

## CASE REPORT

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# Zoon vulvitis in a woman of childbearing age. A case report

## Vulvitis de Zoon en mujer en edad fértil. Reporte de un caso

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### ABSTRACT

Zoon vulvitis or vulvitis circumscripta plasmacellularis is a rare chronic inflammatory disease characterized by well-delimited and shiny erythematous plaques that can affect any location of the vulva. It usually occurs in postmenopausal women and poses a difficult differential diagnosis. The need for a vulvar biopsy is emphasized to obtain a certain histological diagnosis. The case of a 36-year-old patient is presented and the current therapeutic options described in the literature are discussed.

**Key words:** Zoon vulvitis, vulvitis circumscripta plasmacellularis,

### RESUMEN

La vulvitis de Zoon o vulvitis de células plasmáticas es una enfermedad inflamatoria crónica infrecuente, caracterizada por placas eritematosas bien delimitadas y brillantes que pueden afectar cualquier localización de la vulva. Suele presentarse en mujeres posmenopáusicas y plantea un difícil diagnóstico diferencial. Se insiste en la necesidad de realizar biopsia vulvar para obtener un diagnóstico histológico de certeza. Se presenta el caso de una paciente de 36 años y se comenta las opciones terapéuticas actuales descritas en la literatura.

**Palabras clave.** Vulvitis de Zoon, Vulvitis circunscrita de células plasmáticas.

### INTRODUCTION

Zoon vulvitis or plasma cell vulvitis is a rare chronic inflammatory disease described for the first time, in 1954, by Garnier and Zoon<sup>(1,2)</sup>. It is characterized by well-defined and shiny erythematous plaques that can affect any location of the vulva, in the form of single or multiple plaques. It does not usually affect the vaginal mucosa or other extragenital locations. It can involve the genital mucosa of the vestibule, the paraurethral epithelium, periclitoid region or the labia minora. The most frequent symptoms are burning, itching, dyspareunia, and dysuria<sup>(3)</sup>. It usually occurs in postmenopausal women and poses a difficult differential diagnosis.

The objective of presenting the case is to substantiate the need for a histological study for the diagnosis of this vulvar dermatosis of exceptional observation in clinical practice, as well as to update the best therapeutic option.

### CLINICAL CASE

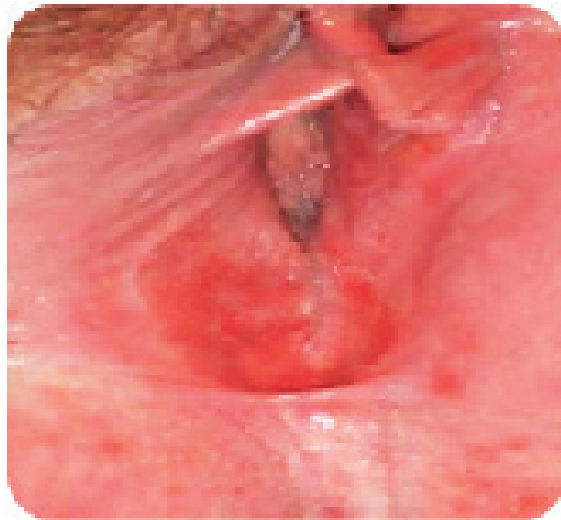
We present the case of a 36-year-old woman with two pregnancies and two eutocic deliveries who underwent surgery for a sacrococcygeal cyst. Rest of history without clinical interest.

The patient referred long-standing chronic vulvar itching, burning, discomfort, and dyspareunia. She had consulted on previous occasions for vulvar itching, not responding to topical moisturizers.

The examination revealed a well-defined erythematous plaque in the labia minora and introitus, approximately 1 cm (Figure 1).



FIGURE 1. ERYTHEMATOUS PLAQUE ON THE LABIA MINORA AND INTROITUS, WELL DEFINED.



The first diagnostic suspicion was vulvar candidiasis, due to the presence of an erythematous plaque. Topical antifungals were prescribed, without obtaining a clinical response.

