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Differential diagnosis of vulvar pruritus: Psoriasis and vulvar lichen

Diagnóstico diferencial de prurido vulvar: Psoríase e líquen vulvar

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ABSTRACT

Objective: To address some of the main pathologies that can bring the complaint of "itching" to the doctor's office, namely: Vulvar Psoriasis, Lichen Sclerosus and Lichen Planus Vulvar. This recurrent complaint in the clinics is characterized by an itching or discomfort in the vulva region, with various causes. Methods: The research was carried out based on a literature review with databases published in the CAPES Portal, SciELO and PubMed. Results: In the literature review, it was possible to clarify that Vulvar Psoriasis, one of the differential diagnoses, is a chronic non-contagious dermatosis, with intense itching, discomfort and desquamation. Lichen Sclerosus, in turn, is a chronic dermal pathology of intense inflammatory character, with itching, pain and irritation in the mucosa, while Lichen Planus Vulvar is a chronic inflammatory disorder, with pruritus, pain, burning, dyspareunia and atrophy of the cultivar and vaginal architecture. Conclusion: In the end, the present study concludes that the non-treatment of this complaint, common to the three pathologies, has consequences that impact the quality of life of the patient.

Keywords: Vulvar prurid, dermatopatias, psoriasis, flat lichen, lichen vulvar sclerosis.

INTRODUCTION

Vulvar pruritus is defined by discomfort and itching in the vulvar region, and is a condition ideally resolved with medical follow-up. It is a common complaint in gynecological consultations, and is often neglected, misdiagnosed, and undertreated (CROWLEY, MARTIN, 2020). In addition, it has various causes, including: infections, such as candidiasis, sexually transmitted infections (STIs), contact dermatitis and atopic dermatitis. The main warning signs for this condition are the presence of ulcers, plaques or nodules on the vulvar skin, in which case biopsy should be submitted after STI are ruled out. These findings raise the suspicion of a possible association with vulvar intraepithelial neoplasia and cancer (MINISTRY OF HEALTH, 2016).

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The diagnosis of vulvar pruritus is clinical, and biopsy is performed in dubious cases. It is known that there is a wide variety of possible differential diagnoses for this symptom, covering several pathologies. Among the possible etiologies, we highlight in this study vulvar psoriasis, lichen sclerosus, and lichen planus, which are relevant and prevalent diseases in Brazil. As soon as an abnormal vulvar pruritus is identified, it is indicated that the patient strictly follow the use of medications prescribed by the responsible health professional, in addition to prioritizing proper hygiene and suspension of possible allergenic products (CROWLEY, MARTIN, 2020).

METHODS

The research was carried out based on a narrative review based on data published in the CAPES Portal, SciELO and PubMed, using descriptors related to Psoriasis, Vulvar Pruritus, Vulvar Lichen Sclerosus, Gynecological Diseases and Signs and Symptoms. A total of 50 articles were collected, 12 of which were selected. The inclusion criterion was the correlation between vulvar pruritus, the related gynecological manifestations, and the differential diagnoses regarding the pathologies studied, and the exclusion criterion adopted was the language of the articles, selecting only those in Portuguese or English. On the PubMed platform, in addition to the descriptors, filters involving the date of publication and the availability of free texts were used in order to obtain updated and complete studies.

DISCUSSION

VULVAR PSORIASIS

Concept

Psoriasis is a chronic non-contagious dermatosis characterized by the presence of erythematosquamous lesions with whitish scales (MINISTRY OF HEALTH, 2016). It is a common, incurable and controllable autoinflammatory disease of the skin, which causes the appearance of lesions periodically (NETTO et al, 2006). This disease affects the skin tissue, in which reddish, hardened and scaling-prone plaques are formed on the skin, which can also affect nails and nails. joints (BETANCOURT et al, 2023), with the most common areas being elbows and knees (LENA et al, 2021). The disease has a familial predisposition (CROWLEY, MARTIN, 2020).

