1</sup>, Dr. Katharine Warburton<sup>1</sup>

1. Leeds Centre for Dermatology, Leeds Teaching Hospitals NHS Trust

## Introduction

Syringomas are benign intraepidermal eccrine sweat-gland tumours more commonly seen in women, usually presenting in puberty although they can occur at any age.

The most frequently involved sites are the lower eyelids and malar regions, but they can present anywhere on the skin.

Cases of vulval syringomas have been rarely reported, describing localised 1-3mm vulval papules. More extensive plaque-like variants are recognised extra-genitally, but have not been reported in vulval skin.

## Case History

- Well 30 year old para 0 gravida 0 female
- Presented with a 10-year history of progressive textural change to both labia majora associated with pruritis.
- Reported pre-menstrual flaring of her symptoms.
- No improvement with super-potent topical steroids or Ovrnette (combined oral contraceptive pill).
- Differentials considered were atypical lichen simplex chronicus, extramammary Paget's disease or adnexal tumours.

## Examination findings

Bilateral, symmetrical, plaque-type textural change to the labia majora.

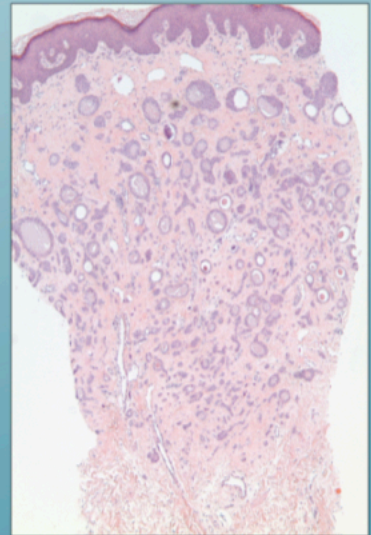
Labia minora, vaginal introitus and perineum were unaffected.

Full cutaneous examination revealed no extra-genital or facial lesions.



## Histology

Biopsy of a representative area showed dermal proliferation of multiple small ducts lined by a bi-layered cuboidal epithelium, with comma shaped extensions, typical of a syringoma. The clinical features suggested a unifying diagnosis of plaque-type syringomas.



## Discussion

- Four suggested clinical variants of syringomas include localized, generalised, Down's Syndrome-associated and a familial form<sup>1</sup>. Less common variants include lichen planus-like, milium-like and plaque-type syringomas.
- Plaque-type syringomas are most commonly seen on the face. Cases of these mimicking other tumours or found as collision lesions with basal cell carcinomas have been reported, providing intraoperative challenges during MOHs micrographic surgery<sup>2</sup>.
- The symptomatic correlation with menstruation has been described with other cases of vulval syringoma, and in a separate case during pregnancy, suggesting a hormonal relationship. This is further supported by cases with positive immunohistochemical staining for oestrogen and progesterone receptors<sup>3</sup>.
- There are reports of varied success with topical steroids, tretinoin and atropine; oral isotretinoin; and destructive methods including surgical excision, electrodesiccation and curettage, cryotherapy and laser. Treatment for vulval plaque-type syringomas poses a difficult challenge with risk of significant scarring particularly from the destructive therapeutic options, where use has only been reported in discrete vulval lesions. Our patient has opted to implement simple vulval skin care. She declined a trial of progesterone only contraceptive pill to address the potential hormonal causal relationship.

## Key Summary Points

- The prevalence of vulva syringoma may be under recognised given that many presentations are asymptomatic and not recognised by the physician. They should be considered in the differential diagnosis of vulva papular lesions and those presenting with vulva pruritis.
- Although recognised extra-genitally, this is the first case reported of the plaque-type variant bilaterally on the vulva.
- Most syringomas do not require treatment as they are asymptomatic. Vulvar syringomas presenting with pruritis have varied success with medical therapy, but the benefits seen in some cases with surgical options need to be outweighed against the risk of scarring, recurrence and sometimes persistent pruritis despite removal.

## References

1. Friedman, S.J. and Butler, D.F. Syringomas presenting as milia. *J.Am.Acad.Dermatol.* 1987, 16:310-314.
2. Yang, Yi BBA, MPA; Srivastava, Divya. Plaque-Type Syringoma Coexisting With Basal Cell Carcinoma. *Dermatologic Surgery*; November 2018 - Volume 44 - Issue 11 - p 1464-1466
3. Wallace, M.L. and Smoller. B.R. Progesterone receptor positivity supports hormonal control of syringomas. *J. Cutan. Pathol.* 1995, 22:442-445.