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Common Vulval Dermatoses in Clinical Practice

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Abstract

Pathology of vulva is common due to structure, nature and its susceptibility to friction. Lesions can be results from infection, trauma, neoplasia or immune response. As a result, symptoms may vary from pain, dyspareunia, discharge, pruritus or bleeding. Most of the woman with vulval problems who referred to gynaecology and vulval clinics are suffer from lichen scleroses, vulval candidiasis, lichen simplex chronicus and lichen planus.

Keywords: Vulval; Dyspareunia; Dermatological; Prepubertal; Hidradenitis

Abbreviations: ISSVD: International Society for the Study of Vulvar Disease; LS: Lichen sclerosus

Introduction

Benign vulval skin conditions are common and community based survey found nearly 20 percent of women suffer significant vulval symptoms [1]. Vulva consist of external genital organs of female includes mons pubis, labia majora, labia minora, clitoris, urinary meatus, vaginal orifice and hymen (Figure 1). The pruritus, vulval pain and dyspareunia are the commonest symptoms. It needs to assess about the aggravating and relieving factors of the condition. Allergic history is important. Underlying medical or surgical problems can manifest as vulval problems. Examination of the vulva needs to be performed with adequate illumination in the gyanecological couch. Vulvoscopy needs in suspect cases of vulval intraepithelial neoplasia. Assessment of other sites with condition associated with extragenital manifestation such as oral cavity, flexural surface, neck, elbow and knees. It is better to perform the swab tests, iron studies, thyroid functions and sexually transmitted disease screening in relevant cases. Skin biopsy indicated to rule out malignancy, inflammatory condition and failed to respond to standard treatment. The procedure of performing the vulval skin biopsy and general advice for vulval care has been given in Tables 1 & 2 respectively. International Society for the Study of Vulvar Disease (ISSVD) has described a new classification in 2011 based on skin features in a consistent and systematic manner. In thus review article we discussed common vulval dermatological problems and their management.

The area wish to biopsied should clean with the antiseptic solution. Then infiltrate with the 2% lidocaine solution and with the punch biopsy forceps make a cut. With the tooth forceps raise the skin edge and with the fine dissecting scissors undermine the circular biopsy and then free it. Specimen should send in a formal saline solution for histological analysis.

Table 1: How to performed a vulval biopsy?.

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Avoid soap, gel, bath products and antiseptic solutions in the vulval area.
Avoid tight fitting undergarments. White cotton underwear will be benefitted.
Sleep without underwear.
Avoid wearing panty liners or sanitary towels on regular basis.
Shower rather than a bath.
Avoid washing powders to wash the cloths.
Use the white toilet papers (Avoid colour papers)

Table 2: General advice on Vulvar Hygiene.

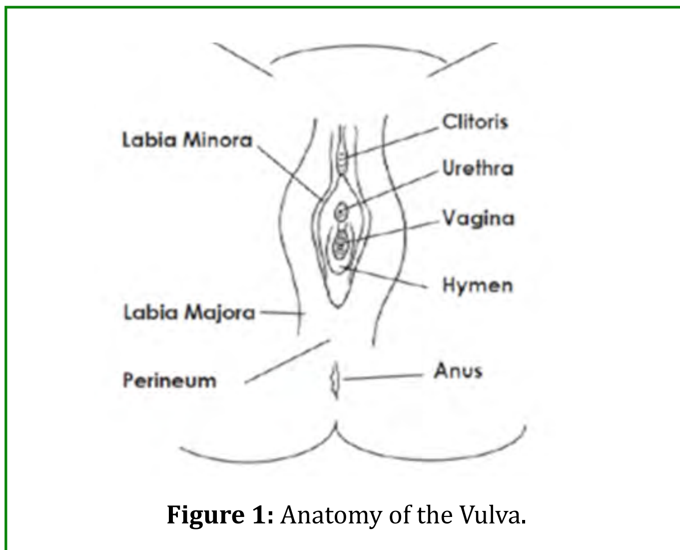

Figure 1: Anatomy of the Vulva.