Epidemiology

Psoriasis is a disease of uncertain incidence, being found in different age groups and in both sexes. According to the Brazilian Society of Dermatology (SBD), in 2020 psoriasis affects 1.15% of the female population in Brazil. In addition, about 60% of adults with psoriasis develop lesions in the genital area at least once in their lives (MINISTRY OF HEALTH, 2016).

Symptomatology

Patients with vulvar psoriasis commonly complain of severe itching, local discomfort, and peeling mainly on the labia majora, perineum, and labia minora. According to the SBD, these erythematousquamous plaques on the vulva usually have a symmetrical, thin, and well-defined character. In addition, lichenification and abrasions are recurrent, and may or may not have manifestations of the disease in other areas of the body (SBD, 2021).

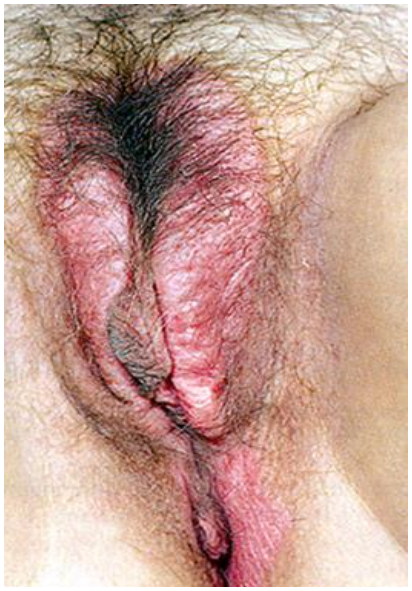
Because it is not a scarring dermatosis, there is usually no damage to the anatomy of the vulva, although some patients have partial damage to the labia minora. In addition, due to intense pruritus in the affected region, it is possible to trigger Koebner's phenomenon, which culminates in the development of new lesions in areas subject to trauma (JOHNSON et al, 2022).

Some studies have shown that physical sensations and injured areas in visible places generate psychological discomfort in the patient. Among the signs and symptoms are: sadness, frustration, shame, anxiety, and fear of forming affective, professional, and social bonds (VIEIRA et al, 2022).

Clinical presentations

- Thick confluent plates covering the mound of venus and labia majora forming an almost solid, horseshoe-shaped pattern with silvery white adherent scales (JOHNSON et al, 2022);
- Thin salmon-pink lesions with small scales of varying size and shapes (JOHNSON et al, 2022);
- Inverse psoriasis: well-defined, smooth, shiny plaques with absent or minimal scales, often confused with fungal or bacterial infections. It can occur in skinfolds or intertivinous skin, such as inguinal folds, crurals, perineum, umbilicus, gluteal cleft, armpits, and inframammary. In addition, it has a bilateral and symmetrical pattern, with erythema, maceration, and fissures extending linearly from the inguinal fold through the labiocrural fold and the gluteal cleft (JOHNSON et al, 2022).

Figure 1- Erythematous and squamous plaques involving the labia majora and perineum in a patient with inverse psoriasis (UpToDate, 2022).



Diagnosis and clinical practice

For diagnosis, biopsy is performed restricted to cases of atypical distribution or lesion restricted to the vulva. It is necessary to investigate family history and the presence of lesions in other areas of the body, in addition to screening all patients for the presence of psoriatic arthritis (SBD, 2021).

It is essential that the physician in charge be aware of the increased risk of these patients developing cardiovascular, autoimmune, inflammatory bowel diseases and osteoporosis. It is essential to carry out an inventory of the medications used by the patient, as the use of systemic glucocorticoids, oral lithium, antimalarials, interferon, and beta blockers can trigger disease crises (JOHNSON et al, 2022).

Treatment

To treat this disease, the severity of the extent of the clinical picture and the patient's psychological impairment must be taken into account (MOSCARDI, OGAVA, 2017). It is important to communicate to the patient the level of commitment to the treatment so that there is complete or almost complete resolution of the lesions. It is also worth remembering that a healthy lifestyle will allow for better control of the disease (TORRES et al, 2021).

