Case report

Neural leprosy: A case report

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Abstract

Neural leprosy is characterized by neurological deficit without skin lesions, with a prevalence ranging from 1% to 17.7%. Diagnosis might be difficult and need a multidisciplinary approach. This is a case of axonal type motor and sensory polyradiculoneuropathy of the peripheral facial nerve. A 26-year-old woman was referred from the neurology clinic with facial paralysis, suspected as leprosy. Physical examinations were as follows: no skin lesions, left eye lagophthalmos, thickening of right lateral peroneal and bilateral posterior tibial nerves, sensory impairment, peripheral bilateral facial palsy, and wasting of bilateral distal small muscles of the hands, with normal autonomic function. Nerve Conduction Study revealed multiple demyelinating mononeuropathy of upper and lower extremities. Her serum anti-PGL-1 IgM level was 1721 μ/mL, but after three months of treatment with MDT-PB regimen, it increased to 2815 μ/mL. Therefore, the treatment was switched to MDT-MB regimen and 30 mg prednisone. The patient is still undergoing treatment. There has been a slight improvement after treatment with MDT-MB regimen. Nerve biopsy is the gold standard for diagnosis but has its limitations. However, serological test of anti PGL-1 can be a marker and a useful tool as an additional test to confirm the diagnosis, especially for patients with nerve impairments. Difficulties are due to the absence of skin lesions and neuropathy which may be caused by other diseases. Both diagnosis and treatment require multidisciplinary approach. Treatment given is intended to correct nerve damage and prevent further disabilities.

Key words: Neural leprosy, anti-PGL-1, systemic corticosteroid, multidisciplinary approach

Abstrak


Kata kunci: Kusta neural, anti-PGL-1, kortikosteroid sistemik, pendekatan multidisiplin

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**Introduction**

Neural Leprosy or neural type Morbus Hansen (MH) is a leprosy characterized by loss of sensory function in the area along the nerve trunk distribution without a skin lesion.\(^1\)\(^2\) The prevalence of neural leprosy ranges between 1-17.7% of all leprosy cases.\(^3\)\(^4\)

Definite diagnosis cannot be established based on clinical features alone, and it can be a challenging case for physicians.\(^3\) Slit skin smear examination is usually negative. Diagnosis can be made based on clinical examination, with thickening of the nerve at the predilection site. However, peripheral neuropathy can be caused by many other etiologies, such as metabolic diseases, drug reactions, genetic disorders, and other infections. Until recently, nerve biopsy is the gold standard in the diagnosis of neural leprosy, but the implementation is not easy and it has its limitations.\(^2\)\(^4\) Anti-PGL-1 can be used as an additional examination in establishing neural leprosy cases.\(^3\) In addition, collaboration with related fields of science is needed in order to establish an integrated management of this disease.

**Case Illustration**

A 26 years old woman came to the Dermatology and Venereology Outpatient Clinic of dr. Cipto Mangunkusumo National Hospital (RSCM). She was referred from the neurology outpatient clinic with a chief complaint of facial stiffness starting seven years ago. Her initial complaints were that her eyes and eyebrows could not be moved, her mouth could not be closed, and her cheeks could not be bulged. The patient was treated at the neurology outpatient clinic for two years, but not in a regular manner. The patient then went to Neurology Outpatient Clinic (Department of Ear Nose and Throat/ ENT), because of facial muscle stiffness, suspected as a peripheral facial nerve paralysis. The facial stiffness gradually worsens, coupled with paresthesia on bilateral upper and lower limb. Since an autoimmune disorder was suspected, the patient was also referred to the Allergy and Immunology Division, Department of Internal Medicine.

History of contact with MH patients was denied, and no one in the family has had a similar sickness. Lagophthalmos in both eyes, lower facial asymmetry, disability to close the mouth and puffing cheeks were findings of the physical examination. No disturbance in sensibility and skin lesions were found. The enlargement of bilateral posterior tibia nerve and right lateral peroneal nerve were found. Autonomic nerve function was not compromised and there were no abnormal nerve reflexes. Laboratory tests showed anti-PGL-1 IgM of 1721 μ/mol (cut off <605 μ/mol) and Anti-Nuclear Antibody (ANA) were positive. The Department of Neurology performed electromyography (EMG) and Nerve Conduction Study test and concluded that there were mononeuropathy and multiple demyelination of superior and inferior extremities, peripheral bilateral facial palsy and wasting of bilateral distal small muscles of the hands. Positive result of ANA test is used to help determine whether the patient has an autoimmune disorder, however ANA test alone could not diagnose a specific disease.

