Case Report

Plasma cell vulvitis

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Abstract

Plasma cell vulvitis is a very rare inflammatory disorder of vulva, characterized by a bright-red mucosal lesion of significant chronicity, which may be symptomatic. Very few case studies of this condition are reported in literature. We describe one such classical patient, who presented with slight dyspareunia. The diagnosis was confirmed on histopathological examination. It is important for clinicians to accurately diagnose this alarming condition in time.

Key words: Dyspareunia, plasma cell vulvitis, vulvitis circumscripta, Zoon's vulvitis

INTRODUCTION

Plasma cell vulvitis (PCV) is an extremely rare idiopathic inflammatory disorder of female genitals. Less than 50 cases have been reported in English literature.[1,2] It usually presents as a chronic itchy mucosal condition affecting the vulva, with or without dyspareunia. Histopathologically, it is characterized by dense plasma cell infiltration in the subepithelial zone of the affected skin. It may mimic vulval intraepithelial neoplasia.[1] In presenting this case, we aim to draw the attention of dermatologists, venereologists and gynecologists for having a high index of suspicion on this entity, which might often be misdiagnosed.

CASE REPORT

A 44 year old female patient presented with minimally itchy but frequently annoying red patch on the vulva of 6 months duration. It was not accompanied by pain or bleeding. She was otherwise healthy with unremarkable gynaecological and obstetrical history and had 2 children born out of full term normal delivery. There was no history of surgical procedures or local trauma in past. Her general examination did not reveal any abnormality. There were no major illnesses in past or family. There was no history of dysuria in her but soreness and dyspareunia were significant complaints.

Clinical examination revealed an ill- defined, bright erythematous, slightly elevated plaque on the inner surface of left labia minora almost encircling the urethral meatus [Figure 1]. There were no fissures, excoriations or oozing. The lesions were non-tender but she was quite apprehensive while palpating the affected area. There was no regional lymphadenopathy. Differential diagnoses of erosive lichen planus, genital psoriasis, lichen sclerosus et atrophicus and erythroplasia of Queryat (squamous cell carcinoma *in situ*) were considered.

Her investigations including hemogram, blood sugar and urinalysis were all within normal limits. Venereal disease research laboratory and HIV antibodies were non-reactive. Her biopsy revealed atrophic epithelium, dense lichenoid

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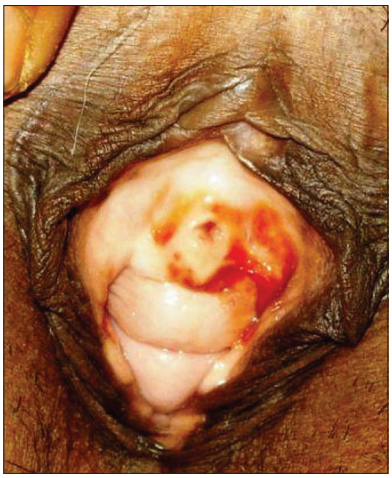

Figure 1: Bright red erythematous plaque chiefly on left labia minora almost encircling the urethral meatus giving a peculiar lacquer paint appearance

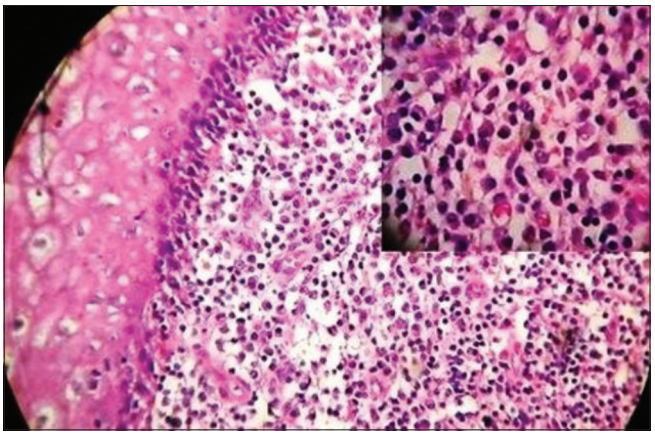

Figure 2: Histopathological examination shows dense infiltrate of plasma cells in the upper dermis. Inset shows high power view revealing abundant plasma cells and extravasation of red blood cells

