

Case Report

A Rare Case of Cutaneous Leishmaniasis Presenting as Rhinophyma

Arghya Bandyopadhyay, Kousik Bose

Dept. of Pathology, Burdwan Medical College, Burdwan, India

ABSTRACT

Cutaneous leishmaniasis (CL) is referred to a group of diseases because of the varied clinical presentation, ranging from small cutaneous nodule to wide spread mucosal destruction. The nose is rarely involved by CL in even in endemic region. In this report we describe a rare rhinophymatous presentation of CL from a non-endemic region which was diagnosed by fine needle sampling and treated with Miltefosine.

KeyWords : Cutaneous Leishmaniasis, Rhinophyma

Introduction

Cutaneous leishmaniasis (CL) is a parasitic disease caused by parasitic protozoa of the genus *Leishmania* and spread by female sandfly. *Leishmania* species primarily affect cells of monocyte-macrophage lineage and exist in two forms during their life cycle: a flagellar (promastigote) and an aflagellar (amastigote) stage. During the bite of infected sand fly promastigote enters the skin and transform into amastigote within histiocytes. Cutaneous lesion develop if the histiocytic response confined to the skin, if dissemination of the protozoa occurs, internal organs becomes involved (1).

CL is very rare in India and few cases have been

reported from Rajasthan and Himachal Pradesh (2). It usually presents as an asymptomatic solitary erythematous papule, tends to become nodule centrally covered by yellow scabs, and produces ulcers on the exposed parts of the body, such as the face, arms and legs. Nasal involvement in cutaneous leishmaniasis is seen only in 9.82% cases (3,4). Nasal CL may present as psoriasiform plaques, furunculoid nodules and lupoid plaques but rarely present as rhinophyma (5). Rhinophyma like presentation of CL has been described only once in English literature (6).

We are presenting the second case of CL with rhinophyma like presentation in young female from West Bengal, India, diagnosed by fine needle sampling (FNS) and confirmed by histopathology.

Received: 30 Oct 2013

Accepted: 27 Jun 2014

Address communications to: Dr. Arghya Bandyopadhyay, Department of Pathology, Burdwan Medical College, Burdwan, India.

Email: drarghyabanerjee@yahoo.com

Case Report

A 20 year old female from Birbhum district (West Bengal), India, presented with soft painless swelling of her nose clinically resembling rhinophyma for last 2- 3 months (Fig. 1). She had severe pallor but other systemic examinations were unremarkable. No organomegaly or lymphadenopathy was detected. No history of visceral leishmaniasis (VL) was present. FNS and skin biopsy were done in our department. FNS smears showed *Leishmania* amastigote in a chronic inflammatory background, subsequent histopathological examination showed *Leishmania* amastigote, chronic lympho-plasmacytic infiltration and ill-defined non caseating granuloma (Fig. 2, 3a and 3b). She was positive for rk-39 and negative for HIV. She had microcytic hypochromic anaemia with Hb of 7.7 gm/dl, other biochemical investigations were within normal limits. Her bone marrow smears did not show *Leishmania* amastigotes. She was diagnosed as CL and treat-

ed with miltefosine with complete clinical cure in three months.



Fig. 1: Cutaneous leishmaniasis give rise to enlarged bulbous nose (Rhinophyma like) with satellite lesions in surrounding skin.

