Leprosy and Neurofibromatosis 2: What is Common?

TO THE EDITOR:

The neurofibromatoses are a heterogeneous set of genetic disorders having clinical manifestations that involve the skin, the nervous system or both. Although this condition is unrelated to leprosy, common clinical as well as histological features have aroused the interest of clinicians in the field of leprosy. Leprosy in association with von Recklinghausen neurofibromatosis (NF1) has been reported previously ([2, 3, 8, 9]), but to the best of our knowledge, the association of leprosy with neurofibromatosis 2 (NF2) has not been reported in the past. We wish to share our interesting observations of histoid leprosy occurring in a patient with NF2.

CASE REPORT

A 36-year-old male patient presented to us with complaints of progressive sensory loss over the hands and feet for a duration of 3 years. He also noticed multiple papulonodular lesions over his body and deafness for the last 6 months. There was no symptom suggestive of leprosy reaction in the past. The patient had not received any treatment for his complaints previously. One examination, multiple skin-colored to erythematous papulonodular lesions were seen over the face, trunk and extremities, varying in size from 0.5 cm to 1.5 cm. The nodules were waxy, firm, nontender and were present over apparently normal-looking skin. In addition there was diffuse infiltration of the face and ears (Fig. 1) and thickening of the peripheral nerve trunks involving ulnar and common peroneal (lateral popliteal) nerves bilaterally with glove-and-stocking type of anesthesia.

A clinical diagnosis of histoid leprosy was made which was confirmed by histopathology. A biopsy of one of the nodules on histopathological examination showed numerous spindle-shaped histiocytes forming interlacing bands and whorls in the dermis. Numerous acid-fast bacilli were seen that were slightly longer than normal along with few foamy histocytes. Slit-skin smear examination from the nodules and the earlobes demonstrated a bacterial index (BI) of 5+. Routine hematological, biochemical and radiological investigations were within normal limits. Initially, the deafness was thought to be due to ear pathology secondary to leprosy, but on suspicion of associated vertigo, brain stem-evoked response audiometry (BERA) was done, which suggested a retrocochlear pathology. On further investigations, a magnetic resonance imaging (MRI) scan (Fig. 2) revealed a well-defined, approximately 2 × 2.5 cm heterogeneous mass lesion in both CP angles with a well-defined intracanalicular component of the lesion on both sides, features sugges-

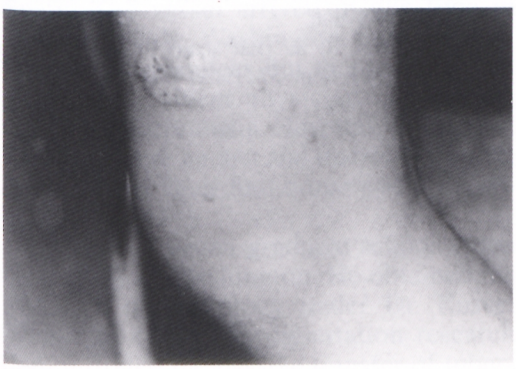

FIG. 1. Papulo-nodular and plaque lesions of histoid leprosy.

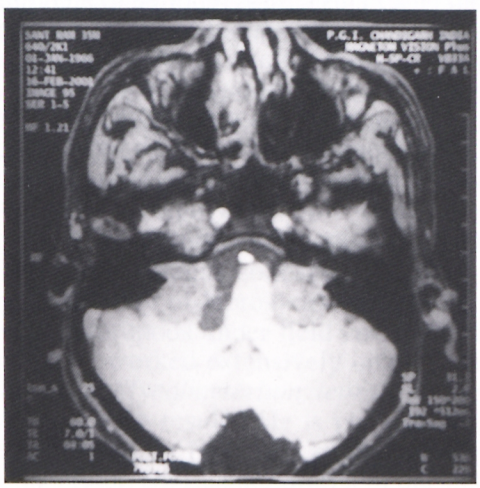

FIG. 2. MRI showing heterogeneous mass lesions in both CP angles suggestive of bilateral acoustic schwannomas.

tive of bilateral acoustic schwannomas. There was another cystic lesion along the posterior part of the cervicomedullary junction with an enhancing nodule anteriorly, suggestive of ependymoma. Orbits, basal ganglia, thalami, sella and corpus callosum were normal. All of these radiological findings confirmed the diagnosis of NF2, which was probably responsible for the vertigo and deafness in the patient.

DISCUSSION

The co-existence of leprosy and neurofibromatosis in the same patient is interesting in that Schwann cells are involved in the pathogenesis of both and masquerade each other clinically. Cutaneous and subcutaneous neurofibromas may be mistaken for nodules of lepromatous leprosy ([1]). Peripheral nerve trunk thickening in neurofibromatosis has been recognized as a clinical differential diagnosis of leprosy ([5, 6]), but such clinical attention is not shared by tumors of NF2 arising in the central nervous system. The literature reveals contradictory findings about the affection of the audiovestibular system in leprosy. There are reports of high incidence of hearing loss ([10]) and vestibular hypofunction ([7]). Mann, *et al.* ([4]) in a case control study concluded that leprosy selectively affects the cochlear end organs without affecting the vestibular functions. Bilateral vestibular schwannomas, the hallmark of NF2, can mimic audiovestibular involvement of leprosy as in our case. This case is reported to alert clinicians that audiovestibular symptoms in leprosy may not always be related to the primary disease. Although the frequency of central neurofibromatosis (NF2) is much less compared to NF1, a high index of suspicion should be kept in a leprosy patient who has vestibular symptoms associated with sensorineural deafness.

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