Tic Douloureux as a Presenting Feature of Facial Leprosy: Diagnostic Enigma in Taiwan

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Abstract-

Purpose: Leprosy is rarely seen in Taiwan. We herein report a foreign worker concomitantly with facial borderline tuberculoid leprosy presenting with trigeminal neuralgia.

Case Report: A 26-year-old male foreign labor from Indonesia, presented with 1 year history of a hypoanesthetic erythematous plaque of right face and subsequent 6 months constant, severe pain in the right side of his face over the nasolabial groove. Biopsies and histopathological examination confirmed the diagnosis of leprosy. We treated the patient with a multidrug regimen including dapsone, clofazimine, and rifampine since April of 2012 with a good response.

Conclusion: We report a rare case of new-onset leprosy presenting with trigeminal neuralgia in Taiwan and suggest leprosy should be listed in the differential diagnosis of unusual skin manifestations and neuralgia.

Key Words: trigeminal neuralgia, tuberculoid leprosy, leprosy, ticdouloureux, dapsone

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INTRODUCTION

Trigeminal neuralgia, also known as tic douloureux, is a chronic and disabling facial pain. The condition has no clear-cut cause[(1)]. In all cases, though, an excessive burst of nervous activity from a damaged nerve causes the painful attacks. A more recent notion is that the cause stems from biochemical change in the nerve tissue itself[(2)].

The presentation is not always typical and variations may not always be easy to diagnose. Leprosy is one of the most important causes of peripheral neuropathy of the world but scares in modern Taiwan. A population increase from 3.3 to 21.7 million, several tides of immigration and national leprosy control programs, from 1910 to 1997[(3)]. Leprous neuropathy is characterized by the involvement of superficial nerve trunks in cooler regions of the body. Cranial nerve involvements, including the trigeminal nerve, are also commonly seen in patients of leprosy[(4)]. Trigeminal neuralgia has been reported to mimic leprosy or vice versa[(5)]. This case report describes a patient with trigeminal neuralgia, who

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later diagnosed as mononeuritis due to leprosy.

CASE REPORT

A 26-year-old foreign foreign labor from Indonesia, presented with 1 year history of a hypoanesthetic erythematous plaque of right face and subsequent 6 months constant, severe pain in the right side of his face over the nasolabial groove. The pain had been progressively worse and was usually precipitated by brushing his teeth, shaving, washing his face, eating, and talking. Cold water over his face made the pain worse as well as cold breeze hitting his face. The pain at times would radiate to the rest of his face. He has had no further pain in the past 10 years. He did not consume alcoholic drinks or smoke. Empirical treatment with an antibiotic, with ketoconazole, and with acyclovir was unsuccessful. Family history was not contributory.

Clinical examination demonstrated mild sensory loss over the right V2 and V3 dermatome. Corneal reflex was absent at right side. There was one well-defined hypopigmented lesion measuring 2×2.5 cm, with impaired sensation to touch, pain, and temperature on right side cheek (Figure 1). Skin biopsy was obtained for histopathology and was reported as showing borderline tuberculoid pathology and the absence of acid fast bacilli. A biopsy of the cutaneous plaque revealed well-defined epithelioid cell granulomas in the upper and mid dermis (Figure 2). Immunohistochemical study for S-100 revealed a recognizable nerve with infiltration of perineurium by lymphocytes and/or formation of intraneural epithelioid cell granulomas. Nerve damage was observed with some fragmented strips within perineurium. (Figure 3) which correlated with fragmented type (pattern B) S100 staining[6].

The x-ray films of the chest were normal. The following laboratory tests were negative or normal: blood cell counts, serum electrolytes, liver and kidney functions tests, antinuclear antibodies, rheumatoid factor, Serological tests for HIV 1 and 2, Anti-Neutrophil Cytoplasmic Antibodies (ANCA) and hepatitis B and C were also negative. Computed tomography imaging of brain, orbit, and cervical spine revealed no abnormality. The clinical impression was trigeminal neuralgia and mononeuritis of trigeminal nerve due to leprosy.

The patient was put on WHO multidrug therapy for paucibacillary leprosy, dapsone 100 mg daily plus rifampin 600 mg monthly, for 6 months[7]. The patient was deported to Indonesia within one month. Skin lesions and trigeminal neuralgia regressed within one week.

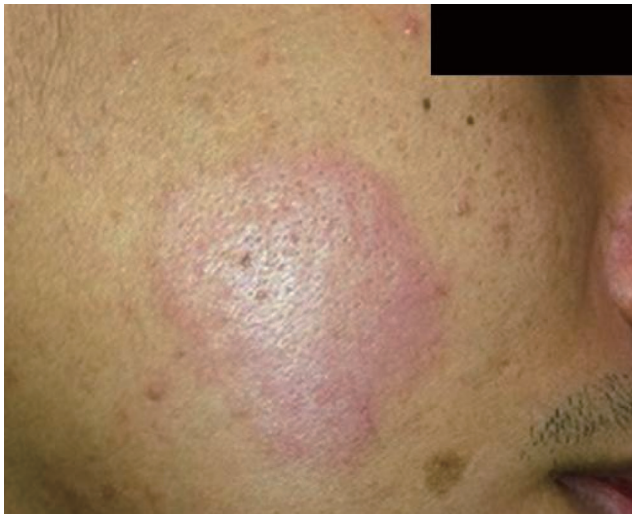

Figure 1. An annular erythematous, indurated plaque 2×2.5cm in size, with anesthesia in right face was noted.