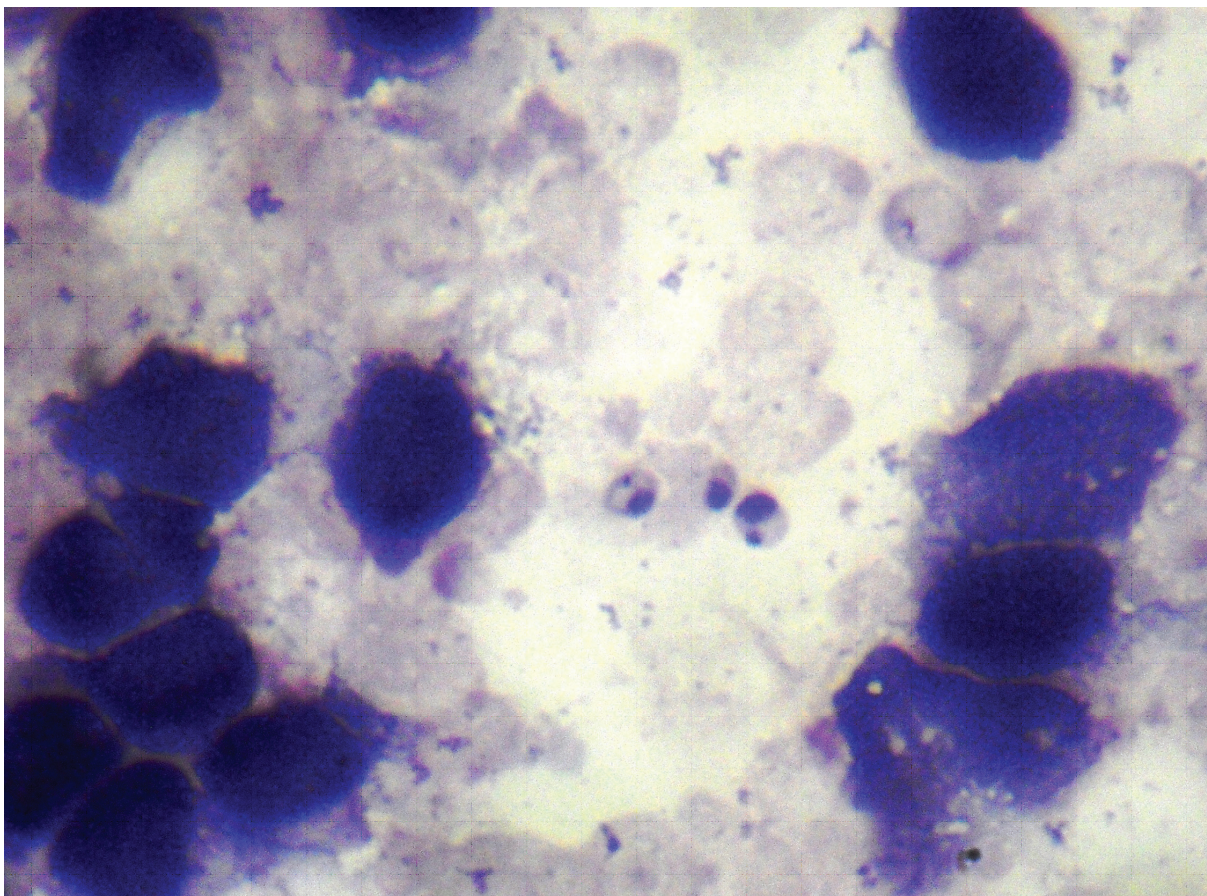


Fig. 2: Cytology smear showing *Leishmania* amastigote in a reactive lymphoid background. (H & E, $\times 1000$)

