ABSTRACT
Hansen’s disease is on the verge of being eliminated from India and often missed by clinicians due to low index of suspicion. We present an unusual case in which greater auricular nerve thickening masqueraded as enlarged lymph node in the neck. The patient was referred for fine needle aspiration cytology, which revealed epithelioid cell granulomas suggestive of Hansen’s disease. Further clinical examination and investigations including the skin biopsy confirmed the disease, highlighting the role of pathologist in the management of such unusual presentation of a common disease.

Key words: Epithelioid cell granuloma; fine needle aspiration cytology; greater auricular nerve; Hansen’s disease

Introduction
Hansen disease continues to be a challenge to health worldwide. Despite widespread implementation of effective multidrug therapy, this disease is yet to be eradicated.[1] According to World Health Organization (WHO), the global registered prevalence of Hansen disease at the end of the first quarter of 2013 was 189,018 cases, while the number of new cases detected during 2012 was 232,857, with the Indian subcontinent recording one of the highest prevalence. One of the principal causes resulting in nondiagnosis and mismanagement is an unusual clinical presentation with a low index of suspicion of the disease.[2]

Although Hansen disease usually presents with hypoesthetic patches and nerve thickening, it may manifest with a wide spectrum of signs and symptoms. The most frequently affected nerve is the ulnar while the greater auricular nerve (GAN) is the most commonly involved sensory nerve.[3] We present a case of Hansen disease who presented with painless neck swelling simulating cervical lymphadenopathy. Fine-needle aspiration cytology (FNAC) raised the suspicion of Hansen disease, which was later confirmed by skin biopsy.

Case Report
The patient, a 48-year-old male, presented with swelling on the left side of the neck of 1-month duration [Figure 1a]. He denied any history of fever, weight loss or pulmonary symptoms or any other co-morbidities. Clinical examination revealed a 3.5 cm × 0.1 cm oval, firm, nontender swelling in the left posterior triangle. He was investigated for cervical lymphadenopathy. Routine hematological and biochemical tests were within normal limits, and he was seronegative for HIV.

Subsequently, during FNAC using a 23-gauge needle, patient complained of severe pain radiating to the left pinna and mastoid process, suggesting sampling from a neural tissue. Though the aspirate was scanty, the smears revealed numerous scattered epithelioid cells with few epithelioid cell granulomas admixed with lymphomononuclear cells [Figure 1b] without any lymphoglandular bodies. Owing to the meager yield and reluctance of the patient for a repeat procedure, stain for acid-fast bacilli could not be done. In view of the cytomorphological assessment suggestive of Hansen disease, a dermatological consultation was sought to evaluate skin lesions.
Detailed dermatological examination revealed two well-defined hypopigmented patches on the left side of the forehead measuring 0.5 cm × 0.5 cm and 0.4 cm × 0.4 cm. In addition, an ill-defined hypoesthetic patch was also noted in the lower 1/3rd of left pinna extending to the angle of mandible. Examination of nerves revealed grossly thickened left GAN (Grade II) and right GAN (Grade I). Bilateral ulnar, right radial and right posterior tibial nerves were also uniformly thickened and nontender. Overall dermatological evaluation was supportive of Hansen disease.

Biopsy from the hypoesthetic patch showed perivascular, periadnexal and perineural lymphocytic infiltration [Figure 1c] with numerous CD-68 immunoreactive histiocytes in the dermis [Figure 1d]. Fite Faraco stain of skin biopsy and skin scrape smear was negative for lepra bacillus. A diagnosis of borderline tuberculoid (BT) leprosy was confirmed and the patient was started on multidrug regimen, to which he responded with improvement in the condition.

Discussion

Hansen disease encompasses a wide clinicopathological spectrum ranging from localized paucibacillary tuberculoid form with anesthetic hypopigmented skin patch (TT) to the generalized multibacillary, lepromatous.[4] Between the two poles are unstable variants viz. BT, borderline and borderline lepromatous.

The skin biopsy in BT leprosy reveals granulomas with peripheral rim of lymphocytes affecting the neurovascular bundles and infiltrating sweat glands and erector pili muscles. Langhan type of giant cells vary in number with scanty acid-fast bacilli. Granulomas along the superficial vascular plexus are frequent, but they do not infiltrate into the superficial epidermis.[5]

The primary neuritic variety of leprosy is seen in 5-9% of leprosy patients in India and Nepal, which usually presents as peripheral neuropathy without any associated characteristic skin lesions or acid-fast bacilli on skin smears. Nerve biopsy is often required for diagnosis. However, few of these patients develop visible skin lesions during follow-up.[6] Although various different nerves may be affected, but the most commonly involved nerve is the ulnar nerve. The GAN, which is the most common of all the purely sensory cutaneous nerves, has been reported to be involved in 18.9%. The GAN is a branch of the cervical plexus (C2, C3), and an involved nerve may be clinically apparent as a swelling or thickening in the posterior triangle of the neck.[7]

The present case highlights an unusual presentation of Hansen in the form of a GAN thickening masquerading as a cervical lymphadenopathy. The case also emphasizes that should any patient complain of severe referred pain during the process of FNAC, a suspicion of Hansen should be considered, and the patient thoroughly evaluated for any skin lesions. Thorough clinical examination along with a high index of suspicion will ensure obtaining a definitive diagnosis and initiating appropriate therapy at the earliest.

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