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Original Research Paper

Dermatology

AN INSIGHT TO DERMATITIS CRURIS PUSTULOSA ET ATROPHICANS – A CASE SERIES

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ABSTRACT Dermatitis cruris pustulosa et atrophicans is a distinct type of folliculitis that presents as pruritic, diffuse tiny follicular pustules with perifollicular edema and alopecia commonly occurring over the legs most commonly caused by Staphylococcus aureus. This was first described by Clarke in his book titled Skin diseases in the African as Nigerian shin disease in the year 1952. Dermatitis cruris pustulosa atrophicans is therapy resistant and recurrences are quite common in spite of proper treatment. Self-resolution occurs as scarring sets in. Despite being a well-recognized entity, it has been less commonly reported or studied in dermatological literature. In this article, we highlight four classical cases of dermatitis cruris pustulosa et atrophicans and its review of literature. Further studies on a larger scale need to be done to elucidate the exact etiopathogenesis and its proper management.

KEYWORDS : Dermatitis cruris pustulosa atrophicans, chronic superficial folliculitis, resistant folliculitis, staphylococcus aureus, Lupoid sycosis

INTRODUCTION:

Dermatitis cruris pustulosa et atrophicans (DCPA) is a form of chronic superficial folliculitis which presents as extremely pruritic, multiple tiny follicular pustules that involves almost all follicles with surrounding perifollicular edema and shiny skin. Due to the perifollicular edema, the angulation of hair is lost and it gives a wiry feeling on touch. It is commonly seen in males and almost all cases involve the legs (Latin – Crus means legs) starting from the lower 1/3rd of the anterior shins.[1] In its due course it can extend upwards involving the whole of the legs & thighs. Rarely can affects the face & upper limbs. The classical clinical features of DCPA are highlighted in **table 1** and the difference between folliculitis and DCPA are highlighted in **table 2** . Pruritus is an important feature of this condition it can be a sign of relapse.[2]

This was first reported in Nigeria, hence also known as Nigerian shin disease. Other synonyms include lupoid sycosis of legs,[3] Folliculitis cruris pustulosa et atrophicans[1] & therapy resistant pyogenic folliculitis of legs.[4]

The most commonly isolated organism in this distinctive folliculitis is staphylococcus aureus. However, there can be isolates of bacterial species of beta hemolytic streptococcus, Klebsiella & pseudomonas species. Fungi like trichophyton rubrum have also been associated with DCPA. [2]

Table 1: Shows clinical features to clinch the diagnosis of DCPA

- Chronic and recurrent
- Extremely pruritic
- Symmetrical involvement of legs
- Numerous tiny follicular pustules
- Perifollicular edema and scaling
- Loss of skin marking
- Wiry feeling on palpation
- Heals with scarring alopecia
- No Post inflammatory hypo/hyperpigmentation
- Absence of systemic symptoms like fever / lymphadenopathy despite extensive infection

Table 2 : Showing the differences between folliculitis and DCPA

Folliculitis	DCPA
Can occur over any site including the trunk and pubic region	Most of the cases are located over legs and spares the trunk (can occur over face, neck and forearms[5])
Few follicles are involved	Almost all follicles are involved in the affected site
Pruritus is less common. Pain is the predominant presenting symptom	It is extremely pruritic rather than painful
Recurrences less common once the precipitating factors are eliminated	Recurrences are common. Most of the patients don't have any associated precipitating factors
Not associated with perifollicular edema, shiny skin or scarring alopecia	Associated with edema & shiny skin followed by scarring alopecia.
Can heal with Post inflammatory hyperpigmentation	Doesn't heal with post inflammatory hyperpigmentation
Responds well to routine antibiotics	Resistant to routine antibiotics

Table 3: Shows grading of Dermatitis cruris pustulosa et atrophicans by Suganthan et al[5]

GRADE	CLINICAL CHARACTERISTICS
GRADE 1	Follicular pustules with perifollicular erythema
GRADE 2	Principal lesions are follicular pustules. There is also presence of papules with broken hairs, wiry roughness on palpation, excoriation marks, crusting, scaling and visible (not marked) alopecia.
GRADE 3	Infiltrated papules with scaling & some scattered pustules. Surrounding skin is shiny & shows marked alopecia & atrophy.
GRADE 4	Shows near total alopecia with few scattered papules in periphery. No pustules are seen

Case Series:

Case 1:

A 58 year old male who is a gardener came with the complaints of recurrent extremely pruritic follicular pustules with surrounding perifollicular edema and scaling which started first over both legs then progressed to involve the extensors of forearms(figure 1) & legs on and off since 6 months. He was earlier managed with amoxicillin, cephalexin, cotrimoxazole, doxycycline and dapsone without much relief. Microbiological examination revealed growth of trichophyton rubrum. Histopathological examination revealed intrafollicular abscess with papillary edema. Patient was managed with 200 mg of itraconazole capsules and anti-histamines for 2 weeks. Skin lesions showed near complete resolution.

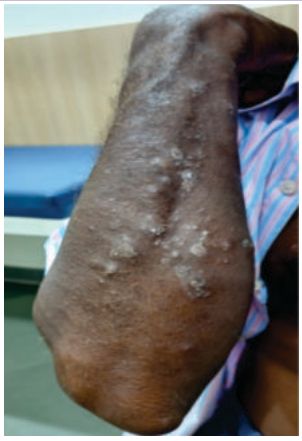

Figure 1: Showing follicular pustules with perifollicular edema & scaling over the extensor of left forearm

Case 2:
A 24-year-old male who is an IT professional male came with the complaints of recurrent, itchy pustular eruptions over the anterior aspect of skin of both legs for past 6 months. On examination, there was a skin-colored edematous plaque with skin color studded with few follicular pustules which showed a wiry feeling on palpation with loss of hair in the center (figure 2). Microbiological examination revealed heavy growth of staphylococcus aureus. Patient was managed with cotrimoxazole and anti-histamines. Patient lost follow up.

