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Case Report

Zoon´s Vulvitis: A Rare Differential Diagnosis of Vulvar Pruritus

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A B S T R A C T

Zoon´s vulvitis, also known as plasma cell vulvitis (PCV) is a rare inflammatory disorder of the female genital tract, which typically affects the vulva. Its real prevalence is supposed to be underreported as typical symptoms can mimic and be misdiagnosed as other more common vulvovaginal disorders. It usually presents as a well-circumscribed and erythematous patch/macule that can cause itching, burning, dysuria and dyspareunia. Histologically, it is characterized by a thinned epithelium with infiltration of more than 50% of polyclonal plasma cells in the underlying dermis. Here we present a 73-year-old patient referred to a gynecology consultation due to complaints of persistent vulvar itching and discomfort for the past 6 months. She was initially treated with topical antifungal and a course of corticosteroid therapy with persistence of complaints. On examination, she had an ecchymotic macule on the anterior one-third of the transition between the left labia minora and the labia majora. This lesion was biopsied and a Zoon´s vulvitis diagnosis was confirmed. Currently, she maintains topical estrogen therapy and regimens of topical corticosteroids when the condition worsens. Vulvovaginal erythema and/or pruritus may be caused by a range of conditions that varies from infectious, immunological, or even malignant causes. In cases of resistance to treatment or persistence of lesions it is therefore essential to perform a biopsy in order to avoid delays in instituting therapy and potential long-term complications. Although rare, this condition can cause serious discomfort to patients; therefore, more research is needed to establish its most cost-effective management.

Keywords: Zoon vulvitis; Plasma cells; Pruritus; Vulva

1. Introduction

Zoon´s vulvitis, also known as plasma cell vulvitis (PCV) is an inflammatory disorder of the female genital tract, which was first described by Zoon in the 1950s. Its true etiopathogenesis is unknown, although a variety of triggering factors have been hypothesized (autoimmune, irritative, hormonal). It typically affects the vulva, but there are also several case reports of vagina and cervix´s involvement by this condition[1-5]. Its real prevalence is not known, but it´s supposed to be underreported as its typical symptoms can mimic and be misdiagnosed as other more common vulvovaginal disorders, such as lichen sclerosus, lichen planus, genitourinary syndrome of menopause, contact dermatitis or infectious vaginitis[6-11].

It usually presents as a well-circumscribed and erythematous patch/macule with a fainted red-orange hue involving the vulvar vestibule, periurethral area and labia minora and majora; it can cause itching, burning, dysuria and dyspareunia. When the vagina is affected, it can trigger a yellow leukorrhea-type discharge. Some patients can also be completely asymptomatic. Histologically, it is characterized by a thinned epithelium with infiltration of more than 50% of polyclonal plasma cells in the underlying dermis along with diamond shaped keratinocytes and extravasation or hemosiderin deposition[1-4,6,12,13].

The condition is often difficult to treat, and standard treatment has not been established[9,10]. It may include topical, oral, intralesional and surgical options in refractory cases.

2. Materials & Methods

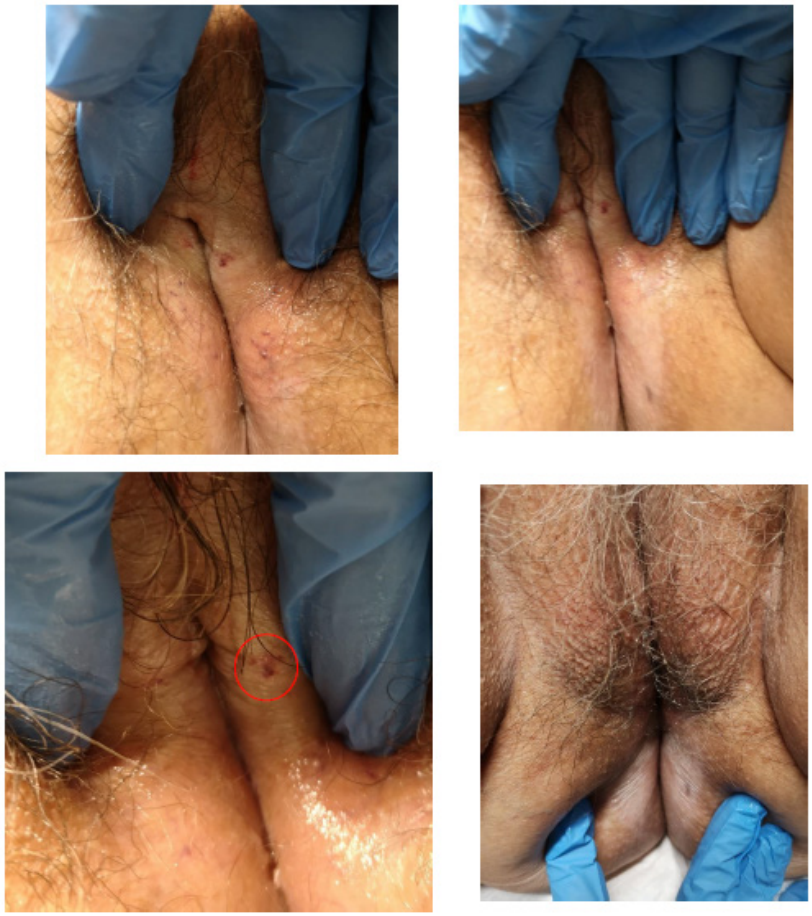
We present a case of Zoon´s vulvitis which was treated with corticosteroid therapy resulting in symptom relief.

