DERMATOLOGICAL VULVOVAGINAL CONDITIONS CAUSING PAIN

Infections
- Herpes simplex infection
- Group A streptococcal cellulitis

Inflammatory conditions where the dominant symptom is pain
- Lichen planus
- Desquamative inflammatory vulvovaginitis
- Chronic fixed drug eruption
- Immunobullous disease
- Nonspecific acute genital ulceration
- Hidradenitis suppurativa
- Crohn's disease of the vulva
- Behçet's disease
- Hailey–Hailey disease
- Drug reactions

Inflammatory conditions where the dominant symptom is itch
- Dermatitis
- Psoriasis
- Lichen sclerosus
- Chronic vulvovaginal candidiasis

Neoplasia
- Eroded extramammary Paget's disease
- Ulcerated VIN or SCC

Other conditions
- Atrophy
- Chronic idiopathic fissures
- Scar tissue
- Vulval varicosities

ABBREVIATIONS: SCC = squamous cell carcinoma; VIN = vulvar intraepithelial neoplasia.

TABLE. DIFFERENCES BETWEEN LESIONAL AND NONLESIONAL PAIN

Lesional pain	Nonlesional pain
Pain and itch	Pain, no itch
Pain is described as sharp like a cut	Pain is described as burning and other dysaesthesias
Worse with walking	Improved with walking
Worse at night (in bed)	Improved with rest
Usually bilateral	May be unilateral
Well localised	Poorly localised
Normalising the lesion usually improves the pain	If there is a lesion, symptoms are unchanged when vulva normalises

covered the aetiology, pathophysiology and clinical presentation of vulvovaginal pain.[1]

PAIN WITH AN OBSERVABLE LESION

Vulvovaginal pain is usually caused by infections and inflammatory dermatological conditions that result in inflammation, ulceration, blisters, fissures and adhesions (Box). Less commonly, tumours, benign and malignant, cause the pain.

The three most common vulval dermatoses – dermatitis, psoriasis and lichen sclerosus – usually cause itch. Pain may also occur, due to excoriation from scratching or fissuring. Dyspareunia, if present, is usually due to friction causing raw areas.

Chronic vulvovaginal candidiasis (CVVC) is also principally an itchy condition but pain often occurs, particularly with tampon insertion or intercourse. CVVC characteristically flares premenstrually, and improves during menses. Examination may be normal in the first two weeks of the menstrual cycle.

Dermatological conditions and infections that are predominantly painful rather than itchy are uncommon, and are listed in the Box.

Vulval varicose veins are another possible cause of vulvovaginal pain. They can cause a dull ache, particularly after long periods of standing.

Patients with vulval skin conditions usually have observable signs. However, recognising these signs involves knowing what is normal and abnormal for a patient's age, hormonal status and racial group, and then the performing of a thorough examination that includes stretching the skin to look for subtle fissures. A speculum examination is essential to rule out vaginal epithelial causes, but because of the pain this may not be possible at the first visit. Many genital dermatoses are episodic, and thus it may be necessary to examine the patient on a day when she is most symptomatic.

PAIN WITH NO OBSERVABLE LESION

In a patient presenting with chronic pain and its variants (burning, stinging, 'awareness', throbbing, stabbing, formication and sometimes itch) who has a genuinely normal physical examination, evidence is increasing that the most likely explanation is neuropathy, referred pain and/or pelvic musculoskeletal dysfunction.[2,3] The source of this neuromuscular pain may be visceral pelvic problems such as prolapse and constipation, or spinal disease or dysfunction, or both. An important practice point is that nonlesional pain can follow an episode of painful vulvovaginal skin disease (for example, genital herpes or vaginal candidiasis) and persist after the skin problem has been resolved. Even though there has been a historical dermatological trigger for the pain, such patients should be included in this group.

