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Case Report Open Access

Plasma Cell Vulvitis: A Case Report and Review of Literature

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Abstract

Plasma cell vulvitis (PCV) is a rare disease affecting anogenital skin. It is also known by multiple names including Zoon's vulvitis and vulvitis circumscripta plasmacellularis. It is an inflammatory condition that can cause severe local discomfort and psychosocial distress. The diagnosis is challenging and often remains unrecognized. It usually presents as a glistening, red, well-demarcated plaque or patch. The pathophysiology is unknown. Treatment recommendation is mostly based on case reports and case series and this condition is often recalcitrant to topical and systemic treatment.

Keywords: Plasma cell vulvitis; Clinical presentation; Histopathology; Treatment

Introduction

PCV is a rare disease of anogenital skin defined by morphological appearance and histology [1]. It is a benign inflammatory condition but can cause local discomfort and is often refractory in its course [2]. The pathophysiology is unknown, though herpes simplex has been cited as a possible etiology [3]. The condition usually presents as a glistening, red, well-demarcated plaque or patch. The pathology often reveals characteristic findings including sub epithelial band-like infiltrates with plasma cell predominance. However, the diagnosis remains challenging and often unrecognized. Treatment recommendation is based mostly on case reports and case series and this condition is often recalcitrant to topical and systemic treatment. Presented is a case revealing the psychosocial ramifications of PCV in addition to the physical suffering brought on by this disease. The case exhibits the importance of considering PCV in a patient with refractory vulvar pain and red well-defined plaques on physical exam.

Case Report

A 43 year old Caucasian woman presented with vulvar burning for 2 years. She had been treated unsuccessfully with topical clobetasol, triamcinolone and fluocinonide ointments and oral prednisone. She endorsed moderate vulvar pain, vaginal discharge and bloody tinge on the toilet paper with wiping. Patient acknowledged stress within marriage and status of apareunia because of the pain. Physical exam revealed well-defined rusty-red plaques with superficial erosions at vaginal introitus (Figure 1). The lesions were tender to palpation with Q-tip. Wet prep showed normal pH, negative for yeast hyphae or trichomonas. Herpes simplex and herpes zoster PCR were negative. Bacterial and fungal cultures were negative. Pathology revealed a sub-epithelial band-like infiltrate composed of lymphocytes and numerous plasma cells (Figure 2). There was mild epidermal acanthosis, diffuse spongiosis, and dermal fibrosis. The clinical pathological correlation supports a diagnosis of plasma cell vulvitis (PCV).

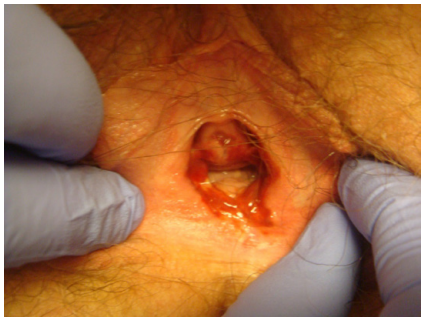

Figure 1: Rusty red, glistering well demarcated plaques of plasma cell vulvitis.

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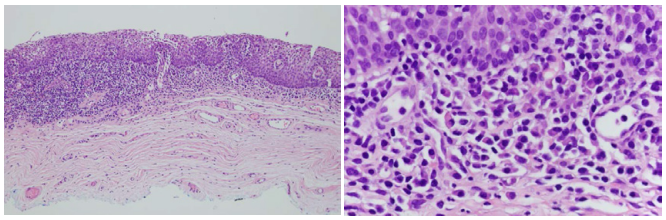

Figure 2: Mucosa epithelium with neutrophilic spongiosis and an underlying band-like infiltrate composed of lymphocytes and numerous plasma cells.

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Discussion

PCV can be asymptomatic or it can present with vulvar soreness, burning, pruritus, and dyspareunia [4-7]. In the present case and another study, apareunia was reported as a result of severe vulvar soreness from PCV [6]. An observational cohort study of 36 patients with PCV found that burning was the most frequent symptom (80.6%) out of the 83.3% that complained of symptoms [4]. PCV appears as glistening, deep to rusty red, well demarcated plaques or patches, solitary or multiple [4,6-7]. These lesions are easily eroded and may bleed [5,6]. Also described in case reports is the presence of an orange hue around the lesion [4,6]. The clinical differential diagnosis for PCV includes squamous cell carcinoma, lichen planus, herpes simplex, psoriasis, and candidiasis.

