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HANSEN'S DISEASE DIAGNOSED AFTER ANTI-CANCER CHEMOTHERAPY (case report)

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Background. *Leprosy or Hansen's disease is a chronic granulomatous disease involving predominantly skin, peripheral nerves and nasal mucosa but capable of affecting any tissue or organ. Histoid leprosy is a very rare well-defined clinicopathological variant of multibacillary lepromatous leprosy, which is very difficult to diagnose due to different specific clinical and histopathological findings that mimic a fibromatous disorder. Histoid leprosy occurs generally after treatment failure and sometimes de novo.*

Objective. *The aim of the study was to explore histoid leprosy throughout a case report.*

Methods. *A case report of histoid leprosy diagnosed after cancer chemotherapy is presented.*

Results. *A 25-year-old healthy male presented with multiple skin coloured, discrete, well defined, painless papules and nodules scattered over nape of neck, right side of the trunk and both arms along with numbness as well as tingling sensation over both the arms and trunk. It was a case of non-seminomatous germ cell tumour (NSGCT), left testis, diagnosed and treated with a high inguinal orchidectomy with adjuvant chemotherapy in 2016. Ziehl Neelsen (ZN) stain for Acid Fast Bacilli (Mycobacterium leprae) – a modified Fite stain method showed numerous acid-fast bacilli. Histopathological diagnosis of Hansen's disease (Histoid) was conducted. The patient was admitted and started on triple drug multi-bacillary multi-drug therapy (MB-MDT). A remarkable improvement was noticed in the lesion status within one month of institution of the therapy.*

Conclusions. *Histoid leprosy is a discrete infrequent form of multibacillary leprosy with distinctive clinical, bacteriological and histomorphological features. Histopathologic examination with modified Fite stain is still the mainstay of diagnosis.*

KEY WORDS: **histoid leprosy; acid fast bacilli; multi drug therapy.**

Introduction

Leprosy is a chronic granulomatous disease involving predominantly skin, peripheral nerves and nasal mucosa but capable of affecting any tissue or organ. Histoid leprosy is a very rare well-defined clinicopathological variant of multibacillary lepromatous leprosy, which is very difficult to diagnose due to different specific clinical and histopathological findings that mimic a fibromatous disorder. Morphologically, histoid lepromas are sudden eruptions of dome-shaped tumours, resembling eruptive kerato-acanthomas or cutaneous metastasis.

The global burden of lepromatous leprosy is shared by Kenya, Cuba, Indonesia and Democratic Republic of Congo. India has eliminated leprosy in 2005 to less than 1 case per 10,000 population. Despite this feat achieved 13 years ago, India is the deemed leprosy capital of the world owing to a massive population of 1.32 billion. In India, Annual New Case Detection Rate (ANCDR) for leprosy is 9.71 per 100,000 population and a Prevalence Rate (PR) of 0.66 per 10,000 population with a Child Case rate of 8.94% as recorded in 2016. Pockets of high endemicity with prevalence rate of >1% are present in many parts of India [1].

Histoid leprosy has been registered to occur at an age of ten years or younger to as old as 84 years. 58% of leprosy patients are between 20 and 39 years of age with male preponderance in Indian studies [2]. Histoid leprosy is quite common in patients on irregular and inadequate dapsone monotherapy. However, the *de-novo* occurrence of the disease has only been recorded sporadically. In India, the overall incidence of histoid leprosy among leprosy patients has been estimated to be between 2.79 and 3.60% [3]. In a study from the state of Rajasthan in India, the incidence of biopsy proven histoid leprosy was 2.8% [4].

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Case report

A 25 year old healthy male, a resident of eastern Uttar Pradesh in India, presented with multiple skin coloured, discrete, well defined, painless papules and nodules scattered over nape of neck, right side of the trunk and both arms along with numbness as well as tingling sensation over both arms and trunk. The lesions were first noticed six months back with presence of a few lesions which progressed within one month, bringing him to seek medical attention. It was a case of non-seminomatous germ cell tumor (NSGCT), left testis, diagnosed and treated with a high inguinal orchidectomy with adjuvant chemotherapy in 2016.