A difficulty in making a diagnosis of nonlesional pain is that some common dermatological conditions causing pain may have subtle, easily missed examination findings. For example, it should always be assumed that fissuring is caused by a dermatosis, even if no rash is seen. In addition, many common dermatoses have observable

examination findings that are intermittent; the classic examples are chronic vulvovaginal candidiasis and vulval psoriasis, both of which tend to be most easily recognisable premenstrually. If a skin disease is present, it should always be assumed to be the cause of the vulvovaginal pain until proven otherwise.

Occasionally, patients with psychiatric conditions can experience vulvovaginal pain as a symptom of their condition (somatoform disorder). Our experience is that pain due primarily to psychiatric disease is not more common in the vulvovaginal region than in any other part of the body.

HISTORY TAKING AND DIAGNOSIS

The features of lesional and nonlesional vulvovaginal pain noted on history taking are discussed in detail in part 1 of this article, as is the physical examination and investigation of women with such pain.[1] A summary of the differences between lesional and nonlesional pain is given in the Table, and an approach to the diagnosis and treatment of vulvovaginal pain is outlined in the flowchart.

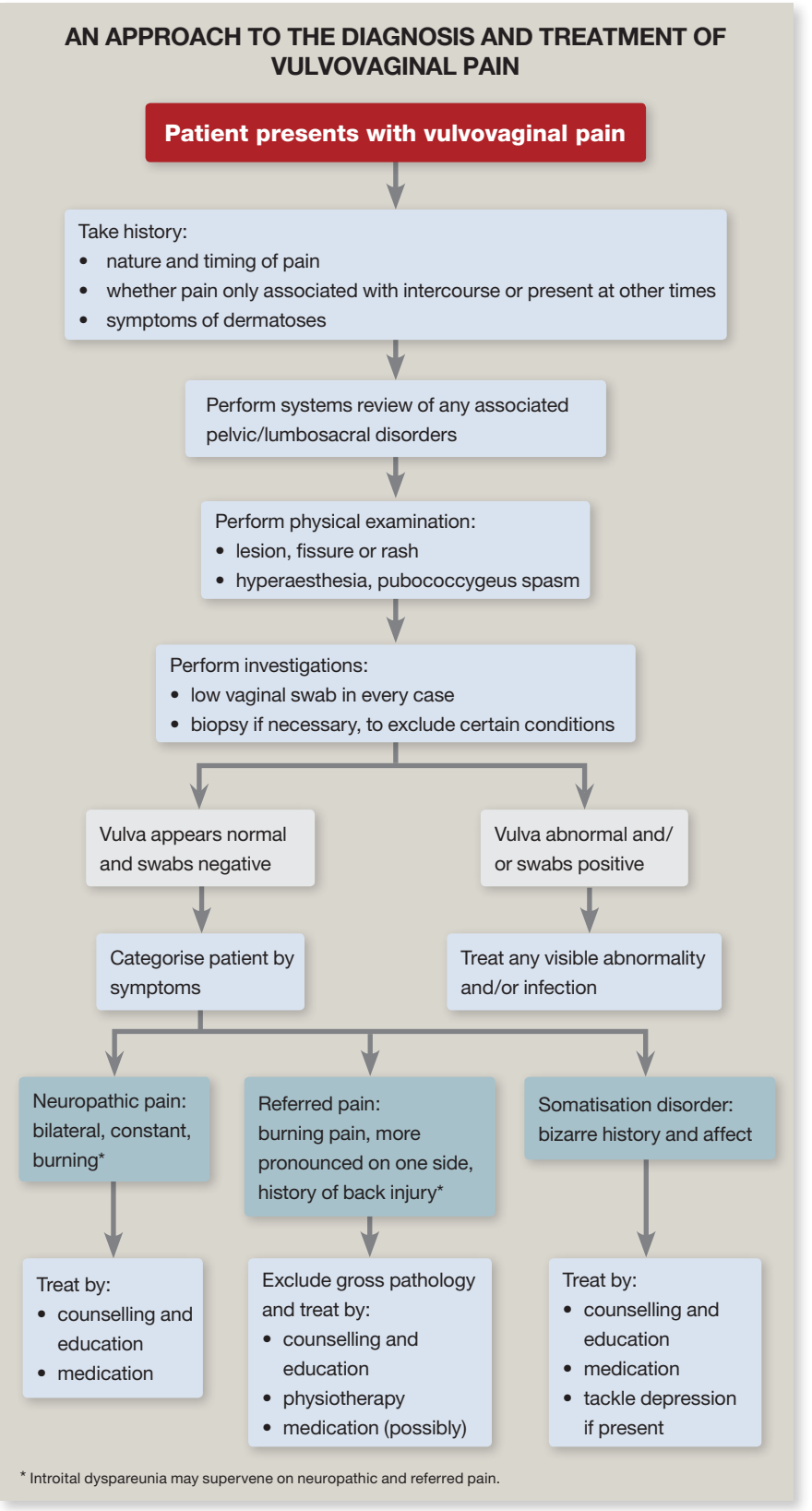
MANAGEMENT OF NONLESIONAL VULVOVAGINAL PAIN