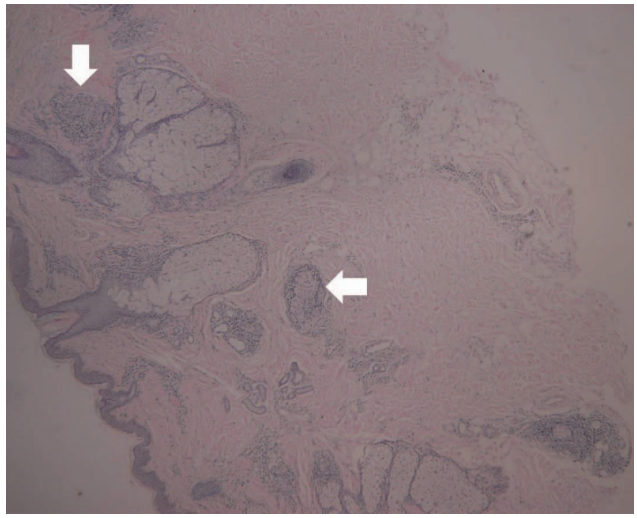

Figure 2. Granulomatous infiltration in the superficial, and deep dermis and around the skin appendages (arrows). (H&E stain, ×20)

DISCUSSION

Leprosy is the rare cause of peripheral neuropathy in Taiwan but probably the most common cause in the world. Only nine cases of leprosy were detected in 2002 [8]. The number of leprosy cases in Taiwan is declining by the year, the importance of cases imported from outside is becoming greater[3]. Leprosy is endemic in Indonesia where this foreign worker came from. The registered cases are Indonesia were 70,961.[9]

Around the world, the most common disorder that might present with these findings is leprosy. Borderline tuberculoid leprosy, produces solitary or regional skin lesions[10] and result in this man's cranial-nerve signs[11]. The trigeminal nerve is the second most common cranial nerve to be affected in leprosy. Damage to the trigeminal nerve leads to corneal anesthesia[4]. In our case, presence of corneal anesthesia suggests trigeminal nerve involvement. Trigeminal neuralgia was reported as a presenting feature of leprosy in endemic area. It is possible that the trigeminal neuralgia response in the present patient may be due to dapsone, as successful weaning of pain-killer was possible only after completion of antileprotic therapy.

In the absence of acid fast bacilli of skin biopsy, differential diagnosis including sarcoidosis and Wegener's granulomatosis deserves careful consideration in this case, principally because it may cause asymmetric subacute or chronic cranial-nerve involvement[12]. Since Mycobacterium leprae are rarely demonstrable in the tuberculoid spectrum of leprosy, a confirmatory diagnosis of leprosy can be made on the basis of finding active destruction of cutaneous nerves by granulomatous inflammation in a skin biopsy using S100 immunohistochemical study (Figure 3)[15]. Immunoperoxidase staining for S-100 protein, which is a marker for Schwann cells, was used to delineate nerves in lesional skin biopsies of patients with tuberculoid and borderline tuberculoid leprosy. Furthermore, the fragmented strips within perineurium positive for S100 staining suggested the active destruction of nerve which was phagocyted by dendritic cell. Four different patterns of nerve damage were observed: infiltrated, fragmented, absent, and intact[6,15].

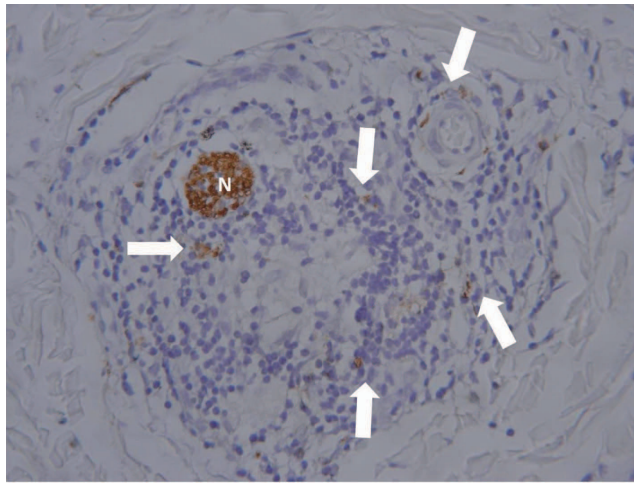

Figure 3. Immunohistochemical study for S-100 revealed a recognizable nerve (N) with infiltration of perineurium by lymphocytes and/or formation of intraneural epithelioid cell granulomas. Nerve damage was observed with some fragmented strips within perineurium (arrows). (S-100 immunohistochemical stain)

Gupta SK et al., addressed such fragmented staining patterns had sensitivity, specificity, and positive and negative predictive values of 100% in diagnosing tuberculoid leprosy[6]. All of the nonleprous granulomatous dermatoses showed only intact nerves, either inside or outside the granuloma, and so S-100 staining can be used to rule in leprosy[15].

In this subject, the skin biopsies failed to reveal sarcoidal granulomas in the superficial subcutaneous or submucosal connective tissue[13,14]. The normal x-ray films of the chest also suggest that sarcoidosis is not the correct diagnosis[12]. Wegener's granulomatosis also may present with unilateral cranial neuropathy and occasionally produces subcutaneous nodules[16]. Could this patient have had a limited form of Wegener's granulomatosis? Even in a limited form of Wegener's granulomatosis, however, some evidence of a systemic illness, such as a history of upper respiratory tract symptoms, evidence of pulmonary or renal involvement, anti-neutrophil cytoplasmic antibodies (ANCA), a normochromic, normocytic anemia, or an elevated erythrocyte sedimentation rate, should be present[17].

In summary, we report a rare case of new-onset lep-

rosy presenting with trigeminal neuralgia in Taiwan and suggest leprosy should be listed in the differential diagnosis of unusual skin manifestations and neuralgia.

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