Inflammatory Diseases of Vulva

Lichen sclerosus (LS)

This accounts for nearly 25 percent of patients in vulval clinic [2]. Usually common in post menopausal women and can be found in reproductive age women and children. More common in Caucasians. Earlier it was named as lichen sclerosus et atropicus ,however, currently we do not used that term as there is no atrophy of the skin in histological assessment. Exact cause is unknown and theory of hormonal, genetics and immunological has suggested [3]. This can be multifactorial. The most common symptoms would be the pruritus itching due to irritation due to inflammation of the fine nerve fiber endings. This pruritus induces itching leads to vicious cycle leads to excoriation and further thickening of the vulval skin. Most of the symptoms confined to a genital regions. With the chronic process it leads to architectural changes, tenesmus, dyspareunia.

On examination there will be erythema and thickened white skin. Some times in advanced cases distorted vulval appearance can be seen such as narrowing of introitus, urethral obstruction, clitoral concealment, fusion of labia minora and majora. This leads to paiful intercourse, difficulty in urination [2,3]. With the advance of the condition lesion can spread to perineum and anus leads to figure of eight or hour glass appearance (Figure 2). Diagnosis of the condition by histology and hyperkeratosis with sub epithelial haemorrhage would be seen. Estrogen deficit leads to thinning of epidermis. However, in chronic cases histology can be non-specific. Prepubertal lichen sclerosus can be improved during puberty. Lifetime risk of squamous cell cancer of vulva is around four percent.

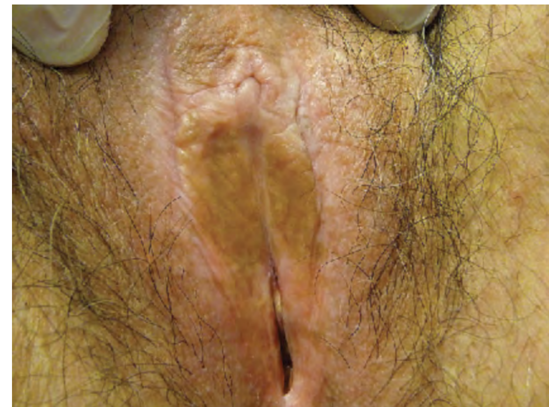
Figure 2: Lichen Sclerosus: Note the Hour glass appearance.

Management

There are no specific curative methods. Aim is to alleviate the symptoms, prevent the disease progress, prevention of cancer transformation and improve the quality of life. In advance cases need multi-disciplinary team approach. The order of the management would be corticosterioids as first line, topical tacrolimus as second line and lazer and surgery for advanced cases [4]. Ultrapotent corticosteroids are the cornerstone of the management and 0.05% clobetasole propionate is the recommended. Ointment preparation is better tolerated due to less allergic properties. Lower potent steroids and topical testosterone are not recommended. There are no randomized trial to support the exact frequency and duration of treatment; however auther recommended that daily treatment for one month and gradual reducing the regime. Early initiating of treatment would prevent the significant scarring and vulval deformity.

Topical estrogen therapy is not recommended, however, it is indicated in atropic changes of vulva for relive the symptoms. Topical retinoids would reduce the hyperkeratosis, induce local angiogenesis, improves dysplastic changes. Nearly 10 – 20 % of patient would not improve the symptoms with steroids alone. The calcineurin inhibitors (Tacrolimus and Pimicrolimus) can use in resistant cases. The mode of action is mainly by reducing inflammation via T lymphocyte response. The regime should be 0.1% tacrolimus twice daily. The advantage is that no steroid related side effects. Surgery and lazer treatment are reserved for restore the anatomy of vulva for urinary and sexual functions and neoplastic changes. Reagglutination can be prevent by ultrapotent steroids and surgical [5,6]. Women should advice on self-examination methods to detect any recurrence or neoplastic changes.