infiltrate in upper and mid dermis with abundant plasma cells in upper dermis as well as around vessels. There was vasodilatation with extravasation of erythrocytes [Figure 2]. There was no basal cell vacuolation. These findings confirmed the diagnosis of PCV. The patient was prescribed topical mid-potent steroid as mometasone fuorate 0.1% cream twice daily application. After 3 weeks, she reported with significant relief in her itching, soreness and dyspareunia. Her examination revealed reduction of erythema, flattening of plaque and reduced apprehension. The frequency of topical steroid application was reduced to once a day and gradually stopped after 4 weeks. There was complete clearing. She remained symptom-free for next 2 months and later, she was further lost to follow up.

DISCUSSION

PCV or Plasmacytosis mucosae et vulvitis is a rare condition, which may involve penis (balanitis circumscripta plasmacellularis, Zoon's balanitis), vulva (vulvitis circumscripta plasmacellularis, Zoon's vulvitis), lips (plasma cell cheilitis), and other mucosal surfaces like oral cavity (plasma cell orificial mucositis, atypical gingivostomatitis, plasmacytosis circumorificialis).[2] PCV, also termed Zoon's vulvitis, Vulvitis circumscripta plasmacellularis, is an extremely rare, idiopathic vulval dermatosis with <50 cases reported in the literature.[1,2] It usually occurs between fifth to eighth decades. It may be asymptomatic or may be associated with symptoms such as pruritus, stinging, burning sensation, dysuria and dyspareunia.

PCV may be the cause of intractable vulvar pruritus.[2] These lesions usually take the form of solitary asymptomatic or itchy, sharply defined red-brown glistening flat but barely palpable patch with "lacquer paint" appearance measuring 1-3 cm diameter. Tiny petechial hemorrhages may be seen. Swelling, discharge, crusting, and similar signs of inflammation are absent.[3] A tumorous variant (plasmoacanthoma) has also been described.[2] The exact etiology is not known but viral (herpes simplex), autoimmune, hormonal or irritant (poor hygiene, sweating, trauma, persistent friction) factors are implicated.[1,2] It is most probably a non-specific chronic, reactive, principally irritant mucositis.[2] Histological features are characterized by dense band-like, predominantly plasma cell infiltrates in the upper dermis, which may extend to the level of the mid-reticular dermis. The number of plasma cells seems to vary with the stage of the disease.[2] There may also be "lozenge- shaped" keratinocytes[4], lymphocytes, mast cells, occasional eosinophils and neutrophils.

Blood vessels are dilated, prominent, may exhibit extravasation of erythrocytes and hemosiderin deposition. With time, fibrosis may ensue within the lesion. The overlying epidermis is usually attenuated, spongiotic but ulceration may rarely occur. A similar condition in men, Plasma cell balanitis, is found almost exclusively in uncircumcised men.

A differential diagnosis of contact dermatitis, erosive lichen planus, lichen sclerosus et atrophicus, pemphigus, mucous membrane pemphigoid, lupus erythematosus, drug reaction, extramammary, Paget's disease, genital psoriasis and squamous cell carcinoma should be considered.[1,2] Mucosal biopsy from the affected area is usually helpful in arriving at the accurate diagnosis. It is important to note that,

the lesions of PCV are chronic, but generally not premalignant.[1,3]

Treatment options include mid-potency topical steroids, topical calcineurin inhibitors like 0.1% tacrolimus ointment or 1% Pimecrolimus cream, topical immunomodulator like imiquimod have been attempted with varying success. Surgical excision, liquid nitrogen cryotherapy and carbon dioxide laser ablation may help in certain individual.[1,3,4] Our case responded very well to topical steroids, but long-term follow-up is desirable in such cases to ascertain the entirely benign nature of the disease.

CONCLUSION

PCV should be considered in a patient complaining of long standing vulvar itching with persistent erythematous mucosal plaque(s).

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Conflicts of interest

There are no conflicts of interest.

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