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CASE REPORT

Plasma cell cheilitis in an elderly female: A case report

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ABSTRACT

INTRODUCTION Plasma cell cheilitis (PCC) is a rare, chronic inflammatory dermatitis of unknown etiology. Due to the limited number of cases reported, no guidelines have been established for its treatment. We present a case of PCC clinically similar to actinic cheilitis or mucosal lichen planus, and squamous cell carcinoma but showed response to topical tacrolimus 0.1% ointment.

CASE REPORT A 62-year-old female with extreme fondness to piping hot food presented with a solitary painful ulceration with some pustules and bleeding on the lower lip with three (3) months duration. Skin punch biopsy revealed a dense band-like infiltrate of plasma cells which is consistent with Plasma cell cheilitis. The patient was given tacrolimus 0.1% ointment and showed significant improvement after a month of treatment.

CONCLUSION PCC is a rare condition that should still be considered in patients presenting with persistent cheilitis. Clinical and histological correlation is advised for proper management and prognostication.

KEYWORDS cheilitis, persistent ulceration, plasma cell, tacrolimus

INTRODUCTION

Plasma cell cheilitis (PCC) is a rare, chronic inflammatory dermatitis of unknown etiology. It is commonly found in elderly males presenting as an erosive plaque or patch with fissures, bleeding, and crusting on the lip mucosa.[1,2] Due to similar presentation to other causes of cheilitis, misdiagnosis is common. Differential diagnoses noted in literature include common causes of cheilitis like infective cheilitis (bacterial, fungal, herpes simplex virus) due to immunocompromised states, actinic cheilitis, squamous cell carcinoma and those associated with systemic diseases such as lichen planus.[3]

Due to the limited number of cases reported, no guidelines have been established for the management of this condition and use of traditional topical steroids has shown varying results.[1] Aside from treatment with oral steroids, intralesional steroid injections, systemic griseofulvin, and topical tacrolimus and pimecrolimus have been explored.[4,5] We present a case of PCC clinically similar to actinic cheilitis, mucosal lichen planus, and squamous cell carcinoma. Our case showed partial remission with topical tacrolimus 0.1% ointment.

CASE SUMMARY

A 62-year-old, non-smoker, businesswoman from Santo Tomas, Pampanga, who came in due to a painful ulceration (8/10 pain scale) with pustules and bleeding on the lower lip of three (3) months duration. Her medical and sexual history was generally non-contributory and the patient noted frequent consumption of piping hot food. The patient claimed to stay mainly indoors and did not use any form of sun protection daily. The initial presentation was a solitary, well-defined erosion with pustules and pinpoint bleeding on the lower lip (Figure 1A).

The patient was initially treated by another physician for herpes simplex infection with fungal superimposition. She was initially given oral acyclovir 400mg every eight (8) hours for seven (7) days, 0.9% sodium chloride compress, miconazole cream, and betamethasone cream twice a day for two (2) weeks. There was a noted complete resolution of pain, however, only partial improvement on the lesions on the lower lip was observed. Hence, the patient decided to seek

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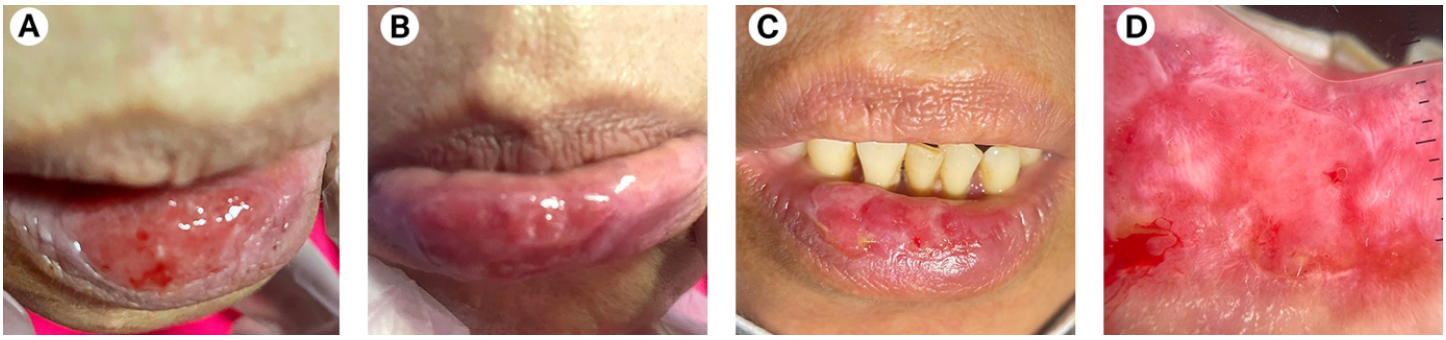