Lichen Planus

This condition can appear in both cutaneous and mucosal surface. More common in oral mucosa. Aetiology is still unknown. However some evidence suggest autoimmunity against basal keratinocytes. Not believe any hormonal relation to this condition. Common in menopause women, however, it can affect any age irrespective of gender. Small risk of transformation to squamous cell carcinoma of vulva is present. Patient typically present with intractable itchiness, pruritus, painful intercourse (either dyspareunia or apareunia), chronic vaginal discharge, post coital or post menopausal bleeding. On assessment there are purple plaques and papules in shiny polygons, with a fine white reticular pattern of lesions (wickham's striae). Papule is common and asymptomatic in 50% of patients [7]. It can be well demarcated in vulva or can be extend in to the vagina which results painful desquamative vaginitis. Histological features include degeneration of the basal layer of the epidermis and lymphocytic infiltration of the dermoepidermal junction.

There may be irregular epidermal hyperplasia resemble the saw tooth appearance. There are three major varieties of lichen planus in vulvar skin- erosive, classic and hypertrophic. Erosive is the commonest type which involve the vulva, vagina and gingival margins (Figure 3). Rarely it can affect to oesophagus, external auditory meatus and lacrimal ducts. This variety is more difficult to cure. Classic variety involves the vulva and perianal skin. Hypertrophied lichen planus is the least common and can be found in anogenital skin. Both classic and hypertrophied lichen palnus can rarely progress to squamous cell carcinoma [8-11]. This condition is usually be resolve within one to two years, however recurrence can be common. Mucosal lichen planus is difficult for complete cure and more persistent. Comparison of lichen sclerosus and lichen planus given in Table 3.

	Lichen Sclerosus	Lichen Planus
Age of onset	Common in post menopausal Childhood disease possible	Common in post-menopausal Any age can possible
Aetiology	Unknown, 40% has other autoimmune condition	Unknown, has some autoimmune back ground. Not related to hormonal
Clinical presentation	Intense pruritus Erythema of vuvla Dyspareunia	Present with purple plaques and papules with fine reticular pattern. (Wickham's striae)
Examination findings	Typical figure of eight appearance due to erythema and thickened white skin appearance.	Well demarcated erythema in vulva
Extra genital manifestation	Rare	Can affect any part of the body. Common in oral
Histology findings	Thinning of epidermis Hyperkeratosis Sub epithelial haemorrhage	Degeneration of the basal layer of epidermis. Lymphocytic infiltration of the dermoepidermal junction.
Treatment	Ultrapotent steroids Topical tacrolimus Lazer and surgery for extensive disease.	Ultrapotent steroids Topical tacrolimus
Progress of the disease	Prepubertal lichen sclerosus improves at puberty. Post-menopausal persist and symptomatic control only	May resolve spontaneously within one or two years

Risk of squamous cell cancer	4% life time risk	Very rarely progress (classic and hypertrophied variety)
Recurrence risk	Common	Common

Table 3: Comparison of Lichen Sclerosus and Lichen Planus.

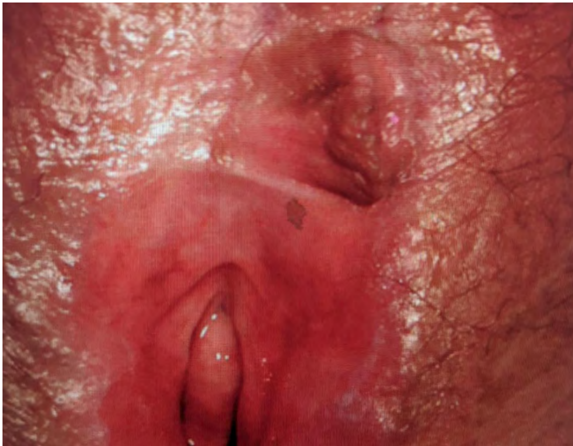

Figure 3: Lichen Planus of the vulva (Erosive Type).

Management

High potent topical corticosteroids are the first line treatment. Clobetasole and topical tacrolimus appear to be effective for vulvovaginal lichen planus. Oral retinoids would be better for non-vulvar lichen planus [12]. Patient with vaginal narrowing and sexual difficulty would be benefited with topical steroids, vaginal dilators and surgical adhesiolysis.

