

Post Kala Azar Dermal Leishmaniasis (PKDL) Presenting with Ulcerated Chronic Paronychia Like Lesion

Jha AK, Anand V, Mallik SK, Kumar P

Department of Dermatology

Katihar Medical College, Katihar

Corresponding Author

Abhijeet Kumar Jha

Department of Dermatology

Katihar Medical College, Katihar

E-mail: drabhijeetjha@gmail.com

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ABSTRACT

A 50-year-old lady, resident of urban Bihar, presented with inflamed proximal nail fold of a single digit for one and half months. The lesion on distal finger was remarkable for ulceration. Mucocutaneous examination revealed multiple hypopigmented macules and patches, notable for absence of scaling. The presence of ulceration was not consistent with chronic paronychia, hence, she was asked for biopsy. She declined and opted for trial of antibiotic and anti-fungal treatment. At two weeks of follow up, no improvement was noted at all. At the same time, she developed multiple juicy papules in perioral area and on neck. She tested positive by rK 39 tests. Histopathology from periungual area showed LD body. She was diagnosed as Post Kala-azar dermal leishmaniasis and was treated with miltefosine 50 mg twice daily for three months, resulting in complete resolution of all lesions.

KEYWORDS

Post Kala azar dermal leishmaniasis, kala-azar, paronychia, ulceration, miltefosine

INTRODUCTION

Post Kala-azar dermal leishmaniasis (PKDL) is a late cutaneous complication of untreated or partially treated visceral leishmaniasis.¹ It is restricted to certain endemic areas like India, Bangladesh, Nepal and Sudan mainly.^{2,3} It is believed that Indian PKDL usually does not ulcerate. However, ulceration in Indian PKDL is being increasingly reported. We report a case of a rare case of PKDL presenting as ulcerated chronic paronychia like lesion on a single digit.

CASE REPORT

A 50-year-old lady, resident of urban Bihar and a maid by profession, presented with inflamed proximal nail fold of left ring finger for one and half months. She had similar lesion for a long time, but it was getting particularly worse since one and half months. The lesion on distal finger was remarkable for ulceration. (Fig 1) On examination, multiple hypopigmented macules and patches were present on trunk. These lesions were notable for absence of scaling. Rest of the muco-cutaneous examination was

non-contributory. The ulceration on distal finger lesion was not consistent with the diagnosis of chronic paronychia. She was advised biopsy and histopathological examination for paronychial lesion; however, she declined. A trial of antibiotics and anti-fungals was given for a period of two weeks. There was no improvement at all at the end of two weeks. However, there was a new development. Multiple erythematous juicy papules and plaques had appeared, mostly in the perioral area and neck. (Fig 2) The appearance of juicy lesions around mouth in a patient with pre-existing hypopigmented lesions was suggestive of Post Kala-azar dermal leishmaniasis. On enquiry, patient revealed history of Kala-azar some four to five years back.

The patient was tested by RK-39 test kit, which came out to be positive. The histopathological findings from both perioral popular lesion and paronychial lesion were similar and revealed many Leishman Donovan bodies. (Fig 3) KOH mount of scrapping from hypopigmented macules and patches was negative. So the diagnosis was revised as post kala-azar dermal leishmaniasis. Treatment with



Figure 1. Inflamed proximal nail fold of left ring finger. Note ulceration on distal part of finger.

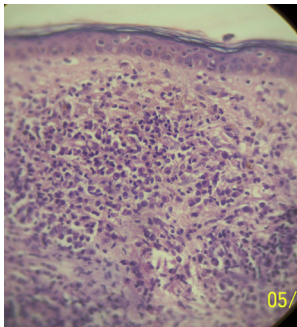


Figure 3. Atrophic epidermis, Grenz zone and dense lymphohistiocytic infiltration in dermis. LD body in upper dermis (arrow).



Figure 5. Lesions healed after three months of miltefosine therapy.

Amphotericin B was considered, but high cost and required hospitalization did not allow it. She was treated with miltefosine 50 mg twice daily, as miltefosine was available free from the National vector borne diseases control program in India. At the one month follow up, chronic paronychia lesion had healed and perioral lesion started subsiding. Treatment was continued for three months, resulting in complete resolution of lesions. (Fig 4 and 5) The patient was under follow up for another six months; no recurrence was noted in this period.

DISCUSSION

Post Kala azar dermal leishmaniasis (PKDL) is a late cutaneous manifestation of visceral leishmaniasis (VL) in untreated or inadequately treated patients, first described by Brahmachari in 1922.¹ In Sudan and India, it has been shown to follow VL in 50%-60% and 5-10% of cases, respectively. The interval at which PKDL follows VL is 0-6 months in Sudan and 2-3 years in India. In Nepal, the



Figure 2. Erythematous "juicy" papules in perioral area and on neck.