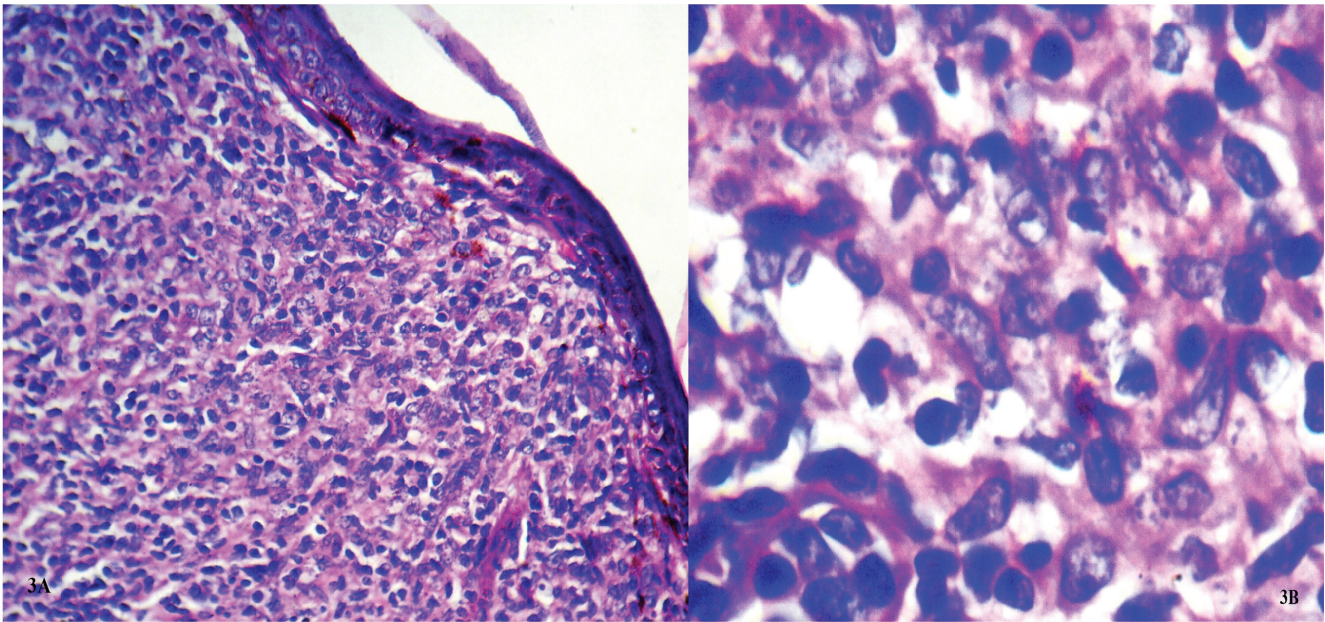


Fig. 3: a) Histopathology of the lesion showing thinned out epidermis and in the superficial dermis there is collection of epithelioid cells forming ill defined non caseating granulomas.(H & E,×400); b) *Leishmania* amastigotes in a background of epithelioid macrophages and lymphocytes. (H & E, ×1000)

Discussion

Localized cutaneous leishmaniasis(LCL) usually caused by *Leishmania tropica* and *L. major* transmitted by bite of sand fly in the Old World. It is endemic in 70 countries, 90% cases are reported from Afganistan, Pakistan, Saudi Arabia, Syria, Brazil, Peru and Algeria. But in south east Asia LCL is caused by *L.tropica* & *L.donovani* (1) and recent reports from India showing *L. donovani* is the predominant pathogen of CL (2). It is imported in nonendemic areas by immigrants, returning travelers and more common in night outdoor workers (3).

A case series from Pakistan, showing nasal involvement in cutaneous leishmaniasis is seen only in 9.82% cases (4). Another case series from India is showing that within face CL most commonly involve cheek (48.5%) and nasal bridges only in 11.4% cases (2). Nasal involvement could possibly due to the fact that nose is a immobile more projected part of face and thus inability to avoid sand fly bite (4).

Typical CL present as an asymptomatic solitary erythematous papule tends to become nodule centrally covered by yellow scabs with raised

indurated margins (5). Nasal involvement usually shows mucocutaneous lesion following cutaneous lesion, leading to ulceration and even erosion of cartilage (4). Though there are many clinical varieties of cutaneous leishmaniasis are known, including impetiginoid, erysipeloid, vegetant, tuberous, verrucosus, nodular, necrotic, lymphangitic or sporotrichoid forms (5), rarely present as rhinophyma. Rhinophyma like presentation of CL has been described only once in English literature (6) as in this case.

The precise pathogenesis of rhinophymatous appearance is not well understood. However, the main factors could possibly be an altered cell-mediated immune host response to the *Leishmania* parasite and the peculiar structural anatomy of nasal tissue. Moreover the laxity of the facial tissue and the great vascularization may enhance cutaneous expression of the *Leishmania* infestation (6).

Rhinophymatous lesion means enlargement of nose, which can give us many differential diagnosis like Inflammatory dermatoses (Granuloma faciale, localising chronic fibrosing vasculitis), benign cutaneous lesion (angioma,

sebaceous adenoma), malignant lesions (sebaceous carcinoma, adenoid squamous cell carcinoma, basal cell carcinoma, angiosarcoma, cutaneous B-cell lymphoma), infective causes (*Mycobacterium*, *Cryptococcus*, Rhinoscleroma, PKDL), following phenytoin intake etc. (7). To differentiate these proper history taking, cytology, histology play a major role. Most importantly the clinicians should have a high level of suspicion and familiarity with different presentations of CL, that direct them to carry out further investigations.

For diagnosis there are several techniques like skin smear examination, fine needle sampling, histopathology, polymerase chain reaction, and monoclonal antibody test (8). According to Sharma *et al.* histology demonstrates a chronic granulomatous inflammation with non caseating epithelioid granuloma, langhans type giant cell in 77% cases as in this case; and in 14.5% cases there are chronic non specific inflammation with dense lymphoplasmacytic infiltration (2). We have seen FNS followed by Giemsa stain is an excellent method to demonstrate *Leishmania* amastigote, known as Leishman –Donovan bodies (LDB) in cutaneous smears (9). In cytology smears or in histopathology the differentials which may be considered with LDB are histoplasma, *Cryptococcus neoformans*, *Candida glabrata* and *Klebsiella rhinoscleromatis* (10). In case of leishmaniasis, recognition of a nucleus and kinetoplast in the organism and the absence of budding will exclude histoplasmosis. Cryptococci are round capsulated organisms with thick capsule, and positive capsular staining on Mayer's mucicarmine while negative staining with India ink preparation. In contrast to LDB these fungal organisms stain with periodic acid Schiff (PAS) and Grocott's methenamine silver (GMS). *Klebsiella rhinoscleromatis*, the inciting agent of rhinoscleroma, also resides in macrophages. In contrast to leishmaniasis, rhinoscleroma reveals a large number of plasma cells with often formation of Russell bodies (11). In our case,