Figure 1. A. Initial clinical presentation before seeking consultation. B. Partial response to initial treatment (given by another physician) with oral antiviral, 0.9% sodium chloride, miconazole cream, and betamethasone cream. C. Clinical photo prior to biopsy. D. Dermoscopy prior to biopsy.

for a second opinion. Upon consultation, physical examination showed a solitary and well-defined erythematous erosion on the lower lip with no Wickham striae (Figure 1B). Skin punch biopsy and syphilis serology was advised. However, due to financial constraints, the patient was lost to follow-up. Two (2) weeks after initial consult, the patient came back for follow-up due to an increase in the size of the erosion and recurrence of pain (7/10) (Figure 1C). Dermoscopy of the lower lip showed a well-defined erosion with milky red structureless areas and multiple linear vessels on the periphery (Figure 1D). These findings were consistent with the diagnosis of PCC.

Rapid plasma reagin (RPR) and venereal disease research laboratory (VDRL) test results were nonreactive. Histopathological examination showed a dense band-like infiltrate of plasma cells, lymphocytes, scattered neutrophils and eosinophils seen in the upper dermis. No hypergranulosis, colloid bodies, solar elastosis, squamous islands, or atypical cells were seen (Figure 2). Histologic findings were consistent with PCC. The patient was subsequently treated with tacrolimus 0.1% ointment twice a day. After a month of daily treatment, significant improvement was observed on the lesions on the lower lip (Figure 3).

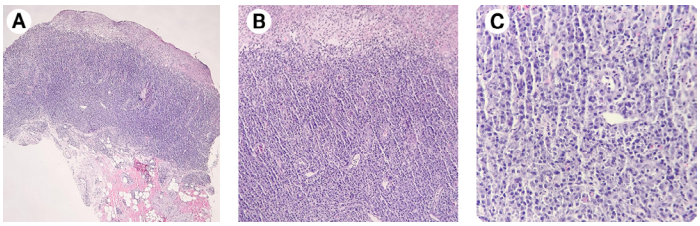

Figure 2. A-C. Histopathology showing dense band-like infiltrate of plasma cells, lymphocytes, and scattered neutrophils and eosinophils.

DISCUSSION

PCC is a rare benign inflammatory condition that was first identified by Zoon in 1952 as the oral counterpart of plasma cell balanitis. PCC may also develop in other orofacial areas such as the vulva, buccal mucosa, tongue, epiglottis, and larynx.[6]

PCC commonly affects the lower lip and presents clinically as erosions, ulcerations, or fissures with crusting or bleeding.[1,6] In a study done by Lee et al, in 2017, the most common primary presentation was a patch accompanied by erosion and the most common symptom reported was pricking.[2] Our patient presented a history of painful erosion on the lower lip for three (3) months similar to the findings described in the literature. However, due to its general appearance, symptoms, and rarity, the diagnosis of PCC may be missed initially. Dermoscopy is a useful tool that can help further investigate cheilitis. Findings of a well-defined border, milky red areas and radial distribution of linear vessels on the periphery, absence of stellate border, and scales at the periphery (suggestive of actinic cheilitis) or Wickham striae (pathognomonic of oral lichen planus) have been identified as important clues in the diagnosis of PCC.[7,8]

Our patient presented with a histology of dense band-like infiltrates with mature plasma cells on the subepithelia consistent with the established literature.[1,9] Plasma cells seen in histopathological findings may also be found in conditions like oral lichen planus, syphilis, and lymphoma. It is important to note that our patient did not show features of hypergranulosis, subepithelial chronic lymphocytic inflammatory infiltrate, and colloid bodies on biopsy. These findings, if present, may point more to a diagnosis of oral lichen planus.[10] Direct immunofluorescence (DIF) can be done to highlight the presence of colloid bodies in these cases. For lymphomas, if a high index of suspicion is present, special