Exclude dermatological or infective causes

Before assuming that a patient has nonlesional pain, ensure that their vulvovaginal examination is truly normal (see the flowchart). If seeing them on a 'good day' and the examination seems normal, reschedule an examination on a 'bad day'. A low vaginal swab should always be taken for culture to exclude candidiasis, even if the examination does not suggest it.

There are patients with observable skin disease in whom the skin disease is not responsible for their symptoms. Nonetheless, a trial of treatment for the observed skin condition must be undertaken to ensure that it has been confidently excluded as a cause for the pain.

Some patients have pain due to coexisting lesional and nonlesional causes. In these



cases, adequate control of an observable vulval dermatosis may not completely improve the pain, and a nonlesional cause should be sought.

Categorise patients

Although it is often challenging, we find it helpful to attempt to categorise patients into those likely to have neuropathic pain, those with referred or musculoskeletal pain and those who have a somatoform disorder. The latter is a diagnosis of exclusion.

The appropriate management of patients in these diagnostic groups is illustrated by the three cases below and summarised in the flowchart.

CASE 1: NEUROPATHIC PAIN

Case scenario

Nora is a 72-year-old woman who complains of a constant and poorly localised burning sensation in the vulva. The pain is least severe in the morning after rest in bed and builds up during the day, especially with physical activity. Certain positions are more uncomfortable than others, particularly sitting.

The pain is not always associated with dyspareunia, but she has developed a distaste for sexual intercourse because she is fearful that it will exacerbate the pain. The burning sensation is not severe, although she finds it debilitating. It rarely wakes her at night but she finds that it is constant and exhausting. She relates that she has 'good and bad' days and that bad days occur after increased stress or physical activity.

When questioned, she reports that she has chronic low back pain, and that the vulval burning started shortly after a lumbar discectomy for sciatica.

Adequate trials of topical antifungal and corticosteroid preparations did not help her, and in fact caused stinging. She self-medicates daily with topical oestrogen cream, which does not sting but is also ineffective.

Vulval examination shows no abnormality other than some degree of atrophy consistent with her age, and vague erythema of both labia majora. Palpation reveals hyperalgesia but no pubococcygeus spasm.

Comment

Nora presents with a typical history and examination for a patient with neuropathic pain. Although atrophy is present, this has been excluded as a cause of her discomfort by an adequate trial of topical oestrogen. The erythema may relate to loss of sympathetic control of skin vasculature and can also be a side effect of daily use of oestrogen and/or topical corticosteroid cream.

Neuropathic pain commonly localises to a horseshoe-shape involving the perineum, introitus and lower labia majora bilaterally. It may radiate to the groin or inner thighs. In some patients the discomfort is unilateral, or even confined to one small spot, usually on the mucosal surface of the labia minora; in others, the area affected is periclitoral.

Management – general advice and therapeutic interventions

Before recommending medication to a patient with neuropathic pain, it is essential to provide an explanation. Many patients present having seen many doctors and tried many treatments without success, and are understandably frustrated and sceptical. Many state that they do not wish to be 'pill-poppers', as if taking medication indicates a psychological weakness. Many have deduced from experience that the problem is perhaps 'all in their heads', and reasonably resist this idea and its implications. The explanation that their pain has a real but not visible aetiology often comes as a welcome revelation.

