Indian J Lepr 2020, 92 : 53-55

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Original Article - Case Series

Greater Auricular Nerve Thickening with Varied Skin Manifestations in Hansen's Patients

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Received : 30.03.2019 Accepted : 17.10.2019

Hansen's disease commonly presents with light coloured numb patches with a peripheral nerve thickening. Sometimes the nerve thickening may be the only clue to the diagnosis of the disease. Hence the peripheral nerve examination holds an important place in the diagnosis of Hansen's disease. Many a times an isolated nerve thickening may be ignored or misdiagnosed. A series of 3 cases with greater auricular nerve thickening with varied manifestations has been presented in this article. The first case had a skin lesion on the face which was mistaken for dermatophyte infection whose thickened Greater Auricular nerve was ignored. The second case had a isolated nerve thickening was misdiagnosed and managed as a case of external jugular vein thrombosis with heparin and hypolipidemic drugs. The third case had a thickened greater auricular nerve which he had ignored but later reported for treatment once his skin lesion had developed. Health professionals responsible for diagnosis of leprosy in peripheral settings as well as specialists working in referral institutions need to be aware of such varied manifestations of Hansen's Disease.

Keywords : Leprosy, Greater Auricular Nerve Thickening, Varied Manifestations

Introduction

Hansen disease, a chronic infection caused by *Mycobacterium leprae* mainly affects the skin, peripheral nerves, mucosa of the upper respiratory tract and the eyes. It usually presents as hypopigmented or slightly erythematous patches on the skin with loss of sensation and nerve thickening. But it can manifest with a wide spectrum of signs and symptoms. Peripheral nerve thickening is one of the cardinal features of Hansen's disease. The ulnar nerve is the most commonly involved nerve. But other nerves may be involved in the absence of ulnar nerve involvement. Sometimes an isolated thickened greater auricular nerve may be misdiagnosed as vein thrombosis (Sharma et al 2006, Ramesh et al 2007) or lymph node (Shilpi et al 2015) or ignored. We are reporting a case series of 3 cases of Hansen's disease with such isolated greater auricular nerve thickening with different presentations.

Case Series

Case 1 : A 22 years old male, resident of Uttar Pradesh presented with red raised lesion over left

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cheek for 3 months. He had been treated for the same with anti fungal and topical steroid creams but there was no response. He had reported to our department for the above complaints. Examination revealed a well defined hypopigmented hypoesthetic plaque on the left side of cheek. He was found to have left greater auricular nerve thickening which was uniform and non tender. Biopsy from the lesion was suggestive of BT Hansen's disease. He was started on Paucibacillary- Multi-drug treatment (PB-MDT).

Case 2 : Second case in the series was a 25 years old male resident of Bihar who presented with a painless swelling on right side of neck which he had noticed while taking bath. He had reported to a hospital for the same where he was admitted and investigated and diagnosed as a case of external jugular vein thrombophlebitis. He was started on Low molecular weight Heparin and Lipid lowering drugs. He was also referred to a Vascular Surgeon for further management. As there was no evidence of any vascular pathology in the external jugular vein on Doppler scan, he was referred to us. 3 months after the onset of swelling, he was reviewed in our department. He was found to have isolated right auricular nerve thickened. No skin lesions were seen. He was diagnosed as a case of Hansen's (Pure neuritic) and started on Paucibacillary MDT.

Case 3 : Third case was a 24 years old male, resident of Uttar Pradesh who reported with swelling over the right side of neck over a period of 6 months which he ignored. After 8 months he had developed a red, raised, numb patch over right side of Face. Examination revealed a well defined erythematous hypoaesthetic dry annular plaque on the right side of cheek. The right Greater Auricular nerve was grossly thickened without any signs of neuritis. He was evaluated and diagnosed as BT Leprosy and started on Paucibacillary-Multi-drug treatment (PB-MDT).

Discussion

The peripheral nerves are the initial sites of pathogenesis of leprosy. *Mycobacterium leprae* gain entry into the Schwann cells and multiply there. When the immunity of the individual is high, the infection may be curtailed within the nerve itself (Weddel & Pearson 1975). In India, primary neuritic type of leprosy is noted in 5-9% of Hansen's patients. They usually present with peripheral neuropathy without any associated characteristic skin lesions or acid-fast bacilli on slit skin smears. Nerve biopsy is often required for diagnosis. Few patients may develop skin lesions later during follow-up (Suneetha et al 2000).

Among the various sensory and motor nerves affected in leprosy, Ulnar nerve is the most commonly involved. Similarly the Greater Auricular Nerve, is the most commonly involved purely sensory cutaneous nerves. This has been reported to be involved in 18.9% (Dharmendra & Chatterjee 1978). The GAN is a branch of the cervical plexus (C2, C3). It runs upward parallel to the external jugular vein. It then bifurcates to provide sensory innervation to the skin over the mastoid process, outer ear and the parotid glands. It may be clinically apparent as a thickening or small mass or swelling in the posterior triangle of the neck if it is involved (Lynch & Johnston 1980, Saxena et al 1990, Neopane et al 2003). The Greater Auricular nerve can normally be seen in thin individuals and in those with good neck mobility but for better visualization the neck is stretched by turning to the opposite side. It is prominent in those with a good muscle mass.

The case series is presented to highlight the various presentations of Greater Auricular nerve thickening in 3 cases of Hansen's Disease. The first case had developed the skin lesion which was mistaken for dermatophyte infection possibly without giving due attention to the thickened Greater Auricular nerve. The second case with

isolated nerve thickening was misdiagnosed and managed as a case of External Juglar vein thrombosis with heparin and hypolipidemic drugs. The third case had a thickened Greater Auricular nerve which he had ignored but later reported for treatment once his skin lesion had developed. This is presented to emphasize the significance of proper peripheral nerve examination in a case of Hansen's disease and also to bring to light the point that even peripheral nerve examination will help in picking up the Hansen cases. Hansen's disease can present as isolated Greater Auricular nerve thickening per se. The skin lesions may or may not accompany the thickened nerve. In patients with facial patch of leprosy, Greater Auricular nerve is often thickened and enlarged along with enlargement of its transverse (anterior) cutaneous nerve (that is perpendicular to main nerve trunk). At times, this may be misdiagnosed and reported as external jugular vein thrombosis by Surgeons / Vascular surgeons / Internists as happened in one of cases in the present series.

In a country like India where leprosy continues to be prevalent, a high index of suspicion should be always there to arrive at a definitive diagnosis and initiate an appropriate therapy at the earliest. In an era of elimination of leprosy, we are still seeing new cases / varied presentation of Hansen's disease. Hence more awareness about the clinical features and varied presentations should be made especially among the budding doctors, doctors working in peripheral settings as well as specialists in referral centres/departments etc. There is a definite need to revamp the leprosy case finding activities in the peripheral and rural areas in the form of sensitizing the health care workers and providers. Special emphasis should be made on early detection and treatment should be made.

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How to cite this article : Sivasankari M, Chopra A and Mitra D (2020). Greater Auricular Nerve Thickening with Varied Skin Manifestations in Hansen's Patients. *Indian J Lepr.* **92**: 53-55.