For proper treatment, it is essential that there is local hygiene, avoiding hot baths. For pruritus, an ice pack and an antihistamine of 25 mg/night are recommended, in addition to the daily use of emollients. When there is an associated fungal infection, Ketoconazole 2% cream is used, and when it is bacterial, mupirocin is recommended (FELDMAN, 2022).

In case of mild to moderate conditions, treatment with 0.05% desonide should be started twice a day for three weeks, and after this period, Triamcinolone 1% should be introduced daily (FELDMAN, 2022). Finally, for severe conditions, treatment should be initiated with Clobetasol 0.05 twice a day for two weeks, followed by Clobetasol 0.05 once a day for two weeks. After the first four weeks of treatment, the use of Clobetasol 0.05 is started every two days for two weeks, and then long-term Triamcinolone is used (FELDMAN, 2022).

SCLEROSUS LICHEN

Concept

Lichen sclerosus (SL) is a chronic dermal pathology marked by intense inflammation, epithelial thinning, pruritus and pain, presenting a progressive character. The most affected region is the anogenital, and extragenital lesions can be found, such as in the breasts, thighs, shoulders, neck, back, wrists and, rarely, oral mucosa (COOPER, ARNOLD, 2022).

Epidemiology and etiology

The epidemiology of lichen sclerosus is not completely elucidated, and is possibly multifactorial. Two major peaks of incidence are considered, namely children and peri- and postmenopausal women (COOPER, ARNOLD, 2022).

It is worth noting that there are case-control studies in relation to the etiopathogenesis of LE and association with autoimmune diseases, it has been shown that the most observed autoimmune disease in patients with LE is thyroid disease, and as with the thyroid, it seems that it is likely that LE is a T-cell-mediated disease. supporting the idea that it is an autoimmune disease (COOPER et al, 2008). However, the exact etiology of LE is still unknown.

Finally, it is known that the factors most studied as possible causes for this pathology are genetic, autoimmune (alopecia areata, vitiligo, thyroid disorders), local (vulvar grafts), hormonal (low estrogen in prepubertal and postmenopausal women) and cell kinetics (tissue hyperproliferation) (COOPER, ARNOLD, 2022).

Symptomatology

The most recurrent symptoms in patients with lichen sclerosus are itching, pain and irritation of the mucosa. In addition to these, LE can manifest with anal discomfort (itching, pain, fissure, bleeding, constipation), dyspareunia, sexual dysfunction, and dysuria (COOPER, ARNOLD, 2022). In addition, LE is an incidental clinical finding and is rarely asymptomatic,

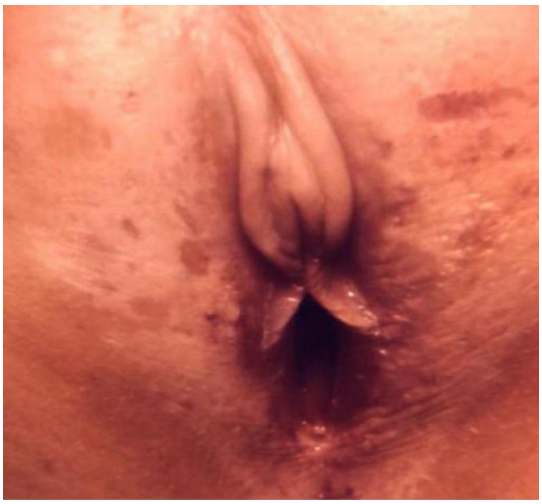
with pruritus, especially at night, being the main complaint of patients during consultations (LEWIS et al, 2018).

