Pyodermatitis-pyostomatitis Vegetans

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Sir,

We are citing this classic case of Pyodermatitis pyostomatitis vegetans (PD-PSV) to emphasize the fact that, to make a concrete diagnosis, we need to feel the importance of detail clinical examination, relevant investigations and we must keep in mind closest differentials and rule them out before giving any final opinion. which are so vital to reach a clinching diagnosis. Hence, a lack of clinical suspicion may land us in making a wrong diagnosis of a not so common similar disease. Adding to it, it teaches us how important is history taking, clinical examination, and laboratory methods in reaching a proper diagnosis.

This case, a 33-year-old married woman, wife of a migrant laborer from Chennai, presented to our outpatient department with painful oral ulcers for 15 days followed by nodules with erosions of surface for 8 days over bilateral axillae, groin, and nipple. The patient was otherwise healthy and active barring mild degree of fever.

As per the patient’s history, lesions started in the mouth cavity 15 days back. She noticed painful small ulcers over hand palate, which subsequently appeared in other areas of oral mucosa [Figure 1]. Skin lesions started after 3–4 days of oral lesions, over both inguinal areas [Figure 2] first, with subsequent appearance over both axillae and nipple [Figure 3]. They started as edematous papules which subsequently increased in shape and size by coalescence. There were no constitutional features. On proper history taking, she denied similar attack in the past and no family history of similar illness. She was nondiabetic and nonhypertensive with all vital parameters within normal range. Sexual history denied any extramarital contact, but her husband had multiple contacts in the past 3–4 years. Even he had a history of genital ulcer 6 months back, which was treated locally with resolution.

On examination, lesions over axillae were big, mild and tender erythematous nodulo-ulcerative lesions with crusting and postulations at margin over trunk, arm, and nipple resembling pyodermas with the underlying edema. In the genital areas, lesions were in the form of moist, edematous, shiny plaques with satellite lesions. Oral ulcers were well-defined, polycyclic-margined erythematous erosions over the hard palate and buccal mucosa. There was an
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ulcerative lesion over the upper lip with crusting. Systemic examinations were normal.

Looking at the lesions and personal and sexual history, we reached a provisional diagnosis of secondary syphilis and *P. vegetans* was kept in differential diagnosis. The patient was admitted and all routine investigations were done including Tzanck smear, liver function test (LFT), renal function test (RFT), HIV screening, and venereal disease research laboratory (VDRL) (in dilution 1:32). Two samples from two different skin lesions were taken by punch biopsy for histopathology (HP)/immunofluorescence (IF) study. The patient was put under plain paracetamol and reports were awaited.

Blood counts, LFT, and RFT were within normal limits except moderate degree of peripheral eosinophilia. Both HIV and VDRL tests were negative. Tzanck smear was also negative for acantholytic cells. We did one dark-ground microscopy which was negative which excluded syphilis. We thought along the line of *P. vegetans* (Hallopeau type), which presents as morphologically similar lesion with benign course. We put the patient under oral steroid (40 mg prednisolone daily), and interestingly within 4–5 days, most of the lesions got dry and developed verrucous pigmented plaques on healing. In the meantime, we got a biopsy report which showed features consistent with *P. vegetans* (epidermal hyperplasia, moderate spongiosis, and suprabasal cleft with multiple intraepithelial abscesses with numerous eosinophils and neutrophils) [Figure 4]. Hence, a diagnosis of *P. vegetans* was made, and the patient was discharged after putting her on gradual tapering dose of oral steroids and was asked for a follow-up after 15 days.

After 3–4 days, we got the report of IF study which was sent to a higher laboratory, but interestingly it was negative. Hence, we were again put on dock and needed to think of another diagnosis. So, we reviewed the case, and looking at the course of disease, clinical presentation, early response to oral steroid, and HP finding, a suspected diagnosis of pyodermatitis-pyostomatitis vegetans (PD-PSV) was made. As we know, PD-PSV is a rare eosinophilic dermatosis of unknown cause, which clinically and histopathologically resembles, *P. vegetans*, the only distinctive feature is

![Figure 1: Ulcer in the oral cavity and upper lip](image1)

![Figure 2: Lesions in groin area](image2)

![Figure 3: Lesions over the left axilla and nipple](image3)

![Figure 4: Epithelial hyperplasia, eosinophilic spongiosis, with focal subepidermal cleft. A mixed inflammatory infiltrate with multiple intraepithelial abscesses, mainly consisting of numerous eosinophils and neutrophils (H and E, ×100)](image4)
negative IF study (direct/indirect). Another important fact is that PD-PSV has a strong association with inflammatory bowel disease (IBD), with ulcerative colitis being the most common association. It is mostly seen in females (3:1) in their 3rd–4th decades of life. Though considered a major association of IBD, it is not always absolute. IBD usually precedes the development of skin lesions by months and years. Hence, we had to wait for the patient to come up for follow-up so that we could take another history regarding her bowel problems.

The patient turned up for follow-up and was examined, and we found that most of the lesions had regressed completely leaving behind brownish pigmentation which is very characteristic of PD-PSV [Figures 5 and 6]. Oral lesion had almost disappeared [Figure 7]. Some leading questions were asked regarding her bowel habits. To our surprise, she revealed a history of recurrent bouts of bloody diarrhea in the past 3–4 years. We referred the patient to a gastroenterologist and a diagnosis of suspected ulcerative colitis was made. Now, we had no problem in reaching the diagnosis and the patient was counseled about the prognosis of disease, which is usually excellent and tends to follow the progression of basic disease.

This teaches us one lesson, we should be careful while taking history; referral of textbooks should be made keeping in mind similar diseases and relevant laboratory. Investigations, how difficult and trivial they may look, should always be done to reach one clinching diagnosis in a situation like this.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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