3. Results & Discussion

A 73-year-old patient was referred to a gynecology consultation due to complaints of persistent vulvar itching and discomfort for the past 6 months. The patient referred dyspareunia and burning but denied any vaginal discharge. When symptoms first began, she was evaluated by her primary care provider, who prescribed topical antifungal, a course of corticosteroid therapy (clobetasol 0.05% ointment) along with an antihistamine (25mg hydroxyzine pills). She was also recommended to wear looser-fitting clothing. However, despite a partial initial improvement, itching persisted, and it got worse over time.

The patient had a personal history of hypertension and dyslipidemia and was taking medication for both. She has had a hysterectomy with bilateral salpingo-oophorectomy at 49 years old because of abnormal uterine bleeding conditioned by uterine leiomyomas. The patient had no known drug allergies.

On examination, the patient presented marked vulvovaginal atrophy, with a whitish color along both labia minora, inner half of the labia majora and extending to the perianal region. In the anterior one-third of the transition between the left labia minora and the labia majora, the presence of a 4mm ecchymotic macule was notable, and this location corresponded to the area in which the patient reported the greatest intensity of complaints. There was a similar, but smaller lesion on the right side **(Figures 1-4)**.



Figures 1-4: Clinical aspect of the patient´s vulva; the circle delimits the area that was initially biopsied.

At this time, another course of corticosteroid therapy was attempted, and the patient began topical estrogen therapy. Testing for bacterial vaginosis and the most common sexual transmitted diseases [trichomonas, chlamydia, gonorrhea, human immunodeficiency virus (HIV), syphilis, and hepatitis] were also done. The results were all negative.

She came back eight weeks later, and the same complaints persisted. The lesion previously described remained like the first description. In this sense, it was decided to carry out a punch biopsy at this specific location. A 9mm length cylindrical punch was obtained, which showed plasmacytosis mucosae consistent with Zoon´s vulvitis.

The patient remains under follow-up at a gynecology consultation, complaining of occasional itching. She maintains topical estrogen therapy and regimens of topical corticosteroids when the condition worsens.

Vulvovaginal erythema and/or pruritus may be caused by a range of conditions that varies from infectious, immunological, or even malignant causes[6,7].

Our patient was older than the average age of patients reported to have this condition (between 52 and 55 years old)[14-16]. This fact, together with the greater probability of other conditions in this age group, may have contributed to some delay in diagnosis.

As performed in this case, patients with severe and unresponsive symptoms should undergo tissue diagnosis to guide the most appropriate treatment. A plasma cell inflammatory infiltrate is the most common finding on histopathology, which was consistent with our results.

As described by Krapf et al.´s review, the most common treatment modalities for this disorder includes topical corticosteroids and immunomodulation with tacrolimus and imiquimod, with 88% of patients achieving symptom resolution[16].

4. Conclusion

This case demonstrates the importance of performing a biopsy in timely diagnosis and treatment, as missed diagnosis can result in delays in instituting therapy and potential long-term complications.

It is advised that patients have regular clinical follow-up as periods of remission and relapse are frequent; although there are no reports of malignant changes of Zoon´s vulvitis cases, moderate dysplasia has been described[17,18].

Although rare, this condition can cause serious discomfort to patients; therefore, more research is needed to establish its most cost-effective management.

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Fernanda Alves contributed to the conception of the case report, acquiring the data and undertaking the literature review and drafting the manuscript.

Both Mara Rocha and Ana Moreira contributed to drafting the manuscript and undertaking the literature review.

All authors contributed to revision of the manuscript and approved the final submitted version.

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