On dermatological examination the dome shaped, nodular lesions were firm and non-tender (Fig. 1).

There was patchy hypo-aesthesia over dorsum of left hand (ulnar distribution), lower 1/3 of the anterior leg and dorsum of the foot. Non tender, uniform, peripheral nerve thickening was noted in right greater auricular, both the ulnar, both radial cutaneous, both common peroneal and left anterior tibial nerves. A healed trophic ulcer over the ulnar border of left hand was also noted. There was no loss of power or muscle wasting or deformity evidenced. His vital status and systemic examination were within normal limits.

His routine investigations such as complete hemogram, liver function test, renal function tests, chest X-ray (PA-view) were all within normal reference range. His split skin smear (SSS) showed a bacteriological index of 5+ with a morphological index of 90%. H&E stained sections from intra-lesional punch biopsy of nodular skin lined lesion showed a thin epidermis with a nodular proliferation of spindle shaped histiocytes with a clear grenz zone in the dermis (Fig. 2).

The nuclei of these fusiform cells are pyknotic with moderate cytoplasm. Occasional epitheloid component forming epitheloid granulomas also noted (Fig. 3).

Ziehl Neelsen (ZN) stain for Acid Fast Bacilli (*Mycobacterium leprae*) – Modified Fite stain method showed numerous acid-fast bacilli (Fig. 4). Histopathological diagnosis of Hansen's disease (Histoid) was performed.

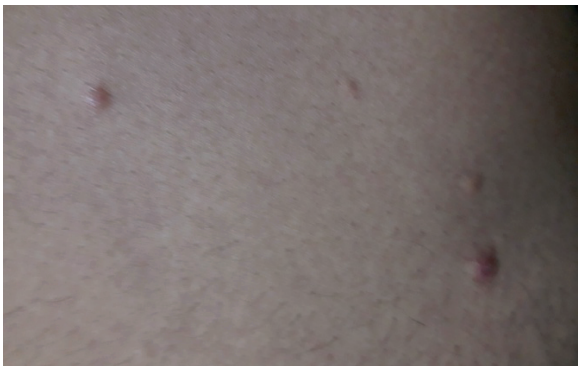

Fig. 1. Clinical presentation: Multiple soft nodules over the skin.

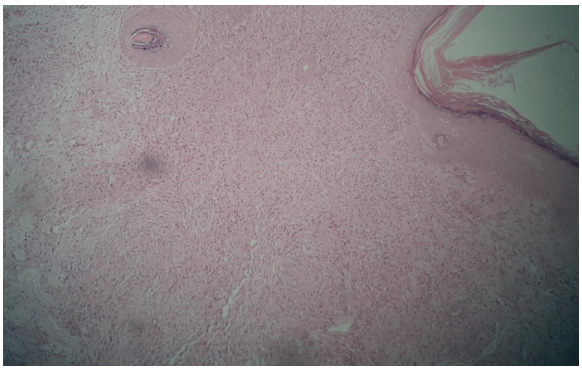

Fig. 2. Photomicrograph: H&E, X 400. Thin epidermis and nodular proliferation of spindle shaped histoid cells.

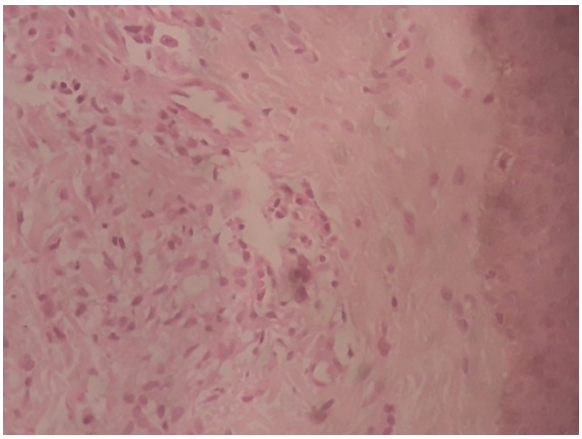

Fig. 3. Photomicrograph: H&E X 1000; demonstration of clear grenz zone.