![Figure 1. Lagophthalmos in both eyes.](image)

![Figure 2. Lower facial asymmetry](image)

Based on history, physical examination, and supporting examinations, the patient was diagnosed with neural leprosy by the Department of Dermatology and Venereology, motor and sensory axonal type polyradiculoneuropathy and functional lesion of the peripheral facial nerve by the Department of Neurology, and Chronic Inflammatory Demyelinating Polyneuropathy (CIDP) by the Department of Internal Medicine. The patient was also consulted to the Department of Ophthalmology with myopia and dry eyes. The patient had also undergone acupuncture therapy as suggested by the neurologist.
The patient was treated with paucibacillary (PB) multi-drug treatment (MDT). After 3 months of treatment, there was an elevated level of anti-PGL-1 of 2815 μ/mL and treatment was switched to MDT-MB. An initial dose of prednisone 30 mg/day was added to the treatment. The level of IgM anti PGL-1 decreased to 1729 μ/mL after the treatment has completed. Currently the patient is still being treated in the Dermatology and Venereology, Neurology, and Internal Medicine Departments.

The consultation to the Department of Ophthalmology serves as a supportive treatment to prevent further damage to the eye. The patient was treated with Oculenta® gel and eye drops which contains carboxymethyl cellulose. Both serve as lubricants. As an adjuvant therapy, the patient had undergone acupuncture therapy for four times, but did not continue because there was no significant improvement.

Allergy and Immunology Division of Department of Internal Medicine prescribed mycophenolate acid 180 mg twice daily as an immunosuppressive agent, calcium and vitamin D3, as well as folic acid. Currently, the patient has completed the multi-bacillary (MB) MDT treatment with good response, as assessed subjectively. EMG and Nerve Conduction Study examination have been repeated after the completion of treatment and the result showed an improvement, despite the fact that there was no significant difference in terms of physical appearances.

Discussion

The diagnosis of neural leprosy in this patient was made based on impaired neurological function, peripheral nerve thickening, Nerve Conduction Study, and the anti-PGL-1 IgM test result. The most commonly found clinical symptoms of neural leprosy are: skin sensory disturbances, numbness, paresthesia, nerve pain, and nerve thickening. Persistent complaints and symptoms (for as long as seven years) can actually determine the patient’s prognosis. When an event of complete paralysis has occurred for more than 1 year, the nerve damage would be irreversible and its function cannot be restored even with medical treatment. Damage to the zygomatic facial nerve branches innervating the orbicularis oculi muscle can be reversible if treated appropriately before 6 months.

Examination of anti-PGL-1 can be used as a marker of Mycobacterium leprae bacterial load. There is a significant correlation between anti-PGL-1 IgM with bacterial index, which showed that the levels of anti-PGL-1 IgM reflects the total bacterial load on leprosy patients and can be used to evaluate treatment. Levels of anti-PGL-1 IgM will decrease after medical treatment. However, nerve biopsy remains the gold standard for diagnosing neural leprosy case. The patient’s anti-PGL-1 IgM level prior to treatment was 1721 μ/mL, which then increased to 2815 μ/mL after three months of treatment with MDT-PB regimen. However, nerve biopsy remains the gold standard for diagnosing neural leprosy case. The patient’s anti-PGL-1 IgM level prior to treatment was 1721 μ/mL, which then increased to 2815 μ/mL after three months of treatment with MDT-PB regimen, but decreased to 1729 μ/mL after completing a regimen of MDT-MB. Elevated IgM level during treatment is probably due to fragmentation of Mycobacterium. It is necessary to educate the patient that the disease was caused by bacteria attacking the nerves, and that when the nerve damage has become permanent, it will be difficult to treat any neurological problems which may arise.

Patient was treated first with MDT-PB regimen due to mononeuropathy as shown in neurological examination. After three months, the treatment was changed to MDT-MB regimen because the level of anti-PGL-1 has increased. Corticosteroids was added to the treatment and then tapered off, with the initial dose being 30 mg prednisone. Corticosteroids work as a nerve protector which can prevent further nerve damage.

Conclusion

Neural leprosy is a type of leprosy which is difficult to diagnose as it resembles other neurological disorders. Neural leprosy needs to be diagnosed early and the treatment requires multidisciplinary approach, especially in cases encountered late in the course of the disease. Together with clinical information, serological tests to detect the level of IgM to PGL-1 are useful to assist in establishing a diagnosis.

References