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Case Report

Surgical management of a persistent lichen sclerosus case and reconstruction of vulva with free rotation and V-Y advancement flaps

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Abstract. Lichen sclerosus (LS) is a lymphocyte-mediated, chronic, inflammatory dermatitis which most commonly involves the anogenital area. Treatment options include topical corticosteroids, tacrolimus, carbon dioxide laser ablation and, as a last resort vulvectomy. The objective of this study is to report a refractory anogenital LS case and describe the use of a V-Y advancement flap for reconstruction of the perineal defect after vulvectomy.

A case of persistent LS, treated succesfully with surgery is presented and the literature is reviewed. The patient tolerated the surgical procedure well and a satisfactory cosmetic result was obtained.

A multidisciplinary team is ideal in the management of patients with persistent LS. Surgical management of LS should be reserved as a last resort for patients who are refractory to other medical treatment options and, in the case of a surgical attempt, techniques for reconstruction of the vulva should be well known.

Key words: Lichen sclerosus, vulvectomy, vulvar reconstruction

1. Introduction

Lichen sclerosus (LS) is a benign, chronic skin disease that most commonly occurs in the anogenital epithelium (1). It affects both genders, children and adolescents and is especially prevalent in women in postmenopausal ages. Although the exact etiology is still unknown, there is a strong association between LS and autoimmune disorders, such as alopecia areata, vitiligo, thyroid disorders and diabetes mellitus (2). Besides the autoimmune diseases in association with HLA class II antigens, hormonal factors, infectious causes, and genetic influence have also been associated with LS (1).

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Classic LS is characterized by marked inflammation, epithelial thinning and distinctive dermal changes with the appearance of fine "cigarette paper-like" wrinkling accompanied by symptoms of pruritus, dyspareunia and dysuria (3,4). A considerable number of patients (33%) are asymptomatic but have signs of LS on physical examination. The diagnosis of LS is based upon the presence of characteristic clinical manifestations, ideally with histological confirmation obtained from a vulvar punch biopsy. A delay in the diagnosis may arise due to patients' embarrassment and/or reluctance of the physician to fully evaluate the symptoms, as well as unfamiliarity with the disease (5).

Optimal management of LS can be achieved with a multidisciplinary approach. Surgical treatment is reserved as a last resort, for example in cases of post-inflammatory sequelae, phimosis or in malignant states. Although the presence of vulvar scars or other characteristics of the defect can affect surgical preference, there are many surgical approaches proposed for perineal reconstruction, including local muscle and fasciocutaneous skin flaps (6-8). We present a

patient with persistent lichen sclerosis, who underwent surgical management reconstruction of the vulva with V-Y advancement flaps.

2. Case report

A 49-year-old gravida 3, para 2, abortion 1 Caucasian woman, has been married for 30 years and suffering from vulvodynia was admitted to our outpatient unit due to persistent vulvar LS for 16 years. Review of her medical history revealed, two cesarean sections, carbamezepine use for restless leg syndrome for the last six years, and three lumbar disc hernia operations, with placement of an internal fixator to the lumbar vertebrae.

She had undergone treatment with local hydrocortisone, estrogen and testosterone since her initial diagnosis of LS. She had one vulvar alcohol injection, and several courses of vulvar betametasone, lidocaine injections and phototherapy. Her history also revealed a vulvar denervation operation of the pudental nerve for her diagnosis of vulvodynia. Moreover, she had undergone three simple vulvectomies with vulvar reconstruction. These treatment modalities failed to alleviate her symptoms, and the sclerosing changes of the vulvar area extended posteriorly, involving the perineum and anus.

Remarkable findings from her pelvic examination included hyperkerotic lichen plaques spreading to the anus, vaginal orifice and periclitoral area (Figure 1a). The vaginal orifice was narrowed, and the labia majora and minora were absent. Hyperkeratotic cicatrices were noticeable on her mons pubis, due to her previous operations and lichen plaques. On speculum examination, the vagina and cervix appeared normal, and the uterus and bilateral ovaries were normal on transvaginal ultrasound. No malignant cells were reported in her pap-smear test. The histopathological diagnosis of LS was confirmed with two previous sequential vulvar biopsies, in both of which the characteristic epidermal atrophy and dermal hyalinization of the upper dermis (homogenization of the collagen) was reported.