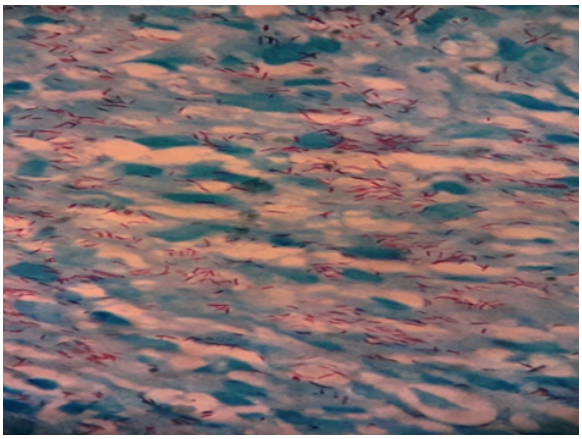

Fig 4. Photomicrograph: Fite Stain, X 1000; Fite stain showing numerous Leprae bacilli.

The patient was admitted and started on triple drug multi-bacillary multi-drug therapy (MB-MDT). A remarkable improvement was noticed in the lesion status within one month of institution of the therapy.

Discussion

Leprosy is caused by *Mycobacterium leprae* and *Mycobacterium lepromatis*, which has been discovered in 2008 causing Diffuse Lepromatous Leprosy of Lucio and Lapati in Central America and the Caribbean. Leprosy is a re-emerging disease de novo as well as in immunocompromised hosts [5]. It can present in the backdrop of immunocompromised states including tumours a seen in this patient [6].

Histoid leprosy has characteristic clinical, histopathologic and bacterial morphological features with an overall male preponderance. Macroscopically, lesions have been variously classified as subcutaneous nodules, deeply fixed cutaneous nodules, superficially placed cutaneous nodules, soft nodules, and plaques or pads over normal skin. Normally the maximum size of the lesion varies between 1.5-3 cm [7, 8]. They may have atypical presentation as giant lesions [9]. Common sites include arms, dorsum of hands, thighs, on the lower part of the back, on the buttocks and over the bony prominences, especially over the elbows and knees. Mucosal and genital lesions have been recorded in histoid leprosy [10, 11]. A single patient can have 3-50 lesions [2]. The smaller nodules are soft and the larger nodules are fibrotic. Such nodules may remain subcutaneous indefinitely or migrate towards the surface to fuse with the dermis. The patient presented almost similar kind of picture with approximately 08-10 lesions over the skin of neck, trunk and arms [12].

The classical microscopic features include epidermal atrophy as a result of dermal expansion by the underlying lepromas and an acellular band located immediately below the epidermis called sub epidermal grenz zone seen in some cases. The most striking and classical feature of typical active histoid nodules is the presence of numerous, thin, spindle-shaped histiocytes forming interlacing/intertwining bands, whorls and at times, tight curlicues giving it a tangled/storiform pattern containing acid fast bacilli. The lesion resembles a fibrohistiocytic tumour. Within the histiocytes there are numerous well-preserved acid-fast bacilli arranged in parallel bundles along the long axis of spindle histiocytes (histoid-habitus) with or without globes formation [13, 14]. The histomorphological picture in our case was in sync with histoid pattern and presence of mycobacterium confirmed on ZN for AFB (L)-modified Fite stain, which showed an abundance of the organisms.