General advice includes reassurance, vulval skin care, minimisation of topical therapy (which is often irritating) and withdrawal of topical corticosteroids. Rest is an important component of treating this condition. As the pain improves, many patients become rapidly more active, causing a temporary relapse.

Therapeutic interventions include oral medication, physiotherapy and, sometimes, counselling, and are often used together. Although some surgical techniques may have a role, we believe they are rarely necessary.

Oral medications

As with other forms of neuropathic pain, the use of oral medication can relieve vulvovaginal neuropathic pain. The most effective medications include:

- the tricyclic antidepressants amitriptyline and nortriptyline (off-label use) – 10 to 50 mg at night
- doxepin (off-label use) – 10 to 30 mg at night appears to be useful in patients whose main neuropathic symptom is itch
- pregabalin – 25 mg at night initially, increasing to 75 mg twice daily (150 mg/day) if tolerated and then increased gradually to a maximum of 300 mg twice daily (600 mg/day)
- gabapentin – 100 to 300 mg at night initially, increasing to twice daily (200 to 600 mg/day) and then slowly increased up to three times a day (300 to 900 mg/day; some patients require total daily doses of more than 1800 mg.

It is very important when commencing all of these medications to start with a very low dose and slowly increase it, as many patients find the side effects initially difficult to cope with.

The side effects of tricyclic antidepressants include drowsiness, disorientation, dry mouth, blurred vision, constipation, hypotension and conduction defects causing palpitations. They should not be used in patients with narrow angle glaucoma. Many patients will not hear of taking a tricyclic antidepressant because of the stigmatisation involved, and it is therefore important to explain that you are not using the drug as an antidepressant but as a pain reliever. Because of this concern, any side effect may cause patients to quickly give up taking amitriptyline or nortriptyline. They need to be encouraged to persevere as it is usually the first two weeks that are the most difficult. The tricyclic antidepressants are best given at night to minimise drowsiness. Dry mouth problems can be counteracted by taking one drop of pilocarpine 1% eye drops in a glass of water in the morning (off-label use).

Gabapentin and pregabalin may be combined with low-dose tricyclics.

If a medication is effective, most patients who respond will notice obvious, if small, improvements in their pain within four to six weeks. The dose should then be slowly increased until no more pain is felt. This can take up to one year. Once the pain has been relieved it may be necessary for the patient to remain on their medication indefinitely, although an attempt is usually made to reduce them once the pain has been adequately controlled for some months.

At the time of writing, we do not believe that there is sufficient evidence for the use of compounded topical neuromodulating agents. An integral part of the effect of these compounds is on spinal neurological function, which topical agents cannot change. Further, topical agents may cause hypersensitivity reactions and they are expensive.

Complementary medicine, counselling and explanation

Some patients appear to benefit from acupuncture and chiropractic treatment and some find that counselling is helpful, particularly if the counsellor has expertise in chronic pain management.

In some patients, just an explanation of what is happening is all that is needed. If the pain is not severe, some patients prefer to live with it rather than take medication or they embark on other therapies.

A note about topical oestrogen

Topical oestrogen is a treatment for symptomatic atrophic vaginitis, which occurs in lactating or menopausal women and, rarely, in women with conditions such as starvation and anorexia nervosa.

Atrophic vaginitis is due to oestrogen deficiency and usually presents with dyspareunia alone, which patients often correctly identify as being associated with dryness. Some hypo-oestrogenic patients, particularly those with a tendency to dermatitis elsewhere or who are atopic, may also develop a mild dermatitis of the introitus and labia minora. They may then complain as well of itching or irritation. The group who are prone to the vulvovaginal effects of postmenopausal oestrogen deficiency, however, are also the age group most likely to experience neuropathic pain. As a result, the atrophy may not be the cause of their vulval pain.