In the clinical follow-up at one month, given the lack of improvement with topical antifungals, study of the vaginal exudate was performed, which was negative, and a vulvar biopsy (Figures 1 and 2) reported predominantly lymphoplasmacytic vulvitis (Figure 2), corresponding to Zoon vulvitis, an entity characterized by a "pattern of inflammation in the mucosa in relation to chronic dermatoses".

Following diagnosis, topical treatment with high-potency corticosteroids, clobetasol 0.05% cream, three times a day for one month, with subsequent downward regimen, was prescribed twice a day the second month and once a day in the third month. The patient reported a significant clinical improvement in the controls at 3 and 6 months. She is currently asymptomatic, with disappearance of the erythematous plaque.

## DISCUSSION

Zoon vulvitis is a benign lesion of the vulva with a low incidence, its equivalent being Zoon balanitis in men<sup>(1,2)</sup>.

Presenting symptoms are usually vulvar itching, dyspareunia and dysuria<sup>(3)</sup>. On examination, the typical signs are shiny, erythematous plaques, and erosions and telangiectasias may appear on the plaques.

TABLE 1. DIFFERENTIAL DIAGNOSIS OF PLASMA CELL VULVITIS.



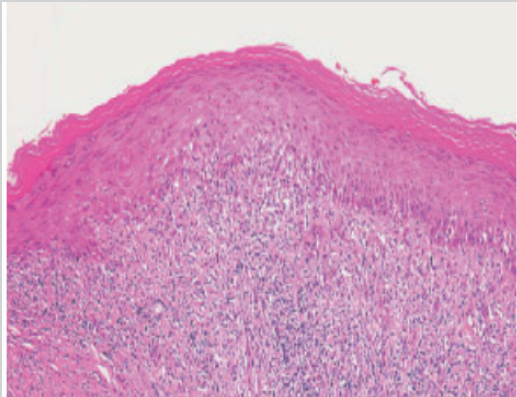
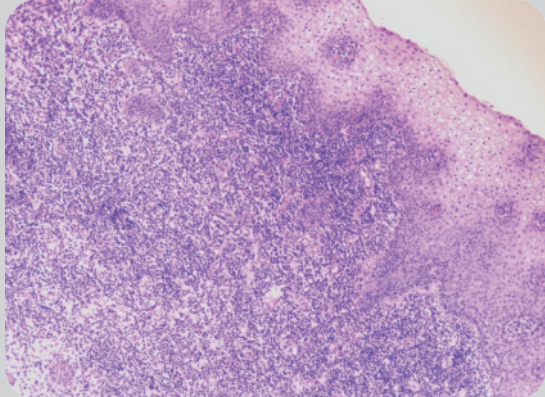
Zoon vulvitis: differential diagnosis	
Vulvar lesions with erythematous macules or plaques	Vulvar lesions with erosions
Candidiasis, atopic dermatitis, lichen simplex chronicus, psoriasis, allergic contact dermatitis, irritant contact dermatitis, lichen planus, tinea cruris, perineal streptococcal infection, vulvar intraepithelial squamous lesion, extramammary Paget's disease, Haley curis disease, tinea versicolor, rosacea pityriasis.	Scratching lesions, fissures, lichen simplex chronicus, herpes simplex, erosive lichen sclerosus, erosive lichen planus, impetigo, irritant contact dermatitis, allergic contact dermatitis, postcoital fissures, herpes zoster, pemphigus, pemphigoid, scar pemphigoid, and toxic epidermal necrolysis.
Lymphoplasmacytic vulvitis usually occurs in postmenopausal women and poses a difficult differential diagnosis.	

Because the symptoms are nonspecific, it is necessary to make a differential diagnosis (Table 1) with other vulvar dermatoses (lichen planus), as well as with systemic diseases (Behçet's disease, Chron's), infections and sexually transmitted diseases (candidiasis, herpes simplex virus, syphilis), precancerous diseases (vulvar intraepithelial neoplasia, Paget's disease) or fixed drug exanthema, being an exclusion diagnosis. Due to the wide range of diseases with which the differential diagnosis must be made, it is mandatory to confirm the diagnosis by biopsy<sup>(4)</sup>.