Histoid leprosy is treated with ROM therapy followed by MB-MDT [15, 16]. High degree of suspicion is warranted for diagnosis of Histoid Leprosy, as it can be easily missed, being completely eradicated in many countries [17]. Leprosy being transmissible by direct contact is a risk in overcrowding [18]. This lesion can be misdiagnosed as a fibrohistiocytic tumour. Histological differential diagnoses include nodular sub epidermal fibrosis, dermatofibroma, and similar skin tumours on routine haematoxylin and eosin stains. Staining for acid-fast bacilli may, however, easily differentiate histoid lesions from such tumours, due to presence of Leprae bacilli in exceptionally large numbers within the fusiform cells of histoid lesions.

Conclusion

Histoid leprosy is a discrete infrequent form of multibacillary leprosy with distinctive clinical, bacteriological and histomorphological features. The appearance of histoid lesions certainly indicates a highly active lepromatous process. Histopathologic examination with modified Fite stain is still the mainstay of diagnosis along with a strong sense of suspicion.

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Conflict of Interests

The authors declare no conflict of interest.

Author Contributions

Hashmi S.A. – conceptualization, data curation, formal analysis, investigation, writing – original draft and review & editing; *Bhadauria G.S.* – formal analysis, investigation; *Rajmohan K.S.* – conceptualization; *Khan I.D.* – conceptualization, investigation, writing – original draft and review & editing; *Gupta A.* – investigation; *Mitra D.* – investigation; *Gupta R.M.* – writing – review & editing; *Rahman M.* – investigation; *Kapoor U.* – formal analysis; *Singh S.K.* – writing – review & editing.

ХВОРОБА ГАНСЕНА (ЛЕПРА) У ПАЦІЄНТА ПІСЛЯ ПРОТИПУХЛИННОЇ ХІМІОТЕРАПІЇ (клінічний випадок)

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Вступ. *Лепра або хвороба Гансена – це хронічне гранулематозне захворювання, яке уражає переважно шкіру, периферичні нерви та слизові носа, однак може локалізуватися у будь-якому органі чи тканині. Гістоїдна лепра – рідкісний, добре описаний варіант мультибацилярного лепроматозного типу лепри, який вкрай важко діагностувати через різноманітні специфічні клінічні на патоморфологічні прояви, що подібні до фіброматозних уражень. Гістоїдна лепра як правило розвивається після невдалого та неефективного лікування, іноді de novo.*

Мета. *Дослідити особливості перебігу гістоїдної лепри, як варіанту лепроматозного типу лепри, на прикладі клінічного випадку.*

Methods. *Описано та проаналізовано клінічний випадок гістоїдної лепри, діагностованої у пацієнта після походження протипухлинної хіміотерапії.*

Результати. *25-річний чоловік звернувся зі скаргами на зміни кольору шкіри, чітко окреслені безболісні папули та вузлики, які розташовувалися навколо шиї, на правій стороні тулуба та обох руках, і супроводжувалися онімінням, поколюванням і порушеннями чутливості на обох руках і тулубі. Перед цим у нього було діагностовано негерміногенну пухлину лівого яєчка. Проведено високу пахвинну орхіектомію з наступною ад'ювантною хімотерапією у 2016 році. При забарвленні взятого клінічного матеріалу за методом Ціля-Нільсена для кислото-стійких бактерій та за модифікованим методом Файт було встановлено наявність численних кислото-стійких бактерій. При проведенні гістологічного дослідження встановлено діагноз хвороби Ганзена, гістоїдної форми. Пацієнтові призначено мульти препаратну потрійну терапію. Значне покращення стану шкіри відмічено через місяць від початку лікування.*

Висновки. *Гістоїдна лепра – рідкісне форма мульбацилярної лепри з характерними клінічними, бактеріологічними та гістоморфологічними ознаками. Гістологічне дослідження та фарбування за модифікованим методом Файт – все ще основні методи діагностики захворювання.*

КЛЮЧОВІ СЛОВА: **гістоїдна лепра; кислото-стійкі бактерії; мультипрепаратна терапія.**

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References

1. Singal A, Sonthalia S. Leprosy in post-elimination era in India: Difficult journey ahead. Indian Journal of Dermatology. 2013 Nov;58(6):443-6.
doi: 10.4103/0019-5154.119952

2. Sehgal VN, Srivastava GO, Beohar PC. Histoid leprosy – a histopathological reapparel. Acta Leprologica. 1987;5(2):125-31.