Examination shows a pale mucosa, with little lubrication and loss of normal rugosity. In younger lactating patients, however, there may be little to see other than a somewhat dry surface.

Atrophic vaginitis responds to topical oestrogen. If there is no response after six weeks of treatment, it is important to consider other causes for the patient's discomfort. Additionally, the speculative use of topical oestrogen vaginally in healthy,

premenopausal women is not only unnecessary but often causes topical hypersensitivity reactions. We frequently see patients who have been prescribed topical oestrogen with no benefit, but who have been told to continue it. We see no logic in this reasoning, and suggest that topical oestrogen should be ceased if it is not helpful.

CASE 2: REFERRED PAIN

Case scenario

Jane is a 42-year-old woman who, in her youth, was a keen horse rider. She has had many injuries, and these have left her with chronic low back pain. She now has two young children and during both pregnancies she experienced right buttock pain and coccydynia. She has gained 15 kg in weight in the past five years.

Jane presents with a constant unilateral burning sensation in the right labium majus associated with right-sided vaginal dyspareunia. Treatment with various topical therapies has been ineffective.

External examination is completely normal. On pelvic examination, however, there is obvious tenseness of the pubococcygeus, which is more pronounced on the right side. Gentle palpation of this muscle reproduces her dyspareunia.

Comment

The nerve supply to the vulva and anus originates from the L1-2 and S2-4 nerve roots via the genitofemoral and pudendal nerves (see pathophysiology in Part 1 of this article).[1] Compression or injury to these nerves may result in pain referred to the vulva, similar to sciatica. There is much overlap between this problem and neuropathic pain, but in referred pain, the pain is often unilateral or much more pronounced on one side, may have a shooting, 'electric shock', stabbing or cramping component and may have been of sudden onset. The patient is often much younger, and may present from early in the third decade of life.

When questioned, patients with pain referred to the vulvovaginal region may also give a history of low back pain or sciatica, and may have a history of a back injury from lifting, falling off a horse, a skiing or motor vehicle accident or falling on the coccyx. Childbearing often worsens the pain. Exacerbations can be associated with episodes of worse low back pain.

Anorectal disease and dysfunction may also cause referred vulvovaginal pain, although in these cases the pain is typically central.

In Jane's case, the lumbosacral dysfunction has caused referred pubococcygeus dysfunction, leading to dyspareunia.

Management – physiotherapy

It is important in women with referred vulvovaginal pain to rule out gross spinal pathology, although this is very unusual. Management of any anorectal problems is also essential.

Management of this type of pain is ideally physiotherapy delivered by a physiotherapist skilled in both the pelvic and spinal regions.[2,4] Neuromodulating drugs may be necessary. General measures such as lifestyle modification and weight loss will help.

The role of physiotherapy in the management of female pelvic pain

Physiotherapy is an essential element of treating many patients with nonlesional vulvovaginal pain. In these patients, retraining of the pelvic floor muscles will reduce resting muscle tone and in turn reduce dorsal horn sensitivity.

The role of physiotherapy is either as an alternative to medication or to allow an eventual withdrawal of medication. It has high levels of patient acceptance and the psychological support of the physiotherapist should not be underestimated. We recommend physiotherapy if the patient is well enough and young enough to embark on it.

Physiotherapy history

The physiotherapy history is similar to the medical history. Physical examination, however, concentrates on the musculoskeletal components of the spine, pelvis and lower limbs. A gentle but thorough assessment may take more than one session, because of the high levels of anxiety and fear in this patient group.

Resting tone, trigger points and pain scores of pelvic floor, hip and relevant spinal muscles are assessed, as well as the presence or absence of prolapse and any apparent fascial defects. A more generalised musculoskeletal assessment is also performed, including assessment of gait and posture, generalised muscle tone and breathing patterns, and specific examination of the lumbar spine and pelvic girdle plus identification of any trigger points in the abdominal, hip, gluteal and back muscles.

