Vulvitis plasmacellularis: two new cases

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Abstract

Vulvitis chronica plasmacellularis or Zoon's vulvitis is a rare benign circumscribed inflammation of the vulvar mucosa. It is found in women ranging in age from 26 to 70 years. Shiny, macular erythematous lesions, which are irregular in shape and sharply marginated are usually observed. The histologic findings show chronic subepithelial dense inflammation composed largely of plasma cells. We here report two cases of vulvitis plasmacellularis with typical clinical manifestations, courses and histopathologic findings.

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Introduction

In 1952 Zoon¹ first described under the name *balanitis chronica circumscripta plasmacellularis* a chronic inflammation of the glans penis or inner surface of the foreskin, clinically resembling the erythroplasia of Queyrat, histologically characterised by an infiltrate of plasma cells. This plasmacellular infiltrate was considered by Zoon to be an essential criterion for the diagnosis. Analogous lesions localised on the vulva are rare; a benign erythematous circumscribed vulvitis with plasmacellular infiltrate was subsequently reported by Garnier in 1954²; since then only 23 other cases of vulvitis chronica plasmacellularis (VP) have been described.²⁻¹⁰ We report here two cases of VP, recently observed in our department.

Case reports

Case 1: A 52 year old woman was referred to our department complaining of pruritus in the vulvar region, which had been present for one year. Physical examination revealed the presence of an erythematous patch, with haemorrhagic spots, approximately 2 cm in diameter and with an irregular edge in the vaginal introitus (fig 1). The remainder of the physical examination was negative. Direct microscopic examination was negative and culture for *Candida albicans* produced no growth. Colposcopy and 6% acetic acid test revealed no lesions associated with human papilloma virus (HPV). A biopsy of the patch was performed and the histological findings showed focal acanthosis and exocytosis of the epithelium and in the tunica propria vascular proliferation with dilatation of vessels and a dense, predominantly band-like infiltration composed of many plasma cells, lymphocytes and extravasated erythrocytes (fig 2). A diagnosis of vulvitis chronica plasmacellularis was made. The patient was treated with topical corticosteroids with only a mild clinical and symptomatic improvement.

Case 2: A 61 year old woman was referred to us for the evaluation of some very pruriginous fixed erythematous patches of the vulva. These lesions had been present for four years and after initial phases of partial remission had become stable. Many local therapies with antifungals and emollients produced only a transient symptomatic improvement. Physical examination of the medial surfaces of labia minora revealed four dark-red, fixed, glistening patches, with a diameter ranging from a few millimetres to 1 or 2 centimetres. Direct microscopic examination was negative and

Figure 1 Dermatological examination of the vulva shows a fixed, glistening erythematous patch.

Figure 2 Focal acanthosis and exocytosis of the epithelium. Band-like infiltration composed of many plasma cells, lymphocytes and extravasated erythrocytes in the tunica propria. (H and E, $\times 6$)

culture for *Candida albicans* produced no growth. Neither colposcopy nor acetic acid test revealed lesions due to HPV. Histological findings in a 4 mm punch biopsy specimen of one of the lesions showed a pattern analogous to the one observed in case 1. Hence a diagnosis of vulvitis chronica plasmacellularis was made. A corticosteroid of class III therapy was administered. Topical application twice daily for one month produced a marked symptomatic improvement without clinical modifications.

Discussion

VP or Zoon's vulvitis is a rare chronic inflammation of the vulvar mucosa, described in women ranging in age from 26 to 70 years and never observed in prepubertal girls. VP is characterised by one or more fixed and not infiltrated shiny erythematous plaques. The lesions, which are irregular in shape and sharply margined, are more often bilateral and symmetrical and tend to join up progressively. Both dark-red¹¹ or ecchymotic^{2 12 13} patches, due to the presence of abundant haemosiderin in the chorion, and teleangiectatic and purpuric^{14 15} varieties are described as well as the typical form. Less frequently erosive lesions^{16 17} and granulomatous varieties with cayenne pepper spots¹⁸ are reported. Elective sites of involvement are labia minora and majora, clitoris, fourchette, urethral meatus and vaginal meatus. The usually asymptomatic disease tends to persist for many years. Patients sometimes complain of pruritus, burning and dyspareunia. The typical histological pattern is characterised by a dense inflammatory band-like infiltrate parallel to the basal membrane, situated in the upper and middle chorion, predominantly composed of plasma cells. A vascular proliferation with dilatation of capillary vessels presenting thickening of the walls and at times some extravasated erythrocytes are also observed. In some cases the number of plasma cells seen in the infiltrate may be low and for this reason some authors do not consider VP as a distinct entity, but as a non specific inflammatory pattern due to exogenous agents such as infections or chronic irritations; the name "Zoon's vulvitis" is therefore preferred.^{15 19-21} The most important differential diagnosis includes erythroplasia of Queyrat; clinically puzzling cases are diagnosed histopathologically. Moreover VP must be differentiated from benign diseases such as candidiasis, fixed drug eruption, allergic contact dermatitis, pemphigus vulgaris and herpes simplex infection. Unsuccessful results in treatment of VP both topically (antiseptics, steroids, antibiotics, emollients, hormones^{22 23}) and systemically (hormones^{24 25} and steroids^{18 26}) have been reported in the literature. Good results with intralesional interferon alpha treatment have been reported in an atypical case of VP, with a vegetating clinical appearance. Destructive procedures (electrocoagulation, CO₂ laser, surgical excision, radiotherapy) are nearly always followed by

recurrence.^{1 2 27} Progression to neoplasia has never been reported. A moderate epithelial dysplasia with hyperchromatic voluminous nuclei was reported by Vilmer *et al*⁷ in repeated biopsies of three patients followed up for periods ranging from 7 to 13 years. This dysplasia, not associated with modifications of the connective infiltrate or clinical variations of the lesions, appeared some years after the onset of VP in these three cases: in one case dysplasia remained stable at follow up and in two cases it resolved. The authors did not consider VP a precancerous condition and ascribed the epithelial alterations only to the chronic inflammation of the dermis. However, they suggested carrying out a clinical and histological follow up of the lesions as well as examinations to exclude HPV infections or other conditions of cervicovaginal dysplasia which might be associated. We think that fixed erythematous lesions of the vulva should be biopsied to exclude erythroplasia of Queyrat. When a diagnosis of VP is made, a follow up with careful examinations should be performed. In patients with a duration of the disease over several years it might be advisable to repeat a biopsy to exclude the onset of epithelial dysplasia. Finally periodic gynaecological examinations should be made to exclude a concomitant HPV infection or other cervicovaginal dysplastic conditions which could promote the degeneration of VP, acting as cocarcinogenic factors.

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