Eczema and Dermatitis

Chronic Dermatistis or Lichen Simplex Chronicus

Lichenification of the skin (thickening) due to chronic scratching, rubbing and handling. This term used due to isolated lichenification of the skin in the absence of background dermatitis. Common in outer surface of labia majora, mons pubis with hypopigmented or hyperpigmented lesions. It can be a chronic process due to itchy- scratch cycle. It can be aggravated by chemical or contact dermatitis, stress situation and low body iron levels [13].

Management

Antipruritic drugs and topical ultra potent steroids would be helpful to break the itchy- scratch cycle. General vulval care and avoidance of soap, irritant, emollients needed [14].

Contact Dermatitis

This condition classically present with immediate burning and stinging sensation following exposure to offending agent. This results erythema, swelling, vesicle and bullae formation (Figure 4). This can get secondarily infected with bacteria.

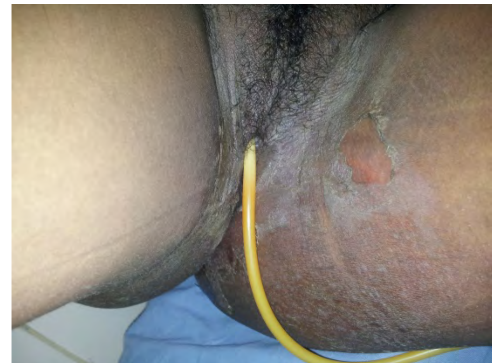

Figure 4: Contact Dermatitis of Vulva.

Management

Avoid offending agent and treat with antibiotics if evidence of infection. Sitz bath with plain water also helpful. Antipruritic medication to relive the symptoms. Rarely need steroids treatment [15,16].

Vulval Psoriasis

Psoriasis is a common skin condition which affect to the 2% population. Aetilogy is unknown and can be immunological mediated. This can be triggered by infection and trauma (koebner phenomenon) and certain medications. Usually presented with silvery white scaly plaques and most of the time diagnose can be clinical. Lesions are commonly seen in inner and outer aspect of labia majora [17]. Some times lesions can be extend to perianal region and gluteal cleft. Histology revealed marked epidermal thickening with acanthosis and inflammatory cells infiltrated into epidermis.

Management

Topical steroids are commonly used as rapid onset of action and effectiveness. In extensive cases need multidisciplinary approach and treat with methotrexate, cyclosporine and biologics (Infliximab, alefacept)

Seborrhoeic Eczema

This is common in childhood and pubertal age group. Aetiology may be fungal in origin (Pityrosporum ovale).

Presented with erythematous lesion and greasy scales. It will affect anogenital skin, inguinal folds and gluteal clefts. This condition is very difficult to differentiate from flexural psoriasis as histological features also compatible with later. The management would similar to psoriasis.

Vulval Candidiasis

Usually present with vulval itchiness, vulval fissuring, vaginal discharge and urinary symptoms. Nearly 90% cases are affected with candida albicans. Other species are the candida glabrata, candida tropicalis, candida krusei and candida parapsilosis. Recurrence are very common [18]. Treated with clotrimazole or fluconazole.

Bullous Disease

Epidermolysis bullosa

It is a group of genetically related skin condition characterized by blistering and fragility of the skin and mucosa [19]. There is no specific treatment for the condition and gene therapy might be helpful.

Benign familial chronic pemphigus

This is an autosomal dominant condition and intraepithelial blisters appear in the groin, axilla, and neck region. Usually patient present with third and fourth decade of life. Friction might worsen the condition and they can get secondarily infected with bacteria. Histology revealed extensive acantholysis [20]. Topical steroids would be useful. Rarely squamous cell carcinoma can arise from these lesions.

