

## Family history repeats itself

### Pain and swelling triggered by sexual intercourse had a genetic basis

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#### Case notes

A 25-year-old woman, gravida 0, who had an 18-month history of severe swelling of the labia minora during and after intercourse was examined. This was extremely painful and associated with intense pelvic pressure and urinary urgency during intercourse only. Swelling usually resolved approximately 12 hours later. The patient also experienced unwanted persistent genital arousal.

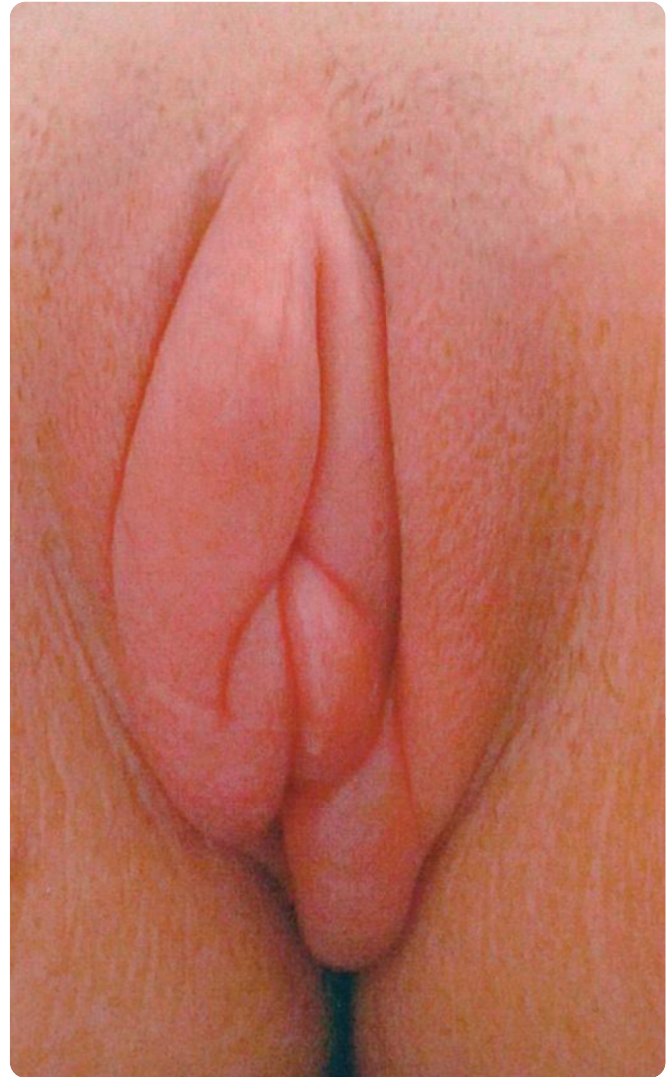
She reported that, before the previous 18 months, intercourse had been symptom-free, and she could not identify any new inciting factors. Her medical history was unremarkable. On physical examination, her external genitalia were exceedingly edematous and tender (Figure 1). A grade 2 cystocele and hypertonicity and tenderness of the levator ani muscles were identified. The uterus and cervix were unremarkable with no apical prolapse. No urinary incontinence was noted with cough.

#### Conclusions

The patient received a diagnosis of intermittent edema of the labia minora, hypertonic pelvic floor muscle dysfunction, and persistent genital arousal disorder. In addition, the patient was referred to a specialist in adult genetics for evaluation of suspected Ehlers-Danlos Syndrome (EDS). In a focused review of systems during the genetics consultation, she reported easy bruising, joint pain, and costochondritis. Her family history revealed a fatal brain aneurysm at age 53 in her maternal grandfather and hyperextensible skin in her mother and brother.

Physical examination showed joint hypermobility with a Beighton score of 5 and hyperextensibility of the skin, which are findings that, combined with a positive family history, led to a diagnosis of EDS, hypermobility type, or EDS type III.<sup>1</sup> The patient was referred for intravaginal pelvic floor physical

FIGURE 1



Severe vulvar edema occurred during and after sexual intercourse.

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therapy, a specialized form of rehabilitation that involves myofascial trigger-point release, mobilization, and massage, that helped to decrease the severity and frequency of labial edema (Figure 2). Treatment with sertraline was initiated to alleviate persistent genital arousal.

EDS hypermobility type is an underreported collagen disorder that may have gynecologic manifestations. Patients with hypermobility syndrome exhibit an increased ratio of type III collagen to type I collagen, which causes increased tissue laxity

FIGURE 2



The patient's vulva had a normal appearance when edema was not present.

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and, subsequently, joint hypermobility and organ prolapse.<sup>2</sup> Abnormal connective tissue in pelvic floor ligaments and fascia is a proposed cause of pelvic floor prolapse.<sup>3</sup> These disorders may cause additional vulvovaginal symptoms that have not yet been described in the literature.

Labial edema and possibly persistent genital arousal may be attributed to venous insufficiency that is associated with EDS. Patients with excessively compliant connective tissue may show increased distensibility of dependent veins that predisposes patients with EDS to excessive pooling of blood. In the genital region, engorgement raises the pressure inside the capillaries and increases transudation of plasma through the epithelium, which leads to fullness and lubrication of the vaginal epithelium and labial edema.<sup>4</sup>

Intermittent profound labial edema should prompt the evaluation of joint hypermobility. Collagen abnormalities might explain uncommon, yet significant, gynecologic presentations such as pelvic organ prolapse in young women and persistent sexual arousal. Because EDS may have other serious sequelae, such as valvular heart disease and aneurysm, it is important for clinicians to identify those at risk. ■

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