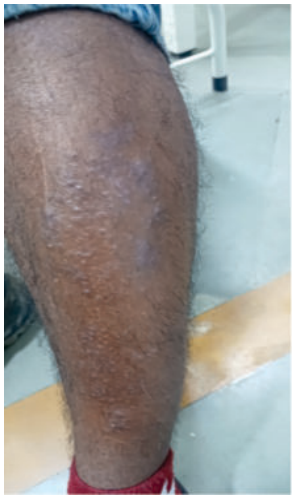

Figure 2: Shows skin colored edematous plaque with central scarring alopecia with few pustules.

Case 3:
A 32-year-old IT professional came with the complaints of recurrent itchy pustular eruptions over the legs extending from knees to foot involving both extensor and flexor aspect (figure 3). Microbiological examination revealed heavy growth of staphylococcus aureus. Patient was managed with a course of oral cotrimoxazole, topical ozenoxacin cream and anti-histamines. The skin lesions resolved in a week and a half again to recur back after a month.

Case 4:
A 44-year-old fisherman came with the complaints of recurrent folliculitis over the legs extending to the thighs since 1 month(figure 4). Lesions were extremely pruritic that patient was unable to sleep at night. He took OTC medications from the pharmacy and took native treatment with no improvement. His microbiological evaluation showed Staphylococcus aureus and histopathology showed intrafollicular infiltration with neutrophils and lymphocytes with dermal papillary edema

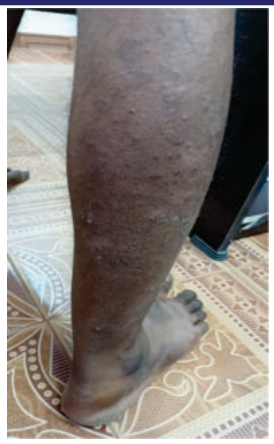

Figure 3: Shows follicular pustules with broken over hairs, perifollicular edema and loss of hair over the right leg involving the posterior aspect of right leg with sparing of the dorsum of foot.

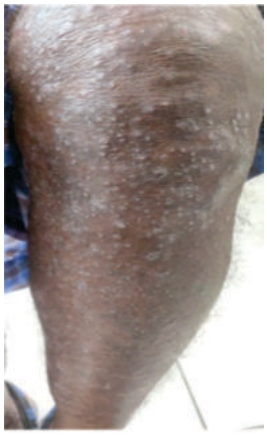

Figure 4: Shows superficial folliculitis with perifollicular scaling with few lesions showing perifollicular edema over the knees.

DISCUSSION:
Most of the cases of DCPA are idiopathic. Various risk factors have been associated with DCPA similar to that of any pyoderma. However, it's the chronicity and the recurrences that makes it a difficult to treat in spite of elimination of the precipitating factors. S.aureus forms one of the common isolates of DCPA and it also forms a common commensal of the skin.

Few factors can pave way for breach in the skin barrier responsible for the infection like excoriations secondary to pruritus.

Increased hydration reduces cases maceration which increases the friction coefficient and predisposes the individual for minor trauma paving the entry of organism as supported by the fact that its common in tropics, summer season, people with occlusive clothing, and people who have prolonged water exposure like farmers and fishermen (irritant effect can occur with sea water).

Oil application adds on to hydration and follicular occlusion. Scrubbing, contact with allergens, atopy, ichthyosis also act as precipitating factors.[2] The risk factors the pathogenesis of DCPA is given in figure 5

Surprisingly DCPA has not been associated with immunosuppressive conditions like diabetes, malnutrition or malignancy however patients with immune dysfunction in the form of hypergammaglobulinemia has shown a tendency towards DCPA.[4][6]

Figure 5: Shows risk factors the pathogenesis of Dermatitis cruris pustulosa et atrophicans

In our case series, one of the patient was a fisherman who had a risk factor for excessive skin hydration and irritation from the sea water. Another patient was a gardener, in whom allergic reaction from the plants could be suspected. The other two were IT professionals. All the patients were healthy and none of the patients had co-morbidities in the form of diabetes or malnutrition. None of them had a history of hyperhidrosis, trauma, oil application or occlusive clothing. There are case reports of DCPA occurring over face and upper limbs.

Histopathological examination of the pustular eruption shows intra, sub-corneal and perifollicular abscess comprising of PMNL, neutrophils or lymphocytes with or without papillary edema. Has a "wine glass appearance" due to abscess formation in the upper part of the follicle and sparing of the lower end involving the bulb. Epidermis can show hyperkeratosis. In later stages when cicatrization sets in, HPE shows atrophy of the rete ridges loss of appendages and upper dermal fibrosis.[1]

Management of DCPA is difficult as most of the cases are resistant to therapy and recurrences are common in spite of elimination of the risk factors. Variable rates of success have been achieved with cotrimoxazole , ciprofloxaxin, minocycline, pentoxifylline, rifampicin, dapsone and mupirocin. In later stages as cicatrization sets in, the hair follicles are lost and it results in self-healing of the condition.[1][2]

CONCLUSION:

Dermatitis cruris pustulosa atrophicans is a rare cause of recurrent resistant folliculitis. Further studies on a larger scale are required to elucidate the exact cause to proper management of this condition.

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