RESEARCH LETTER

Acquired lymphangioma circumscriptum of the vulva secondary to severe herpes simplex infection

ABSTRACT

A 43-year-old woman presented to genito-urinary medicine with skin-coloured, papillomatous papules on the mons pubis and superior labia majora (figure 1). Routine sexually transmitted infection screening was normal. Given the papillomatous appearance, a diagnosis of genital warts was made but treatment with cryotherapy and immiquimod cream was unsuccessful.

Further history revealed that subtle vulval papular changes first appeared 6 years previously, following a severe primary genital herpes simplex virus (HSV) type 1 infection, confirmed by PCR, which had caused extreme pain and marked swelling. Treatment with oral acyclovir was delayed by 7 days as the patient was abroad, where symptomatic relief alone was provided. Further episodes of HSV occurred every 2–3 months over the following 5 years, but antiviral therapy was not initiated on each occasion.

Over the subsequent years, during which the patient underwent two caesarean sections, the papules enlarged and occasional vulval skin redness, swelling and oozing of clear fluid occurred. The patient was referred to dermatology whose differential diagnosis included lichen simplex and allergic contact dermatitis. Patch testing was negative. Vulval skin biopsies were performed.

Histology showed dilated lymphoid channels in the superficial dermis, leading to the diagnosis of acquired lymphangioma circumscriptum (LC), a condition due to local lymphatic disruption. Investigations into underlying causes, including faecal calprotectin, colonoscopy and transvaginal ultrasound, were normal.

Treatment included self-lymphatic drainage techniques and supportive underwear. Intermittent shave excision of some lymphangiomas provided temporary symptomatic relief. Increasingly, the LC became complicated by cellulitis requiring therapeutic and prophylactic antibiotics. Prophylactic 400 mg twice daily acyclovir successfully prevented further HSV outbreaks.

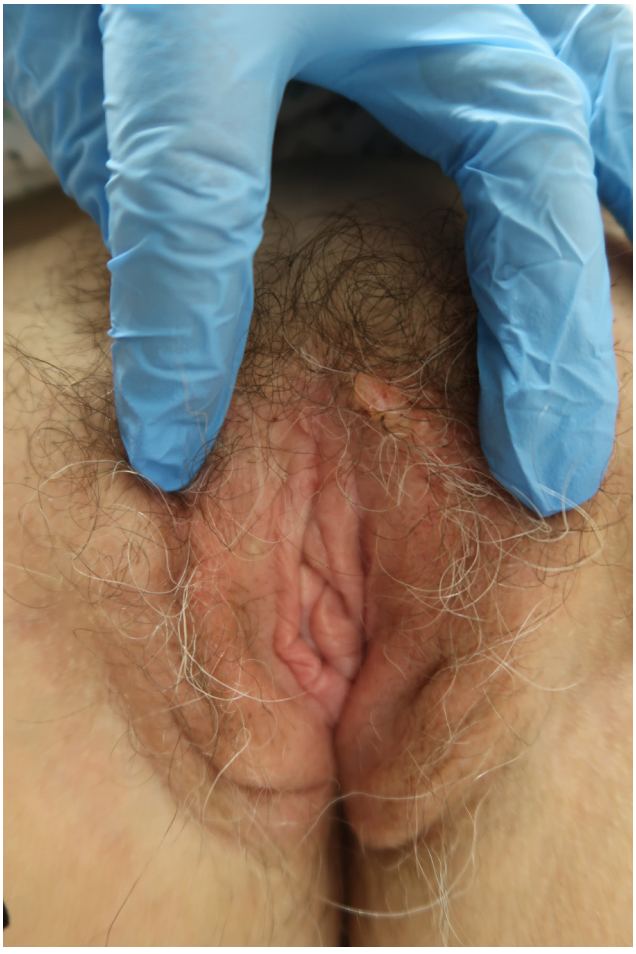

Figure 1 Skin-coloured papules on the superior labia majora. The papules gradually enlarged and were intermittently complicated by lymphorrhoea.

Acquired LC, also called acquired lymphangiectasia, is a rare, benign condition caused by local lymphatic vessel disruption. It typically affects vulval skin, but can also affect the scrotum. It presents with multiple, skin-coloured papules, caused by dilation of cutaneous lymphatic vessels, and may be papillomatous and hyperkeratotic.

Due to the papillomatous nature, LC is frequently misdiagnosed as genital warts[1–3] causing delays in appropriate management; correct diagnosis requires vulval skin biopsy. Acquired LC may be asymptomatic, painful or pruritic. Cellulitis, lymphorrhoea and lymphoedema are common complications. Dyspareunia and psychosexual difficulties can also arise.

Investigations to identify an underlying cause of acquired LC are crucial; reported underlying causes include Crohns disease, local tumours, radiation, surgery and infections such as erysipelas, lymphogranuloma venereum or tuberculosis.[4] In 2016, a literature review by Chang *et al* found that the most common underlying cause was a local tumour, predominantly cervical. Therapeutic options include curettage and cautery, surgical excision, laser, radiotherapy and conservative supportive care.

Given the timing of onset and lack of other underlying pathology in our case, we felt that lymphatic damage probably occurred during initial HSV infection, an association that has not previously been reported, and worsened with recurrent HSV episodes and caesarean sections. Prompt HSV treatment and appropriate prophylaxis may help to prevent acquired LC as a complication of genital HSV. Diagnosis of acquired LC should be considered in women presenting with vulval papules particularly if non-responsive to wart treatment or associated with lymphorrhoea or cellulitis.

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Handling editor Anna Maria Geretti

Funding The authors have not declared a specific grant for this research from any funding agency in the public, commercial or not-for-profit sectors.

Competing interests None declared.

Patient consent for publication Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

To cite Callander JA, Davies BM, Hill G. *Sex Transm Infect* 2020;**96**:233–234.

Received 2 August 2019
Revised 22 October 2019
Accepted 7 November 2019
Published Online First 22 November 2019

Sex Transm Infect 2020;**96**:233–234.
doi:10.1136/sextrans-2019-054224

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