Clinical presentations

Initially, lichen sclerosus manifests as white, atrophic papules that tend to coalesce and form erythematous plaques. LE mainly affects the labia majora and minora, and can cause hemorrhagic, purpuric, hyperkeratotic, eroded or ulcerated lesions. In addition, sexual intercourse or increased friction can result in abrasions, hemorrhages, petechiae, and ecchymosis, demonstrating the fragility of the affected skin (COOPER, ARNOLD, 2022).

As the disease progresses, it is common for the architecture of the vulva to be lost with the fusion of the labia minora to the labia majora, fusing the foreskin and concealing the clitoris. The perineum and vaginal introitus tend to atrophy, generating dyspareunia and fissures during sexual intercourse or even in gynecological examination with a speculum. In addition, involvement of the vaginal canal may be present, although it is a rare condition (COOPER, ARNOLD, 2022).

Figure 2- Thinning of the skin and areas with increased or decreased skin pigment on the female genitals (Health Manual, 2021).



Diagnosis and clinical practice

Although histological confirmation is not recommended in all cases of lichen sclerosus, in clinical practice biopsy is performed in adults in cases of uncertain diagnosis, non-response to first-line drug treatment, and suspicion of associated malignancy. When children are affected, the

diagnosis is preferably clinical, and biopsy is used privately in refractory conditions or with atypical characteristics (DE LUCA et al, 2023).

Dermoscopy may be a viable option to support the diagnosis of non-invasive LE, as well as to optimize the biopsy site. In LE, the most prevalent feature is the presence of whitish or yellowish-white spots without structure, placed on a white atrophic background. These dermoscopic changes represent signs of dermal sclerosis and hyalinization (DE LUCA et al, 2023).

Adult women with SL have an increased risk of developing squamous cell cancer (SCC) of the vulva, and it is also not uncommon for patients with genital cancer to find untreated or asymptomatic LE. Finally, the relationship with other neoplasms is still unclear and the area of investigation is broad (COOPER, ARNOLD, 2022).

Treatment

Drug treatment is with ultra-potent topical corticosteroids; calcineurin and methotrexate inhibitors; and topical progesterone. In addition, it can be associated with intralesional corticosteroid therapy when there is a hypertrophic plaque that does not respond to the topic (COOPER, ARNOLD, 2022).

Similar to other chronic and progressive diseases, the patient needs to develop awareness of the pathology and have attitudes that can contribute significantly to the remission process. It is essential that the professional is willing to explain in a didactic way the changes that occur with the LE, addressing the chronicity, the possible complications if there is no treatment and the high rate of control when all measures are correctly applied. In addition, other key components that the patient must present during treatment are good hygiene, self-examination, medical follow-up at least once a year, and correct use of medication (COOPER, ARNOLD, 2022).

When initial treatment is not effective, it is important to check whether the medication is being used as prescribed, exclude superinfection, assess the need for intralesional corticosteroid therapy, reconfirm the diagnosis, look for signs of malignancy, and consider other causes of symptoms. If second-line therapy is required, topical calcineurin inhibitor is used. The other treatments used do not have proven efficacy and completely established superiority for most patients with LE. In this aspect, once the symptoms are controlled, patients who have developed vulvar disconfiguration with adhesions and scars can undergo surgery, seeking to mitigate these complications (COOPER, ARNOLD, 2022).

Following this line of reasoning, limitations were found in the systematic review studies that sought evidence regarding the use of laser as a treatment for SL, with limitations regarding the presence of long-term data on the treatment of genital LE with laser, including lack of information regarding adverse effects and the existence of confounding factors. such as the concomitant use of topical treatments, such as estrogen, during laser treatment (TASKER et al, 2021).

Finally, the chronicity of SL requires at least annual follow-up to assess therapeutic efficacy, remission, complications, and even malignancy. Since the pathology has no definitive cure, treatment aims to achieve complete remission of symptoms and prevent relapses (COOPER, ARNOLD, 2022).