All treatment options were discussed in detail with the patient, and simple vulvectomy with reconstruction of the vulvar defect with V-Y advancement flap (Figure 1a) was decided upon, the patient's persistent symptoms. Informed consent form was obtained from the family. Briefly, the surgical procedure was started with urinary catheter placement to prevent urethral injury, and in order to provide wound hygiene after surgery. Simple vulvectomy (Figure 1b) followed, with V-shape incisions extending to medial portion of the thigh bilateraly, without complication (Figure 1c). To move the flaps freely, deep incisions were made to the level of the superficial genital fascia, with meticulous attention to hemostasis. Subsequently, the mobility of the cutaneous flaps was tested. Following the release of each flap, the excess block of subcutaneous tissue was folded into the defect. The lateral transverse defect, formed when the island flap was moved medially and closed horizontally, thus giving a "horizontal Y shape" to the final scar. We aproximated the subcutaneous tissue with 2/0 polyglactin 910 sutures [Ethicon, Edinburgh, UK], and the skin was closed by interrupted 3/0 polyglactin 910 sutures [Ethicon, Edinburgh, UK] (Figure 1d).

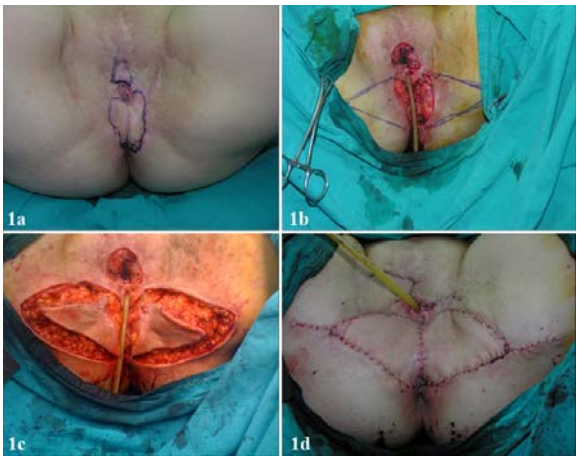

Fig. 1. Pre- and intraoperative pictures of the vulvar lichen sclerosus patient. 1a; hyperkerotic lichen plaques spread to anus, vaginal orifice and periclitoral regions, 1b; vulvectomy was performed, 1c; V-shape incisions extending to medial portion of the thigh bilaterally, 1d. closure of the skin, giving the final scar a horizontal "Y" shape.

The circular defect above the vaginal orifice was repaired with a free rotation flap. No drain was used. Repair of the vulvar defect was statisfactory. The urinary catheter was left in place for two days (Figure 2a), and she was discharged on her third postoperative day, with weekly follow-up. Vulvar healing was uneventful in the following days and completed by the third postoperative week (Figure 2b). We prescribed a high potency topical steroid (clobetasol propionate, Dermovate %0.05 cream; GlaxoSmithKline) nightly following the third postoperative week for eight weeks, in order to prevent reactivation of disease.

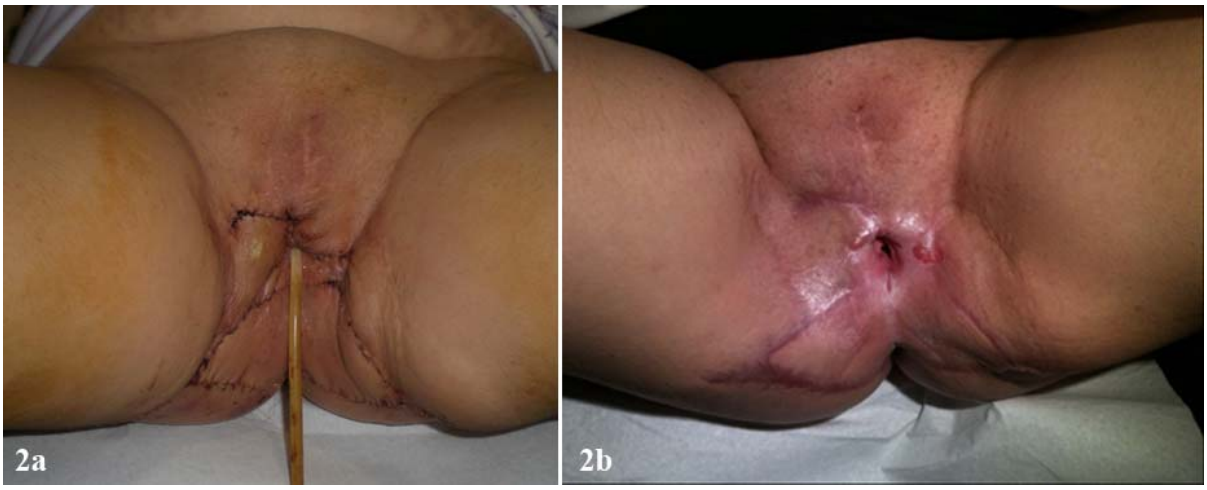

Fig. 2. Postoperative pictures of the vulvar lichen sclerosus patient. 2a; second post operative second day, 2b; completed vulvar healing three weeks after surgery.

3. Discussion

A variety of names and descriptions have been used for the disease that is currently named "lichen sclerosus". In 1887, Hallopeau was the first who described the histological features of the disease (9). LS occurs at all ages except neonates, and is very rare in the first year of life. The disease has a bimodal peak incidence in prepubertal girls and menopausal women. In one study, the prevalence of LS in childhood was found to be 1:900. The majority of LS patients were women between 50-70 years old, while 5-15% of the LS patients were children.