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Biopsy is necessary to confirm the diagnosis of PCV. One review of vulvar dermatoses found 8 out of 15 cases of PCV were originally diagnosed as lichen planus, lichen sclerosis, psoriasis, or eczematous dermatitis [2]. Misdiagnoses may occur because some specimens lack the classic PCV histologic elements. This could also be attributed to coexistence of PCV with lichen sclerosis [8]. PCV was also reported to develop after successful treatment of lichen planus [1]. As the name suggests, PCV usually presents with a subepithelial band-like infiltrate with plasma cell predominance. However, there are cases of PCV with minimal plasma cells [9].Thus, it is important to know the other characteristics seen on biopsy. Other histologic features of PCV include 'lozenge shaped' (diamond shaped) keratinocytes, spongiosis with vascular dilatation, and hemosiderin deposition [2-7,10-14]. Though, one retrospective evaluation of PCV biopsy specimens reported that "lozenge-shaped" keratinocytes were rarely seen [9]. A recent histopathologic review of vulvar dermatoses revealed "basal keratinocytic crowding" to be a new finding significantly associated with PCV. Basal keratinocytic crowding describes crowded, small, uniform basal keratinocytes [1].

Although the morphologic and pathologic features have been well described, patients undergo antibiotic, anti-viral, and other treatments before a diagnosis of PCV is made. Topical and oral steroids are often tried first with varying results. High potency topical steroids have been shown to be effective in some cases [5,12]. With most of the literature consisting of case reports, there is a scarcity of evidence comparing treatments of PCV [3], and currently no therapeutic guidelines exist.

However, one recent study compared topical immunomodulatory and anti-inflammatory treatments for PCV [13]. Three topical therapies were compared for efficacy in relieving symptoms and signs of PCV: Fusidic acid 2%/betamethasone valerate 0.1% cream, clobetasol propionate 0.05% ointment, and tacrolimus 0.1% ointment. The study found no statistical difference among treatment groups. All patients in each treatment group had improvement in symptoms at each follow up visit. Disease symptom response was significantly better than clinical signs in all treatment groups. One patient had to discontinue fusidic acid/betamethasone treatment due to increased itching and another patient had to discontinue tacrolimus due to "burning and itching."

Tacrolimus and other topical immunomodulatory agents have been described in case reports and are often tried after unsuccessful steroid use [3]. In a four patient case series evaluating the efficacy of PCV treatment with 0.1% tacrolimus, one patient reported complete resolution and another reported good resolution of symptoms [14]. The patient with complete resolution had to stop treatment after 16 weeks due to onset of pruritus and burning. In another case series, two patients applied imiquimod 3-7 times weekly for at least 2 months [15]. One remained disease free at 6 months of follow up and the other required 1 month of therapy after a recurrence. Both patients experienced erythema at application site. Another case series where two patients applied imiquimod 5% revealed complete resolution of the lesions by the 9th week. Both patients experienced irritation at site of application [16]. Another patient treated with imiquimod experienced intense pain and swelling after three overnight applications. However, she was able to tolerate the imiquimod when applied for only 30-60 minutes. After 2 months of treatment the lesion disappeared [17]. Intralesional interferon alpha was used successfully in one case, which had also been positive for herpes simplex antigen [3]. Topical misoprostol has also been shown to be effective in resolution of symptoms and lesions, though lower doses were necessary to avoid local irritation [18]. Topical immunomodulatory therapy is an option for patients who have failed other topicals such as steroids. However, no regimen has been established as superior but rather must be determined by patient tolerance of any adverse effects.

For refractory cases of PCV, surgery to remove the area of disease has been described in two case reports with subsequent resolution of symptoms. One patient with vulvar pruritus resistant to treatment with numerous agents including antifungals, clobetasol, estradiol, and intralesional triamcinolone was found to have PCV on biopsy [19]. She received intralesional injections of interferon alpha-2b (Intron A) without improvement. The patient then underwent surgical resection of the lesion and afterwards became symptom free. Another patient with refractory PCV failed oral prednisone and multiple topical therapies, including tacrolimus 0.5%, silver sulfadiazine, estrogen cream and 5-flurouracil. She was then given three courses of laser ablation therapy, which had been reportedly successful in another case but only gave her brief symptomatic relief. Finally, a modified posterior vulvectomy was performed and the patient had complete resolution of her symptoms [20].

Conclusions

Though the clinical presentation and the pathology of PCV are well described in many case reports, therapeutic studies are lacking. Physicians should consider PCV when evaluating vulvar dermatoses. Further studies on the treatment of PCV are needed in order to develop guidelines for practitioners to manage this often recalcitrant disease. Although rare, PCV can cause serious discomfort and psychosocial distress to patients and warrants further investigation.

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