3. National Leprosy Eradication Program (NLEP). Annual Report for the year 2015-16. Central Leprosy Division, Directorate General of Health Services, Ministry of Health and Family Welfare, Government of India.

4. Kalla G, Purohit S, Vyas MC. Histoid, a clinical variant of multibacillary leprosy: report from so-called nonendemic areas. International Journal of Leprosy and Other Mycobacterial Diseases. 2000 Sep 1;68(3):267-71.

5. Khan ID, Sahni AK, Bharadwaj R, et al. Emerging organisms in a tertiary healthcare set up. Med J Armed Forces India. 2014 Apr 1;70(2):120-8.
doi: 10.1016/j.mjafi.2013.09.005.

6. Khan ID, Dogra PM, Ramphal SK, et al. Polymicrobial infections in a teenaged Renal Transplant Recipient. Journal of Basic and Clinical Me_ dicine. 2015 Jul 30;4(1):37-9.

7. Murthy SV, Rao SM, Thejaswini KM. De-novo histoid leprosy. Journal of Laboratory Physicians. 2011 Jul;3(2):110-2.
doi: 10.4103/0974-2727.86844

8. Chan GJ, Tang WY, Lam WY. Histoid lepromatous leprosy presenting as keloid-like nodules: a case report. Hong Kong Journal of Dermatology and Venereology. 2006;14(2):83-6.

9. Nigam PK, Singh G. Mucosal and genital lesions in histoid leprosy. International Journal of Dermatology. 1990 Apr;29(3):207-8.
doi: 10.1111/j.1365-4362.1990.tb03802.x

10. Kamath DH, Sukumar D, Shetty NJ, Nanda-Kishore B. Giant lesions in histoid leprosy – an unusual presentation. Indian Journal of Leprosy. 2004;76(4):355-8.

11. Price EW, Fitzherbert M. Histoid (high-resistance) lepromatous leprosy. Int J Lepr. 1966;34:367-74.

12. Bhat YJ, Hassan I, Yaseen A, Wani R. De novo histoid leprosy: A case report from a post-elimination area. Indian Journal of Dermatology. 2015 Mar;60(2):214.
doi: 10.4103/0019-5154.159666

13. Sehgal VN, Srivastava G. Histoid leprosy: a prospective diagnostic study in 38 patients. Dermatology. 1988;177(4):212-7.
doi: 10.1159/000248566

14. Sehgal VN, Srivastava G. Status of histoid leprosy – a clinical, bacteriological, histopathological and immunological appraisal. The Journal of Dermatology. 1987 Feb;14(1):38-42.
doi: 10.1111/j.1346-8138.1987.tb02993.x

15.Annigeri SR, Metgud SC, Patil JR. Lepromatous leprosy of histoid type. Indian Journal of Medical Microbiology. 2007 Jan 1;25(1):70-1.
doi: 10.4103/0255-0857.31069

16. Manoharan R, Madhu R, Srinivasan MS. Histoid Hansen-A case report. J Indian Soc Teledermatol. 2008;2(2):12-6.

17. Khan ID, Gupta N, Rangan NM, et al. Evaluation of pre and post analytical variables in clinical microbiology services in multidisciplinary ICU of a medical college and tertiary care hospital. Journal of Basic and Clinical Medicine. 2016 Apr 5;5(1):2-4.

18. Khan ID, Khan SA, Asima B, et al. Morbidity and mortality amongst Indian Hajj pilgrims: A 3-year experience of Indian Hajj medical mission in mass-gathering medicine. Journal of Infection and Public Health. 2018 Mar 1;11(2):165-70.
doi: 10.1016/j.jiph.2017.06.004

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