Physiotherapy management

Education is an important first step when using physiotherapy techniques in women with referred vulvovaginal pain. A clear explanation of the possible causes of their pain and the nature of chronic pain is essential. Behaviour and lifestyle modification such as correct posture, good back care, good bladder and bowel habits, vulval hygiene, avoidance of aggravating factors, stress reduction and incorporation of appropriate general exercise and relaxation activities/techniques into daily life is important.

Physical techniques aim to increase awareness and proprioception, normalise tone, improve muscle discrimination and relaxation, desensitise and increase tissue elasticity and reduce the fear of vaginal penetration.[2] Techniques may include myofascial massage and trigger point release. Vaginal dilators may be used both during sessions and as part of the patient's home program. Once normalisation of resting pelvic floor muscle tone had been achieved, further attention may need to be directed to improving the strength, endurance, co-ordination and timing of these muscles.

Treatment of coexisting lumbar spine and sacroiliac joint dysfunction, using joint mobilisation and muscle energy techniques, is often necessary.

CASE 3: SOMATOFORM DISORDER

Case scenario

Alison is a 19-year-old woman who presents with her mother. She has been suffering from vulvovaginal pain since she was 14 years of age, and says that the pain can radiate to involve her entire body at times. It has interfered greatly with her quality of life. She is unable to have intercourse and has not been able to sustain a relationship. Although she started a university degree, she dropped out half way through her first year and now rarely leaves the house because of her pain.

Alison and her mother have visited over 20 doctors and she has been extensively tested to rule out a sexually transmitted or other infection. She has had three biopsies, two EUAs (examination under anaesthetic), a laparoscopy and a cystoscopy. No abnormality has been detected. She has taken large doses of neuropathic pain-relieving medications with no improvement. Most of the history is given by Alison's mother, whose communication style reveals despair. Alison's affect is one of passive aggression.

Examination is normal but Alison is exquisitely sensitive to even the lightest touch. Your attempt to examine her is met with minimal co-operation. It takes some time to permit a brief examination and after a few seconds she clamps her legs together and pushes you away, stating that she is sick of being 'poked and prodded'.

Comment

Patients with psychogenic pain are a very difficult group to diagnose and in many cases impossible to treat. It takes experience to be able to recognise this condition, and even so it is inevitably a diagnosis of exclusion.

Typically with a somatoform disorder, the complaint is of constant pain, often with a burning quality, that does not fit easily with physiological pain patterns. There is a bizarreness to the descriptions, which may change with each consultation. The pain is often not confined to the vulva and may generalise to involve the whole body.

Patients with psychogenic pain rarely come to the consultation alone. They are brought in by relatives who are desperate for an end to the misery and expense that the patient inflicts on all the family members, who are inevitably controlled by the patient. Often these patients have seen numerous doctors and are angry and frustrated. If a patient does come by herself, she often remarks that her family has insisted that she sees a doctor. These patients may be quite unco-operative when examination is attempted.

This type of patient experiences pain as a manifestation of an underlying psychiatric illness, and may have a conversion disorder or Münchausen syndrome. They derive secondary gain from the pain, and do not wish to recover as the pain satisfies their needs; it is their relatives who want a solution. A psychiatrist is one doctor that these patients usually flatly refuse to see. They medicalise their problem, and as soon as they encounter a doctor who refuses to legitimise their medical model, they seek help elsewhere.

Our approach is to help the relatives cope with the patient. It is best for both parties if the secondary gain in truncated. Unfortunately, the relatives are often unwittingly part of the problem, and patience and persistence are needed to convince them of the real nature of the illness.

Depression and other conditions affecting pain-coping ability

Depressed patients may also experience pain as a manifestation of their depression, but a more common scenario is a patient with a painful physical vulval condition who copes poorly due to a concurrent depressive illness. As a result, a physical condition that would normally be mildly painful is experienced as excruciating.