Lifelong surveillance of all women with LS is considered essential because of the risk of malignant progression (10). Although women with vulvar LS are at increased risk of developing invasive squamous cell cancer (SCC) of the vulva (4), the estimated risk is thought to be less than 5% (11). No evidence currently exists that regular check-ups reduce the risk of malignant progression, nevertheless, patients with vulvar LS should be examined at least once a year, and localized, persistent, non-resolving lesions should be biopsied (4). In contrast to vulvar LS, extragenital LS lesions are not associated with an increased risk of malignancy (9,12).

There is not a universally accepted management strategy for women with LS. Most treatments provide symptomatic relief, without necessarily correcting the underlying disorder. The treatment of LS consists of education and support, behavioral modification to maintain good perineal hygiene and avoidance of any local irritants, medication and surgery. Various conservative therapeutic options, such as topical testosterone, progesterone, local corticosteroids, estrogens, retinoic acid, retinoids, vitamin A, chloroquine and a short course of systemic corticosteroids, are all acceptable treatment options (13).

All published data on the management of LS indicate the use of moderate to strong topical steroids is the treatment of choice (12,14,15). Mild to moderate potency topical corticosteroids are also commonly used for treatment of adult vulvar LS (3). Recently, super potent topical steroids were found to be an effective treatment option with both short- (16) and long-term (17) efficacy. However topical steroid therapy is not without complications, and includes the possibility of atrophy, contact sensitization, skin changes, and secondary infection.

Although there are no well studied therapies for women who fail to respond to corticosteroids, progesterone, testosterone, retinoids, topical immune system modulating agents (tacrolimus, pimecrolimus) and cyclosporine have been used in some randomized trials (4). Small randomized trials have been found that progesterone and testosterone creams are less effective than corticosteroids (clobetasol), with more side effects (18). Treatment with an oral retinoid (acitretin) was found to be effective in one randomized trial (19). Successful treatment with tacrolimus ointment was reported (20), but it was often discontinued due to burns on application sites. As presented in our case study, refractory subjects have been treated with intradermal alcohol injections with variable results (21). Another concern is psychological problems that may appear (i.e. narcotic abuse) in persistent LS patients, due to chronic vulvar pain. The

American College of Obstetricians and Gynecologists suggests annual examinations for patients whose LS is well controlled, and advises more frequent visits for those with poorly controlled disease (15).

Surgery does not play a key role in the treatment strategy of LS, and should be considered a last resort to treat complications secondary to LS (11,22,23). Moreover, cicatrice and contracture formation after surgery limit its use. Surgical intervention in LS should not be aimed removing the disease but ıt should be aimed resolving complications of the disease, for example to release a buried clitoris, to separate fused labia, or to widen a narrowed introitus in the case of pain or sexual dysfunction (22). If surgery is the preferred treatment modality, it is important to know how to reconstruct the vulva. Patients usually wish to continue their sexual life as soon as possible. At this point V-Y advancement flap is an effective method for the reconstruction of perineal region. This technique will provide better blood supply and nerve sensation and will allow the expansion of vaginal orifice.

The rationale behind surgical therapy is primarily to treat those patients who did not respond or responded poorly to medical treatment, and secondarily to prevent the development of invasive carcinoma of the vulva (23). Our patient had a very long history of LS, for which she had received several local therapies, phototherapy, systemic medications and surgical attempts over the course of 16 years. Although she hadn't received any premalignant or malignant diagnose from her previous vulvar biopsies, she preferred to undergo another surgery, due to her exhaustion and expectation of a better cosmetic result. Ultimately, her course postoperatively was uneventful, without any recurrence of LS at one year, and she was happy with the cosmetic result. Sexual intercourse resumed three months after surgery, without any annoyance, confirming the success of our surgery.

The data about the recurrence rate of LS after surgery is still limited. In a current review by Gurumurthy et al. (24) improvement of symptoms was reported in 80% of women in a 10-year follow-up period after Fenton's procedure (median perineotomy) and laser division of adhesions in 25 patients. This high success rate in their study was likely due to carefully selected LS cases for surgery (i.e. patients with LS complicated by adhesions) (24). Rouzier et al. (25) reported an 86% success rate in 64 LS women following perineoplasty with a median 34 months follow-up. However, as recently mentioned in a clinical guideline; studies on the surgical management of LS are limited (26), and there is still need for further investigation.

Consequently, surgical intervention should be deferred until LS has been controlled with medication, and should be reserved for the management of postinflammatory sequelae or malignancy. If an excisional procedure (i.e. vulvectomy) is performed, reconstruction of the vulvar defect with V-Y advancement flap seems to be an applicable and highly effective surgical technique, with good cosmetic results and rapid healing after surgery.

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