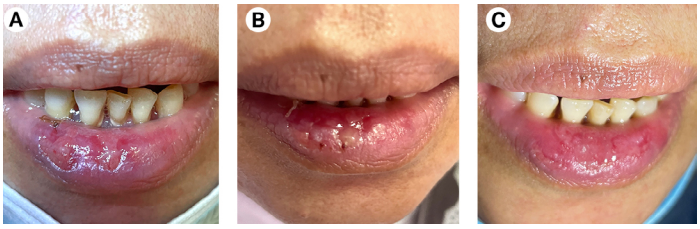

Figure 3. A. 1 week post treatment B. 2 weeks post treatment C. 1 month post treatment

stains for CD 19, CD 20, CD 5, and CD 10 may be done.[11] As for syphilis, the patient presented with a clinically non-contributory sexual history as well as non-reactive RPR-VDRL results. However, a biopsy specimen could be sent for further testing with Warthin-Starry stain to visualize spirochetes if present. Our patient's specimen was not sent for further testing as the correlation of history, physical examination with dermoscopy, results of syphilis serology, and biopsy findings all pointed strongly to clinching the diagnosis of PCC.

Lugović-Mihić et al., proposed a classification for cheilitis that can help simplify the approach to the diagnosis and management of similar cases.[12] The timing, reversibility, and association with other systemic diseases must be carefully considered. Given the presentation and development of persistent cheilitis seen in our patient, there is a need to take into account the following differential diagnoses: actinic cheilitis, squamous cell carcinoma, and PCC. A biopsy is warranted in order to come up with a definitive diagnosis. Finding a band-like infiltration of mature plasma cells in the upper dermis is necessary for the diagnosis of PCC.[2,6,12]

The etiology of PCC is still unknown. However, there have been studies that suggest associations with mechanical damage or accumulated solar damage.[9] As seen similarly in other published studies, our patient presented with lesions on the lower lip. The lower lip is commonly the focal point of accumulated sun damage or mechanical trauma.[6] Although our patient claimed to have no significant occupational or recreational sun exposure, her frequent consumption of piping hot food may have contributed to accumulated mechanical damage to her lower lip, leading to this condition. T-cells and macrophages have been identified to influence cellular differentiation leading to this nonspecific inflammatory response.[5]

The exact mechanism of action that tacrolimus may have on plasma cells remains unexplained. However, its known influence on immunomodulation of inflammatory cytokines such as mast cells, Langerhans cells, T-cells, and eosinophils have been identified. It is suggested that the eroded nature of the lesions contributes on tacrolimus' enhanced efficacy.[2] Although PCC is a benign condition, treatment may be challenging and clinical outcomes may vary. Topical therapeutic options include the use of topical corticosteroids and immunomodulators among others.[1] A 2017 case series done with 13 patients from 2011 to 2016 in Seoul Korea, 8 out of 13 patients used topical tacrolimus 0.1% and intralesional steroid injections. Five (5) patients were given additional methylprednisolone (0.5-1.0mg/kg/day) due to poor response and continued progression of symptoms. Other patients in the case series received topical steroids, pimecrolimus, and cryotherapy. Of the 13 patients, 23.1% of patients experienced complete resolution and 38.5% experienced partial remission. However, 38.5% of patients relapsed after complete or partial remission with a median follow-up duration of 11.2 months.[6] In a case series in Chosun University Hospital involving 20 patients from 2012 to 2019, 90% of the patients received intralesional triamcinolone injections and 60% additionally received topical tacrolimus while the rest were given methylprednisolone (0.1mg/kg/day), dapsone (100mg/day), and cryotherapy. Of the 20 patients, 45% experienced complete resolution with no recurrence, 40% of patients experienced relapse (average time for relapse was 10.75 months), while 15% did not respond to treatment.[9]

Our patient experienced partial improvement with the use of betamethasone cream for two (2) weeks. However, the patient was shifted to topical tacrolimus 0.1% ointment twice a day for maintenance therapy. The patient did not report any adverse events, burning, or irritation during the entire duration of the one-month treatment period. There was no reported recurrence of symptoms after 12 months of partial remission as well.

Performing dermoscopy and skin punch biopsy play an important role in finalizing the diagnosis for patients who present with recalcitrant cheilitis. Although rare, PCC must be considered for patients who present histopathological findings with a dense band-like infiltration of mature plasma cells. An accurate diagnosis is important for proper patient counselling and improved patient outcomes. Currently, there have been no studies that reported on the malignant potential of this condition. However, given its relapsing nature, it is therefore important to counsel patients regarding the probability of recurrence. Thus, regular follow-up amidst initial control of symptoms is important.

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