Patients with obsessive compulsive disorder may also find physical vulvovaginal conditions extremely difficult to cope with. Washing rituals may further irritate their skin, they may become irrationally focused on infection, particularly genital herpes, and because disgust is a major issue for them, they may experience great difficulty touching themselves to apply creams or insert tampons. They may insist on wearing pads and liners at all times because they feel this is hygienic, which further exacerbates their skin problem. As a result, they may find themselves in a vicious cycle. Not surprisingly, anxiety and depression commonly supervene.

Unlike patients with a somatoform disorder who are commonly angry, these patients present as sad and anxious. Even when they lack insight into their condition they are usually receptive to the suggestion of psychiatric help and antidepressant medication.

Any patient with a longstanding vulvovaginal problem, particularly where pain is part of the symptomatology, may become depressed and the depression will exacerbate the pain. For many patients, the concept of long-term control of a condition that cannot be cured is very difficult to accept.

SURGICAL TREATMENT FOR PATIENTS WITH VAGINAL DYSPAREUNIA

Five surgical approaches have been advocated in the medical literature for women with vulvovaginal pain:

- Fenton's procedure, where the superficial muscles of the posterior introitus are incised to increase the dimensions of the introitus
- vestibulectomy, where the sensitive area of the introitus is excised, and the lining of the vagina is undermined and advanced to cover the defect
- surgical excision of persistent skin fissures
- intramuscular injection of botulinum toxin
- surgical decompression of the pudendal nerve.

Vestibulectomy

The rationale for vestibulectomy is that the inside of the vagina is not sensitive to pain, and replacing sensitive introital skin with vaginal lining will eradicate the pain.[5] Advocates of this procedure claim a very high success rate where other treatments have failed. The difficulty is the patients have all been characterised as having

'vulvodynia', which is a descriptor rather than a diagnosis, and therefore represent a heterogeneous population.

In our practice, we have never needed to resort to this procedure. We are of the opinion that vestibulectomy may not be necessary when an accurate diagnosis leads to effective (nonsurgical) treatments.

Surgical excision of skin fissures

Surgical excision of skin fissures is rarely indicated. Persistent fissures should always be assumed to be due to a skin condition, but the clinical signs may be very subtle indeed, and easily missed. Surgery carried out on chronically inflamed vulval skin may result in a worsening of the underlying condition.

There is a very small group of patients in whom the primary problem appears to be fissuring without any evidence of underlying inflammation. This is the only group where such surgery is indicated.

Botulinum toxin treatment

Intralesional botulinum toxin, which is injected directly into the levator ani muscle, is used to temporarily paralyse the muscular spasm that causes the pain. It does appear to be effective as an adjunct to physiotherapy, enabling patients who have a great deal of trouble releasing the pelvic floor to progress with exercises. This therapy is still in an experimental stage. The effect lasts about three months, and the injections have to be repeated. Side effects include the possibility of urinary and faecal incontinence, and the cost is not inconsiderable.

At this stage we still see it as something to be considered when more conservative therapy has failed.

Surgical decompression of the pudendal nerve

Pudendal neuralgia has been suggested as a cause of pain in the distribution of the pudendal nerve. The diagnosis is confirmed with pudendal nerve block. Surgical decompression of this nerve where it runs in Alcock's canal is sometimes performed. As with vestibulectomy, we consider that careful and comprehensive nonsurgical treatment usually suffices.

CONCLUSION

The differences between lesional and nonlesional vulvovaginal pain are discernible if a careful and comprehensive history and examination are performed. Causes of lesional pain include infections and inflammatory dermatological conditions that result in inflammation, ulceration, blisters, fissures and adhesions, and also tumours. Lesional pain usually resolves with resolution of the underlying problem. Evidence is accumulating that the majority of women presenting with nonlesional vulvovaginal pain have a neuromuscular disorder that is amenable to treatment with a multidisciplinary approach that includes neuropathic pain-directed medication, physiotherapy and counselling.

The patient presenting with vulvovaginal pain who has an apparently normal examination should never be dismissed. An initial search should always be made for a dermatological/lesional cause for their symptoms. Once lesional causes are ruled out, an attempt should be made to categorise these patients into nonlesional diagnostic groups so that specific targeted therapy may be initiated. MT

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