PLANO VULVAR LICHEN

Concept

Lichen Planus (PL) is a chronic inflammatory disorder, which has four clinical varieties: erosive, papulosquamous, hypertrophic, and plane pilar (COOPER, ARNOLD, 2022). Erosive Vulvar Lichen Planus (PVL) is the most frequent presentation, 70% of cases (RUIZ, et al, 2014), manifesting with erythematous papules or plaques, eventually eroded, where pruritus is its main manifestation. It corresponds to an inflammatory process of the vulvar skin and/or mucosa, and can be observed in a localized or even generalized form (FEBRASGO, 2010).

Epidemiology and etiology

The prevalence of vulvovalginal LP is estimated at 0.5 to 2% (COOPER, ARNOLD, 2022), which is lower than that of lichen sclerosus and rarely affects children. It is more common in the peri- or postmenopausal period (MIRANDA, et al, 2014), with peak incidence between 30 and 60 years of age (RUIZ, et al, 2014).

Its pathogenesis is unknown, however the most accepted theory considers that activated T lymphocytes are recruited to the dermal-epidermal junction and induce apoptosis of basal keratinocytes (FEBRASGO, 2010).

Both CD4+ and CD8+ T lymphocytes are found in the lichenoid infiltrate, but with a predominance of the latter. The interaction between T lymphocytes and basal keratinocytes is enhanced by increased expression of intracellular adhesion molecules (ICAM-1) by keratinocytes (MIRANDA, et al, 2014). In addition, it was shown that thyroid diseases, alopecia

areata, and celiac disease were significantly more prevalent in patients with erosive vulvar LP, which suggests a strong association of LP with autoimmune diseases (MIRANDA, et al, 2014).

Symptomatology

Erosive Vulvar Lichen Planus manifests with itching, pain, burning, dyspareunia and destruction of the vulvar and vaginal architecture, if it affects the vagina. In addition, it may present in isolation or associated with an oral lesion or generalized skin rash, with a predilection for the trunk and curvature surfaces. Approximately 25% of women with the oral form also have vulvovaginal involvement (MIRANDA, et al, 2014).

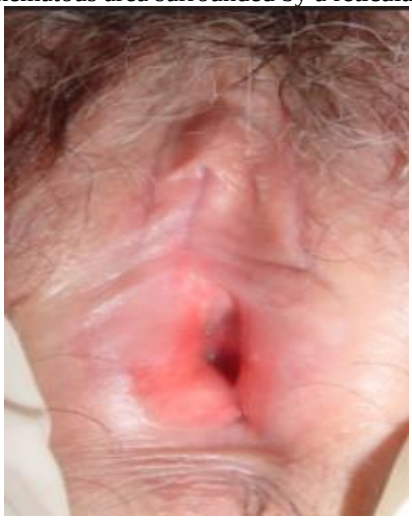
Clinical presentations

The lesions evidenced in PVL are characterized by long-lasting erythematous flat papules and, sometimes, distinct purpuric vascular lesions, which may be discrete or unite to form plaques. They tend to be scaly and eventually erosive. They most frequently occupy the inner surface of the labia minora in an erosive form (FEBRASGO, 2010). Although these vaginal lesions are not routinely observed, the possibility of the formation of vaginal bridles, caused by the evolution of the lesions, is routinely mentioned.

In erosive LP, vulvar lesions are typically located on the vaginal introitus, clitoris, foreskin of the clitoris, and labia majora and minora. They are erythematous, shiny and are associated with Wickham's striae - it is the best area to perform the biopsy and obtain diagnostic confirmation (FEBRASGO, 2010). Usually, there is a whitish border separating the lesions from the areas of healthy skin. Vaginal involvement is manifested by a large and yellowish discharge, as well as bleeding after sexual intercourse. In the most severe cases, synechiae may form, obliterating the vaginal lumen (MIRANDA, et al, 2014).

