Histoid Leprosy Mimicking Eruptive Xanthomas-2: Case Studies

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ABSTRACT
Histoid leprosy is a rare clinical variant of leprosy. Several cases have been reported from India even in post-elimination era. Histoid leprosy mimics variety of dermatological conditions like xanthoma, neurofibroma, dermatofibroma, sarcoid and post-kala-azar leishmaniasis. We report two cases of histoid leprosy mimicking eruptive xanthomas in different age groups.

Keywords: Histoid leprosy, Clinical features, Xanthomas, Skin smears

INTRODUCTION
Histoid leprosy (HL), a variant of lepromatous leprosy (LL), was first described by Wade in the year 1960 [1]. Large number of cases has been reported from India and other endemic countries, even in post elimination period [2,3]. In India its incidence among patients with leprosy is estimated to be 2.79%-3.60% [4].

CASE 1
A 65 year old non-diabetic male of Indian origin, consulted a dermatologist in New Jersey, USA for asymptomatic, multiple, skin colored, shiny, nodules over the back, trunk and extremities. He was thoroughly investigated. All the hematological investigations were normal. The biochemical tests were: fasting blood sugar: 104 mg/dl; HbA1c: 7%; lipid profile (cholesterol: 230 mg/dl; triglycerides: 300 mg/dl; HDL: 35 mg/dl; LDL: 145 mg/dl). Other biochemical tests like liver function tests, blood urea, serum creatinine were within normal limits. Skin biopsy report showed a collection of CD68 positive foam cells in upper part of dermis. CD117 highlights rare mast cells. Immunohistochemical (IHC) markers showed scattered Langerhans cells expressing S-100. Skin smear examination was not done. A diagnosis of xanthoma was made. He was advised regarding diet and regular exercise. After 6 months, he came to his native place in India and consulted three more local dermatologists as his condition worsened. They were also of the same opinion upon the USA reports and he was not advised any specific treatment. As his clinical condition was worsening with development of newer lesions, he visited our department for opinion. On our examination, we found multiple, discrete, skin colored, shiny, succulent, nodular lesions and plaques all over the back, trunk, and both upper and lower extremities on apparently normal skin (Figure 1). A few nodules were also seen on the face and both ear lobules. Edema of both hands and feet with non-healing ulcers over the fingers were also present. Both ulnar and lateral popliteal nerves were thickened and sensory impairment over the lesions and glove and stocking anesthesia were present. There was no lymphadenopathy and organomegaly. Our clinical diagnosis was histoid leprosy or lepromatous leprosy. In addition to routine hematological and biochemical investigations, skin smears for acid fast bacilli (AFB) were taken from ear lobes as well as from nodules and plaques and skin biopsy was taken from the nodules. Skin smears showed multiple solid staining AFB, no bundles and globi were observed (Figure 2). Bacterial index was 6+. Lesional skin biopsy specimen revealed the following features, epidermis was thinned out and multiple granulomas comprising histiocytes, lymphocytes, plasma cells and foam cells were present in the dermis. Occasional spindle shaped histiocytes were also present (Figure 3a). Fite stain of the specimen showed solid AFB (Figure 3b). A final diagnosis of histoid leprosy was arrived on the basis of clinical, histopathological and bacteriological features. He was advised multibacillary-multidrug therapy (MB-MDT) WHO regimen comprising dapsone 100 mg and clofazamine 50 mg daily with rifampicin 600 mg and clofazamine 300 mg once in a month under supervision for 2 years. In view of high bacterial load, ofloxacain 400 mg twice daily for 8 week
was also included in the regimen. After 8 weeks of MB-MDT, majority of the nodules regressed. Patient is currently under follow up.

Figure 1. Discrete nodular lesions on apparently normal skin over the back.

Figure 2. Skin smears showing multiple acid fast bacilli (AFB staining, 100X).

Figure 3a: Spindle shaped histiocytes, lymphocytes, plasma cells and foam cells (H&E 400X).
CASE 2
An 8 year old male child with multiple asymptomatic skin colored nodules was referred from the department of pediatrics with a diagnosis of xanthoma for our opinion. On physical examination, there were multiple, discrete, asymptomatic skin colored nodules over inner thighs, scrotum and penis. In addition, there were infiltrated plaques over buttocks (Figure 4). Detailed history revealed the child started developing these lesions initially on the thighs 6 months ago and multiple lesions over the other areas during the course of illness. Sensory impairment over the lesions and lower extremities was equivocal. Bilateral popliteal nerves were thickened and non-tender. There was no regional lymphadenopathy. Systemic examination was normal. All hematological and biochemical investigations were normal. Skin smears from the nodules were positive for AFB and bacterial index 6+. Lesional biopsy histopathological features were suggestive of histoid leprosy. A diagnosis of histoid leprosy was made. Child was given MB-MDT WHO regimen comprising of rifampicin 300 mg and clofazamine 100 mg once monthly and daily Dapsone 25 mg with clofazamine 50 mg twice weekly for 2 years. After 10 months, majority of the lesions subsided. Child was under regular follow up.

DISCUSSION
Histoid leprosy (HL) is a well-recognized clinical entity and is an expression of multi-bacillary (MB) leprosy. It is characterized by typical cutaneous nodules or subcutaneous plaques present over apparently normal skin with unique histopathology and characteristic bacterial morphology.

Histoid leprosy may appear de-novo or as a manifestation of drug resistance following irregular or inadequate treatment with dapsone monotherapy [6,7]. Histoid leprosy mimics several dermatological conditions like xanthoma, sarcoid, neurofibroma, dermatofibroma and post-kala-azar dermal leishmaniasis [5]. A few cases of HL masquerading as xanthomas have been reported in the
literature [8,9]. The presence of typical spindle shaped histiocytes, foam cells and AFB differentiates this condition from other conditions.

Though, the first case was thoroughly evaluated in USA histopathologically with IHC markers, simple investigation like skin smears for AFB was not done. Approximately 1-1.5 million people visit USA every year from India and other south eastern countries, which are known endemic areas for leprosy. When the patients from these areas present with nodular lesions or hypopigmented patches, leprosy should be considered as one of the differential diagnosis. Knowledge and awareness of leprosy are essential for all dermatologists for early diagnosis and prevention of disabilities. Skin smear examination is still a gold standard bed side investigation for diagnosis of lepromatous leprosy and its variants like histoid leprosy.

**CONCLUSION**

Though leprosy is no longer a public health problem in majority of the countries in the world, cases of multibacillary forms of leprosy have been frequently encountered in post-elimination-era.

Knowledge and awareness of leprosy are essential for all dermatologists and physicians for early diagnosis and management to prevent permanent disabilities.

**REFERENCES**