

# Lepromatous leprosy masquerading as neurofibromatosis

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## Introduction

Leprosy is fairly common in the Commonwealth of the Northern Mariana Islands (CNMI) as it is throughout Micronesia. The disease can have widely variable manifestations. Skin lesions may be hyper- or hypo-pigmented, may be anaesthetic or with sensitivity, may be flat, plaque-like, domed-shaped or nodular. In addition, leprosy may present with neuritis, iritis, orchitis and nasal congestion.

## The three cases

Presented here are three cases of leprosy in the CNMI which were mistaken initially for neurofibromatosis.

### Case 1

A 30 years old Chuukese male, presented to Commonwealth Health Centre in 1993 with multiple asymptomatic nodules on his trunk and upper arms which are said to have been present for 2 months. A diagnosis of "neurofibromas" was made. The patient returned in 4 months later and was seen by a different physician who diagnosed "neurofibromatosis vs. molluscum contagiosum" and sent a skin biopsy which revealed changes diagnostic of leprosy. The patient then completed 8 months of treatment with dapson, clafazamine and rifampin per WHO protocol. His adherence to treatment was irregular and he was lost to followup after 8 months. He is rumoured to have moved out of the CNMI.

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### Case 2

A 22 year old Carolinian male, presented in 1984 to Commonwealth Health Center complaining of a chronic skin condition. On physical examination "nodules and welts were noted on his trunk. A diagnosis of (1) probable neurofibromatosis, and (2) urticaria, was made and he was treated with an antihistimine. In 1986, 25 months later, he presented again with a "one month history of multiple papules" and exam remarkable for skin "covered with hundreds of 5mm-2cm papules and nodules." Biopsy revealed changes diagnostic for leprosy and the patient was started on a regime of dapson and rifampin. He was lost to followup after 3 months of treatment. Between January 1987 and 1992 the patient had several visits for minor illnesses and lacerations. Physical exam was notable for "skin nodules" and "nodular dermatitis" on two of these visits but it was not until August of 1992 that a biopsy was taken again for "nodular dermatitis, ?etiology" and the presence of acid fast bacilli diagnostic of leprosy was again noted. Therapy was restarted with dapson, clafazamine, rifampin per WHO protocol. Ulnar neuropathy with associated deformity of the right hand was noted at this time. The

patient adhered irregularly but finally completed his treatment in 1995.

### Case 3

A 55 years old Carolinian male, presented in 1987 with "raised, non-pruritic rash" and "pain down right leg." He was next seen in the emergency room for chest pain 25 months later, in 1989

where "multiple neurofibromas" of the upper body were noted. He was seen in clinic 3 months later for bronchitis and also diagnosed with "nodular dermatitis, suspect neurofibromatosis". These skin lesions were noted on 3 additional clinic visits over 6 months until a physician recognized his characteristic "leonine facies" and confirmed the presence of leprosy by skin smear. At that time the patient was also noted to have a neuropathic right foot with partial foot drop. He has since completed a full course of treatment with apparent cure of his disease.

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## Discussion

In these three cases from a single health centre within the past 10 years, leprosy was initially mistaken for neurofibromatosis, leading to delays of 4 months, 25 months and 31 months between the times that the patients presented with skin findings and the times that correct treatment was begun. In case number 2 the patient was lost to follow up after only 3 months of treatment following which almost 6 years and several clinic visits (noting skin abnormalities) elapsed before that case was again recognized as leprosy and treatment reinitiated.

Prompt recognition of leprosy is important for preventing permanent neurological damage as occurred in cases #2 and #3. Prompt recognition and treatment of multibacillary cases such as these is also the key to preventing spread of the disease to others in the population.<sup>1,2</sup>

Lepromatous leprosy and neurofibromatosis may both present with widespread asymptomatic nodular skin lesions. Since the lesions are asymptomatic and since the diagnosis of neurofibromatosis has limited therapeutic implications, physicians who do not consider leprosy in the differential diagnosis of these patients may feel no urgency in confirming their impression with lab tests.

Features of leprosy which distinguish it from neurofibromatosis include change of facial features (ie. "leonine facies as in case #3), associated tenderness and thickening of nerve trunks, areas of anaesthesia, presence of skin lesions such as plaques, papules and areas of induration in addition to the nodules, (as in cases #2 and #3).

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Skin smears for acid fast bacilli are a very sensitive means of diagnosis for advanced forms of multi-bacillary leprosy, such as lepromatous leprosy with multiple nodular lesions. This fast, safe and cheap test should be used in any case of nodular dermatitis of uncertain etiology. Skin biopsy of nodular lesions, which is more expensive and requires a pathologist for interpretation, will also usually reveal changes diagnostic of leprosy.<sup>3</sup>

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## References

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The multitude of the sick shall not make  
us deny the existence of health.

*R. W. Emerson*

'The Conduct of Life'