Erosive LP can also appear as part of a triad, which consists of vulvitis, vaginitis, and gingivitis, configuring vulvovaginal-gingival syndrome. In this case, we observed a scaly gingiva, with edema and erythema, associated with vulvovaginitis. Mucosal lesions are not necessarily simultaneous (MIRANDA, et al, 2014).

Figure 3 - PVL showing an erythematous area surrounded by a reticulated border (FEBRASCO, 2012).



The hypertrophic type, on the other hand, presents as hyperkeratotic and whitish plaques or papules, which can be single or multiple, located in the perineum or perianal region. It resembles squamous cell carcinoma, vulvar intraepithelial neoplasia, and lichen sclerosus, and it is necessary to make a differential diagnosis (LOPEZ, NAVARRO, 2016).

Finally, the classic type, also called papulosquamous LP, is characterized by a violaceous, flattened, very itchy papule that grows on keratinized skin. It is similar to extragenital cutaneous LP (LOPEZ, NAVARRO, 2016).

Figure 4 - Lichen Planus Classico (Revista Deana de la Especialidad, 2014).

Lichen planus pilaris has been described in a single case in the literature, affecting labia majora and mons pubis. It is a rare variant, in which there is perifollicular erythema and hyperkeratosis (COOPER, ARNOLD, 2022).

Diagnosis and clinical practice

In addition to the gynecological examination, the oral mucosa should be carefully examined in search of signs that show the presence of the oral form of the disease (LOPEZ, NAVARRO, 2016).

The suspicion can be confirmed through biopsy of the lesion and negative immunofluorescence (LOPEZ, NAVARRO, 2016), which should preferably be performed in a region adjacent to the edge of an erythematous lesion. If Wickham's streaks are present, they should be included in the sample. Typical histological findings are irregular acanthosis, hydropic degeneration of the basal layer, and band-like lymphocyte infiltrate of the dermis-epidermis junction (MIRANDA, et al, 2014).

Treatment

The treatment of LP is complex and difficult, and should include, in addition to general measures, topical, systemic or surgical treatment in severe cases. It is important to inform the patient about the chronic nature of the disease and the possibilities of treatment and, if necessary, recommend psychological follow-up (FEBRASGO, 2010).

Antidepressants can be used, helping with symptoms and promoting nocturnal sedation. Secondary infections should always be identified and treated. General measures should be adopted, such as the correct use of toilet paper, avoiding the use of scented soaps and lotions, wearing cotton underwear, avoiding tight clothing and preferring ointment medications, which, in addition to causing less aggression to the mucosa, have an emollient effect. Medications that stimulate lichenoid eruption should be suspended, if possible (FEBRASGO, 2010).

The first-line treatment for vaginal involvement is the use of a 25 mg hydrocortisone suppository, vaginally, twice a day, for two months. We then reduced the dosage to once daily for another two months. After this period, maintenance treatment is initiated with the use of one to three times a week until differential clinical improvement (FEBRASGO, 2010). In some cases, after a long period (around 10 years) PVL may go into remission, and may occur in about 10 to 15% of them (FEBRASGO, 2010).

The use of topical calcineurin inhibitors, such as tacrolimus and pimecrolimus, has been explored with varying degrees of success, with a risk of intense burning as a side effect in most women, as already mentioned (LOPEZ, NAVARRO, 2016).

Vaginal dilators can be used in combination with topical medications to prevent vaginal stricture. In cases where there has already been significant vaginal narrowing, surgical treatment

should be instituted to allow the patient to maintain an active sexual life (LOPEZ, NAVARRO, 2016).

CONCLUSION

As presented, vulvar pruritus is one of the most common complaints presented by gynecological patients, and its causes can be diverse, such as vulvar psoriasis, lichen sclerosus and vulvar lichen planus. Thus, it is essential for the gynecologist to know well each of the possible pathologies, signs and symptoms, in order to establish the correct differential diagnosis when required. Finally, vulvar pruritus has a great negative impact on the patient's life, and should be properly managed for a better quality of life and a good prognosis of treatment.

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