Erethema Multiforme

It is an acute reaction pattern which mucosal erosions and ulcers can occur. Steven Johnson syndrome and toxic epidermal necrolysis is the extreme form of erethema multiforme (Figure 5). Vaginal stenosis is a complication. Systemic steroid and immunoglobulin used to treatment [21].

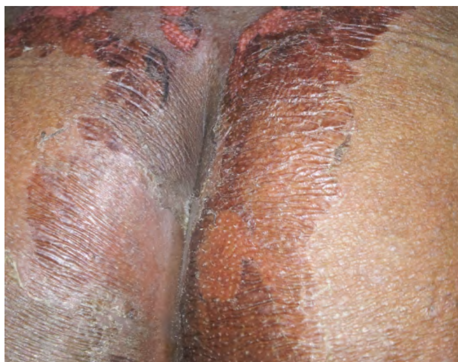

Figure 5: Erethema Multiforme.

Bullous Pemphigoid

Localized vulvar pemphigoid is a rare entity and characterized by tense bullae. Mucus membrane involvement is less common. Complications can be labial fusion, scarring and introital shrinkage which ended up with urinary and sexual problems. Histology showed sub epidermal blisters [22]. Initial treatment would be topical corticosteroids and adhesions need to be surgically divided. Resistant cases immunosuppressive therapy, mycophenolate mofetil and intravenous immunoglobulin would be needed.

Vulval Ulceration

Aphthous Ulcers

Oral aphthous ulcers are the commonest. However vulval aphthous lesions also can occur. It can easily mistake with herpetic ulcers. Typically they are, yellow color small ulcers (few millimeters) and surrounded by erythematous rim [23]. Treatment can be topical steroids, antibiotics and topical anesthetics agents.

Herpes Ulcers

Lesions are caused by both type 1 and 2 variety, however, latter is the commonest. Nearly 90% lesions are acquired through sexual contact. Characteristically lesions are appearing as aggregated vesicles later rupture and form the ulcers (Figure 6). Lesions are very painful and burning sensation [24]. Diagnose by clinically and viral cultures. Treated with acyclovir or valcyclovir. Recurrences are common.

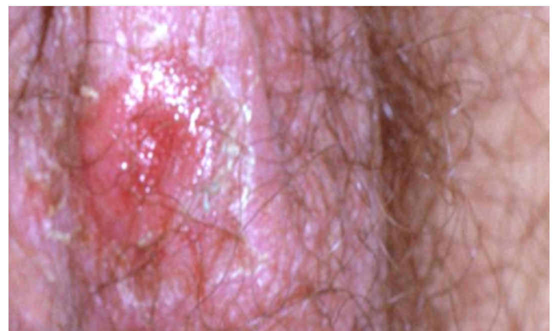

Figure 6: Herpes Lesion of a vulva.

Hyper or Hypo pigmented lesions

Acanthosis Nigricans

This is more common among obese women and thickened hyper pigmented plaques seen in labia, inguinal folds, neck and axillary regions [25]. This can associated with insulin resistance or may be a paraneoplastic manifestation of underlying malignancy.

Vulval Melanosis

There are small pigmented lesions can be seen without prior history of inflammation. Always need to perform a biopsy to exclude underlying malignancy. Histology revealed increased melanocytes [26].

Vitiligo

Common autoimmune disorder with characterized with depigmentation of the skin (Figure 7). Usually not associated with change of the anatomy of the vulva and can be seen other parts of the body as well [27]. No effective treatment available at the moment.

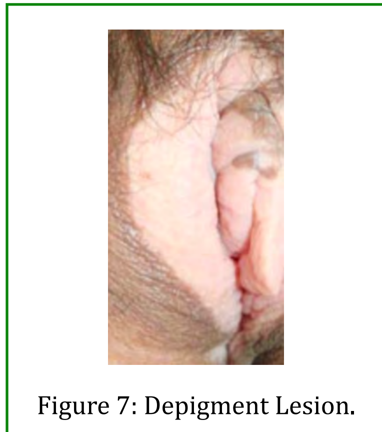

Figure 7: Depigment Lesion.