Perhaps due to the similarity in the presentation of the lesions, the main differential diagnosis should be made with lichen planus, an erosive subtype (Figure 2), this being its most frequent form of presentation (70%). It appears as bright erythematous erosions associated with white or Wickham striae, especially on the labia minora and the vestibule, being relatively frequent the involvement of other locations such as the oral mucosa (forming part of the vulvovaginingival syndrome), the skin or the scalp<sup>(5,6)</sup>. Meanwhile, Zoon vulvitis presents with well-defined and shiny erythematous plaques that can affect any location on the vulva in the form of single or multiple plaques. It does not usually affect the vaginal mucosa or other extragenital locations<sup>(7)</sup>. Recently, it has been proposed that to make the diagnosis of erosive lichen planus, the presence of at least three of the nine clinicopathological criteria (Simpson)<sup>(8)</sup> is required: well-defined erythematous areas, hyperkeratotic border and/or Wickham's striae in the periphery of the lesions, pain/burning, scar changes/loss of normal architecture, vaginal inflammation, involvement of other mucosal surfaces, band



FIGURE 2. MAIN DIFFERENCES BETWEEN ZOON VULVITIS AND LICHEN PLANUS.

Lichen planus	Zoon vulvitis
 <p data-bbox="235 719 794 791">Erosive, well delimited, bright red plaques with a whitish-serpiginous hyperkeratotic border (white or Wickham striae) on the labia minora, clitoris, vestibule and vaginal introitus.</p>	 <p data-bbox="910 732 1275 753">Erythematous plaque in introitus and labia minora.</p>
	
<p data-bbox="428 1221 602 1249"><b>Lichenoid pattern</b></p> <ul data-bbox="298 1268 733 1372" style="list-style-type: none"> <li>Lichenoid interface dermatitis</li> <li>Inflammatory band infiltrate at the dermoepidermal junction</li> <li>Lymphocyte-predominant inflammatory infiltrate</li> <li>Degeneration of the basal epidermal</li> </ul>	<p data-bbox="987 1221 1198 1249"><b>Vasculopathic pattern</b></p> <ul data-bbox="976 1257 1208 1385" style="list-style-type: none"> <li>Dense inflammatory infiltrate</li> <li>More than 50% are plasma cells</li> <li>Endothelial proliferation</li> <li>Red cell extravasation</li> <li>Hemosiderin deposits</li> </ul>

inflammatory infiltrate that compromises the dermoepidermal junction, predominantly lymphocytic inflammatory infiltrate, signs of basal epidermal degeneration.

The etiopathogenesis is unknown, although there are different theories (autoimmune, hormonal, irritative).

The definitive diagnosis is histological and is characterized by the presence of a dense inflammatory infiltrate where more than 50% of the cells are plasma cells<sup>(4)</sup> (Figure 2). There is significant endothelial proliferation, with erythrocyte extravasation and hemosiderin deposits. According to the classification of the International Society for the Study of Vulvar Diseases, ISSVD

2006, plasmacellular vulvitis belongs to the vasculopathy pattern<sup>(8)</sup>, unlike lichen planus, that corresponds to the lichenoid pattern.

Because the etiology is unknown, multiple different treatments have been chosen, the response to them being inconsistent. The first line of treatment would be high-potency topical steroids such as clobetasol propionate 0.05%, with surgery being the treatment of choice in highly refractory cases<sup>(9)</sup>.

Among the medical treatments used we find first-line treatments such as high-potency topical corticosteroids<sup>(10)</sup> and medium-strong topical corticosteroids, second-line treatments such as calcineurin inhibitors, tacrolimus<sup>(11)</sup>. Imiquimod,



interferon alfa, and even cryotherapy have also been used<sup>(12)</sup>. If there is no clinical response to previous treatments, excisional CO2 laser treatment can be used and even excisional treatment in the operating room<sup>(13)</sup>.

Given that plasma cell vulvitis is a chronic inflammatory disease and is often associated with periods of remission and relapse, clinical follow-up of the patient is essential.

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