Figure 4. Lesions healed with atrophic scarring.

incidence rates of PKDL has been reported as 2.0-4.0 per 10,000 person-years in and it is particularly prevalent in districts of Jhapa, Morang, Sunsari, Saptari, and Siraha. The risk of developing PKDL after VL has been estimated as 1.4% within two years, and 3.6% within 8 years, which is lower than that reported in other VL-endemic areas in the Indian subcontinent.²⁻⁵

PKDL is characterized by wide variations in clinical lesions and presentations. In a recent cross-sectional descriptive study of 105 patients with PKDL in eastern Sudan, a papular or nodular rash was most frequently seen (51%); other types of rash were maculopapular (23%), micropapular (measles-like) (17%) and macular (9%).⁶ From India, three main presentations have been described, of which one or two may predominate: erythema and induration on the butterfly area of the face that shows photosensitivity; multiple symmetrical hypopigmented macules that may coalesce; and combinations of papules, nodules, and plaques.^{7,8} Although there are differences in description of clinical findings from Sudan and other areas; however, in most reports, macular, papular, and nodular lesions are considered the hallmarks of PKDL. Ulceration although is not considered to be a feature of Indian or Sudanese PKDL. Usually, PKDL lesions typically appear around the mouth and spread to other parts of the face; subsequent spread to upper arms and chest may follow. This pattern is most consistent in papular and nodular PKDL, but to a lesser extent in macular lesions. In most severe cases the whole body may be affected sometimes with mucosal lesions on the lips or palate.⁹ The typical pattern of distribution has resulted in the description of three clinical grades of severity. In grade one, a scattered maculopapular or nodular rash occurs mainly in the face with or without

some lesions on the upper chest, and arms. Grade two is defined as a dense maculopapular or nodular rash covering most of the face and extending to the chest, back, upper arms, and legs, gradually becoming less distally, with only scattered lesions on the forearms and legs. Grade three is defined as a maculopapular or nodular rash covering most parts of the body, including hands and feet. In grade three crusting, ulceration, sloughing, scaling, and spreading to the mucosa of the lip (cheilitis) and the palate may occur.⁶

However, atypical and unusual presentations are not uncommon. Bari et al have noted unusual presentations in 5.7% cases among 718 patients with cutaneous leishmaniasis. The commonest among unusual morphologies was lupoid leishmaniasis 14 (34.1%), followed by sporotrichoid 5 (12.1%), paronychia 3 (7.3%), lid leishmaniasis 2 (4.9%), psoriasiform 2 (4.9%), mycetoma-like 2 (4.9%), erysipeloid 2 (4.9%), chancriform 2 (4.9%), whitlow 1 (2.4%), scar leishmaniasis 1 (2.4%), DLE-like 1 (2.4%), 'squamous cell carcinoma'-like 1 (2.4%), zosteriform 1 (2.4%), eczematous 1 (2.4%), verrucous 1 (2.4%), palmar/plantar 1 (2.4%) and mucocutaneous 1 (2.4%).¹⁰

Demonstration of LD bodies in the Slit skin smear or by culture of the skin tissue was considered to be the gold standard for the diagnosis of PKDL.¹¹ Histopathology from the lesion is diagnostic and shows a poorly differentiated infiltrate of chronic inflammatory cells, with a variable number of amastigotes in dermal macrophages.² Recombinant DNA technology has produced serodiagnostic antigens, which include rK39, A2, ORF F, rH2A, rH2B, rGBP, rLACK, and purified lipophosphoglycane (LPG).¹²

There are few controlled studies on the management of PKDL and most data comes from small case series. In addition, there are differences in approach according to geographical area. Treatment with sodium antimony gluconate (SAG) and Amphotericin B have met with considerable success. In India, cure rates as 64–92% have been documented with sodium antimony gluconate (SAG) 20 mg/kg per day for 120 days given parenteral, intramuscularly or intravenously.^{13,14} Amphotericin B given parenteral at a dose of 1 mg/kg on alternate days or daily was shown to be effective and is being considered a first line therapy, especially in endemic areas with considerable SAG resistance.¹⁵ Pentamidine too has been used at a dose of 2 mg/kg on alternate days for seven doses with a cure rate 93%; however, Amphotericin B has been found to be superior to pentamidine.^{15,16} All these treatment modalities are parenteral and require hospital admission and monitoring. Experience with amphotericin B in PKDL is limited and also treatment with this is more expensive and showed some nephrotoxicity.¹⁷ Resistance to treatment is also an upcoming problem which advocates cautious use of these drugs. Drug-resistant parasite strains or immunosuppression (eg, caused by HIV) are usual underlying problem responsible for unresponsiveness to these agents.^{18,19} Moreover, the lengthy course of intramuscular or intravenous injections means that many patients fail to complete their full course of treatment.²⁰

There is one case of successful surgical removal of a localised lesion by shave excision.²¹ However, usefulness of such procedures is limited as they are not suitable for widespread lesions.

The need for oral medicine with less serious adverse effects has been felt for long. Oral Miltefosine has been found to be effective in treatment of both VL and PKDL and has shown some promise. In fact, it is being used as a pilot programme in certain districts in India for treatment of Kala azar and PKDL under National vector borne diseases control program. Most commonly, it has been used in a dose of 2.5 mg/kg for 28 days, although longer periods of treatment may be required. However, miltefosine too is not free from limitations. The drug is contraindicated for pregnant women or women of child bearing age.^{22,23} PKDL developing in miltefosine treated VL cases has been described.²⁴ Also, long term follow up studies of miltefosine treated patients are lacking. Hence, in our opinion, miltefosine should be used with a caution and patient should be asked to come for regular follow up.

In an endemic area, the unusual manifestations of PKDL are not uncommon. High index of suspicion and appropriate investigations are often necessary in proper management of patient. Our patient presented initially with chronic paronychia like lesions, a rare presentation of PKDL. At this stage, diagnosis was missed as we did not find anything suspicious in a chronic paronychia like lesion in a maid. However, the ulceration was proximal nail fold was not fitting with chronic paronychia.

CONCLUSION

With this case report, we want to put emphasis on unusual presentation of PKDL. Also, we recommend maintaining a high index of suspicion in an endemic area, and PKDL should always be kept in the list of differential diagnoses.²⁵

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