Vascular or Lymphatic disorders

Haemangiomas are congenital and usually asymptomatic. Mons pubis and labia majora are the common sites involved. Occasionally, there may be rupture of vessels and leads to ulceration. Propranolol and steroids can be used to shrink the lesions. Lazer treatment is reserved for cosmetic benefit. Vulvsal varicosities are usually associated with varicose veins of the lower limb as well. Common in pregnancy and usually resolve after delivery. May need to exclude obstructive pelvic lesions. Acute lymphedema are associated with sudden onset of swelling of the vulva. Can be associated with type 1 hypersensitivity and sexual intercourse. Contact urticaria due to seminal fluid also recorded. Chronic lymphedema also possible following infection, malignancy and inflammatory bowel disease [28]. Lymphangiectasia means development of small lymphatic vessels with background of chronic lymphedema. This can be due to inherited defects or secondary to malignancy, radiotherapy or inflammatory disease. Lazer treatment can be used to treat the condition.

Manifestation of Underlying Diseases

Extramammary Paget's Disease

This is very rare and commonly seen in post menopausal women. They present with pruritus. On assessment erythematous excoriated lesions can be seen. Usually affect to the hair bearing areas. This can associated with underlying adenocarcinoma. Need to evaluate the breast, gastrointestinal system and urinary system. Treat with surgical excision to rule out the carcinoma [29].

Inflammatory Bowel Disease

Anogenital lesions can occur secondarily to inflammatory bowel disease and more common with crohn's disease than the ulcerative colitis. Usually they present with either unilateral or bilateral vulval oedema. Perianal ulceration can be present. Diagnose the condition by biopsy and histology revealed that granulomatous inflammation [30]. Treated with multidisciplinary settings and initially with steroids, antibiotics, immunosuppressive agents and biologics (Infliximab).

Behcet's Syndrome

This is the triad of symptoms and consisted of oral, genital ulceration and uveitis. Usually they start before the fifth decade of life. Vulval ulcers are usually larger, painful and more common in labia minora. Histology is non-specific. Management in multidisciplinary settings and topical steroid for genital ulcers [31].

Pyoderma Gangrenosum

This is a very aggressive ulcerative disease associated with rheumatoid arthritis, inflammatory bowel disease or myeloproliferative disease. Lesions can be pustular then it can be rapidly to form the ulcers [32]. Histology is inflammatory but nonspecific. Can be treated with steroids, ciclosporin, azathioprine, mycophenolate mofetil or dapsone. Surgery should avoid treating the condition.

Vulval Lumps and Bumps

Bartholin's cyst is formed when the duct of the bartholin gland get obstructed and secretions are filled in the gland and convert to a cyst. If it get secondarily infected abscess can be formed. This is very painful and associated with swelling of the surrounding vulva. Marsupialisation is more effective than the simple incision and drainage. Need treatment with antibiotics and recurrence cases need to remove the gland. Sabaceous cysts are common and generally mobile masses due to fibrous tissue which get filled with sebaceous materials. More common in hair bearing areas such as mons pubis and labia majora. Need surgery as a treatment and need to remove the punctum to prevent the recurrence. Papillomatosis are benign epithelial tumours and grows as a finger like projections. Important to differentiate from the vulval warts. Vulval lipomas are uncommon and they are usually asymptomatic. To correct the disfigurement need surgery. Hidradenitis suppurativa is a chronic skin condition

and associated with skin abscess, fistulas and skin scarring. Treatment includes antibiotics, retinoids and surgery.

Conclusion

Benign vulval conditions are common in gynaecology practice. Vulval itchiness and pain are the commonest symptoms. Skin biopsy is always not necessary to diagnose the condition unless suspect of underlying malignancy. Advice on general care of the vulva is very important to avoid disease progress and symptomatic control. Lichen sclerosus is the commonest condition seen in vulval clinic and it has 4 % risk of progress to squamous cell carcinoma. Most if the benign vulval condition can be manage with steroid treatment and vulval hygiene methods.

Conflicts of interest

There are no conflicts of interest.

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