CASE REPORT

Isolated case of mucosal histoid Hansen’s disease of the nasal cavity in a post-global elimination era

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Summary  Histoid Hansen’s disease is a rare form of multibacillary leprosy with distinct clinical and histopathological features. This type of leprosy is a variant of lepromatous leprosy with a very high bacterial reserve. Of alarming concern is the discovery of an isolated mucosal histoid leprotic lesion inside the nasal cavity of a patient in the post-global leprosy elimination era. Our case had no history of leprosy or exposure to dapsone/multidrug therapy but had a heavy bacillary index. We are reporting this case to highlight the rarity of mucosal lesions due to histoid leprosy and involvement of the nasal cavity, as well as to create awareness and avoid misdiagnosis. This will help facilitate prompt treatment to minimize the complications and deformities of the patient and prevent its spread throughout the community. © 2015 King Saud Bin Abdulaziz University for Health Sciences. Published by Elsevier Limited. All rights reserved.

Introduction

Leprosy is a chronic infections granulomatous disease caused by Mycobacterium leprae with high
morbidity. Histoid leprosy is a rare variant of lepromatous leprosy with incidence varying from 1% to 2% among total leprosy patients and an average age at diagnosis is between 21 and 40 years old [1]. The adult population is most commonly affected with a male predominance [2]. The bacillary load is high in these patients and usually presents in the lepromatous types as well as in patients undergoing dapsone monotherapy and dapsone-resistant cases. Clinically, histoid leprosy presents as asymptomatic, with discrete, firm shiny papules and nodules on relatively normal appearing skin. The sites of predilection are the extensor surface of the extremities and lower trunk [1,3]. It is extremely rare in the genitalia, which occurs in more severe form [3]. However, the etiopathogenesis is unclear. The characteristic histopathological findings in conjunction with a high bacillary index confirm the diagnosis in the clinically doubtful cases. As the bacillary load is very high in these patients, they can form a potential reservoir of infection in the community. The mucosal type of histoid leprosy is extremely rare in the community. Here, we report a case of isolated mucosal histoid leprosy in a 45-year-old man from the post-elimination area of Odisha, India, where the prevalence rate of the disease was reported to be 1.47/10,000 people in March 2014 (NLEP) [4].

Case report

A 45-year-old male presented to the Outpatient Department (OPD) of Ear, Nose and Throat (ENT) with a right nostril block that existed for 6 months and intermittent nasal bleeding from same side (Fig. 1). The patient had neither rhinorrhea nor anosmia and had no history of serious illness. Upon inspection and an anterior rhinoscopy procedure, there was a small reddish mass observed in the anterior part of nasal cavity. A diagnostic nasal endoscopy was performed to confirm the lesion inside the nasal cavity and its site of attachment. A computed tomography (CT) scan of the nose and paranasal sinus was performed to determine the exact size and extent of the mass and showed that the mass was confined to the anterior region of the nasal cavity, primarily the anterior region of the inferior turbinate attached to septum. There was no history of sneezing or hyposmia, and the tar and throat examinations were normal. The general physical examination, systemic examinations and routine blood tests were within normal limits. Infectious diseases such as HIV, syphilis and tuberculosis were ruled out by ELISA, VDRL test and Mantoux test, respectively. The split skin smear examination for Lepra bacilli was performed on the bilateral earlobes, forehead, cheeks and chin and was negative at all of these sites. The mass was excised endoscopically under general anesthesia after taking the patient’s consent, and the mass was sent for histopathological examination.

The biopsy was reported as the histoid type of lepromatous leprosy (Fig. 2a and b). After this unexpected histopathology result, a more in-depth history was collected. The patient did not have any familial background of the disease and did not mention any contact with any persons suffering from leprosy. There were no cutaneous or peripheral nerve involvements, and neither hypoesthetic patches nor infiltration were discovered on the skin. The patient was referred to the outpatient department of dermatology for further evaluation and treatment of his histoid lepromatous leprosy. After confirmation of the diagnosis, the patient was started on anti-leprotic multibacillary therapy with rifampicin, clofazimine and dapsone. The patient was advised for regular follow up and has responded well to the previous 6 months of treatment.

Discussion

Histoid leprosy was first reported in 1963 by Wade [5]. It is an unusual multibacillary form of leprosy having unique clinical, histological and bacteriological findings. Irregular and inadequate therapies with resistance to dapsone and/or mutated organisms (histoid bacillus) are the primary factors that
cause histoid Hansen’s disease [6]. However, the patient reported here has no past history of leprosy or drug therapy for it. This form of Hansen’s disease has public health implications in this post elimination era as it belongs to the lepromatous pole with high infectivity and increased risk of bacillary transmission in the community. Histoid leprosy is considered to be a variant of lepromatous leprosy, and there exists an enhanced immune response against Mycobacterium leprae compared to lepromatous leprosy with respect to both cell-mediated and humoral immunity. Although there are an adequate number of macrophages, one study claimed that the macrophages are unable to kill the bacilli that exist in high numbers in the histoid lesion [6].

Histoid leprosy in nasal cavity may present with nasal obstruction, epistaxis, rhinorrhea and hyposmia. Evidence of nasal mucosa involvement is observed by anterior and posterior rhinoscopy. Diagnostic nasal endoscopy and radiological tests such as a CT scan of the nose and paranasal sinuses are useful tools to assess the lesion inside the nasal cavity.

Histopathological findings of the histoid nodules showed fusiform histiocytes arranged in whorled, criss-cross or storiform patterns. Within the histiocytes, acid-resistant bacilli were observed in large numbers. These bacilli are longer than the normal bacilli, uniform in length, more solid and are arranged in parallel bundles along the long axis of the histiocytes. There was epidermal atrophy and a traditional Grenz zone just below the epidermis [7,8]. There are three histological variants of histoid leprosy: pure fusocellular, fusocellular with epithelioid component and fusocellular with vacuolated cells, the latter of which is most commonly observed [9]. The mucosal histoid Hansen lesion clinically simulates different granulomatous lesions of the nasal cavity such as syphilis, tuberculosis, rhinosporidiosis and rhinoscleroma, as well as inverted papilloma and malignancy in elderly patients. Management of this condition includes treating the patient with a multidrug anti-leprosy regimen consisting of dapsone, clofazimine and rifampicin. Regular follow-up is advised for these patients to complete the treatment and prevent the spread of infection, as India was declared to have eliminated leprosy [10]. The mucosal histoid subtype serves as a reservoir of leprosy and as a source of new cases, which can create a serious threat to our elimination program.

**Conclusion**

The high bacillary load in the histoid variety of leprosy is a potential reservoir of the infection. This rare mucosal lesion in the nasal cavity could lead to increased likelihood of spreading the infection to others. It is essential to watch for new cases and provide complete treatment to achieve our goal of the elimination of leprosy. This case is unique in the sense that this mucosal variety of histoid leprosy is extremely rare, and this patient has no history of either leprosy or ingestion of dapsone/multidrug therapy. Histoid leprosy requires early detection and prompt treatment, which should be made a priority in the national program against leprosy.
Mucosal histoid Hansen’s disease

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Competing interests
None declared.

Ethical approval
Not required.

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[4] National Leprosy Eradication Programme (NLEP); 2014 March, Nlep.nic.in/Orissa.html

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