after demonstrating *Leishmania* amastigotes in FNS smear, the main differential diagnosis was post kala-azar dermal leishmaniasis (PKDL) which is the cutaneous manifestation of visceral leishmaniasis (VL) after apparent clinical cure (9). We favoured the diagnosis of rhinophymous CL as there was no history of previous VL and histopathology showed epithelioid granuloma, where as histology of macular PKDL show inflammatory cells concentrating around vascular plexuses, and papules show a grenz zone, a strip of collagen separating epidermis from infiltrate (12), which was absent in this case. The possible pathogen in this case was *L. donovani* as it is endemic in Bihar, adjacent to West Bengal in India.

Conclusion

The clinicians should keep this condition in mind while examining a case of rhinophyma especially for those, who are from an endemic zone of leishmaniasis or having a history of staying in endemic country, and Pathologists should meticulously search for *Leishmania* amastigote when they see the smears of such case.

Acknowledgement

The authors are thankful to Dr. S Bhattacharya, Dr. K Konar and Prof. U Banerjee for their help and support. The authors declare that there is no conflict of interests.

References

1. Reithinger R, Dujardin JC, Louzir H, Pirmez C, Alexander B, Brooker S. Cutaneous Leishmaniasis. *Lancet* 2007;7(9):581-96.
2. Sharma N, Mahajan V, Kangab A, Sood A, Katoch VM, Mauricio I, *et al.* Localized cutaneous leishmaniasis due to *Leishmania donovani* and *Leishmania tropica*: Preliminary findings of the study of 161 new cases from a new focus in Himachal Pradesh, India. *Am J Trop Med Hyg* 2005;72(6):819-24.

3. Neupane S, Sharma P, Kumar A , Paudel U , Pokhrel DB. Cutaneous Leishmaniasis : report of rare cases in Nepal. *Nepal Med Coll J* 2008 ; 10(1):64-7.
4. Khan MI, Muhammad M, Khan W, Khan N, Noor SM. Nasal involvement of Cutaneous Leishmaniasis. *JPMI* 2010; 24(3): 202-6 .
5. Guarneri C, Vaccaro M, Cannavo SP, Borgia F, Guarneri B. Erythematous-edematous-infiltrative plaque on the face: cutaneous angio-lupoid leishmaniasis. *Europ J Dermatol* 2002 ; 12(6):597-9.
6. Bari A, Ejaz A. Rinophymous leishmaniasis: A new variant. *Dermatol Online J* 2010;15(3):10.
7. Moran AM, Orman S, Helm FK. Localising Chronoic Fibrosing Vasculitis causing a rhinophymatous eruption. *Cutis* 2006; 78(5):325-8.
8. Sharquie KE, Hassen AS, Hassan SA, Al-Hamami IA. Evaluation of diagnosis of cutaneous leishmaniasis by direct smear, culture and histopathology. *Saudi Med J* 2002; 23(8):925-8.
9. Bandyopadhyay A, Majumdar K, Gangopadhyay M, Koley S. Fine-needle sampling provides appreciable diagnostic yield in lesions of post kala azar dermal leishmaniasis: Analysis of four cases from North Eastern India. *Diag Cytopathol* 2013;42(6):525-9.
10. Arghya B, Kaushik M, Mimi G, Subrata C. Cytodiagnosis of cutaneous histoplasmosis in HIV positive patient initially presenting with multiple umbilicated disseminated skin nodules. *Diagn Cytopathol* 2013;41(5):459-62.
11. Shum TK, Whitaker CW, Meyer PR. Clinical update on rhinoscleroma. *Laryngoscope* 1982;92(10 Pt 1):1149-53.
12. Sing A, Ramesh V. Histopathological features in leprosy, post kala-azar dermal leishmaniasis, cutaneous leishmaniasis. *Indian J Dermatol Venerol Leprol* 2013; 79(3):360-6.