Chronic Inflammatory Demyelinating Polyneuropathy In A Patient With A Leprosy Reversal Reaction: A Case Report


Abstract

Chronic inflammatory demyelinating polyneuropathy (CIDP) is a condition that affects the peripheral nervous system with progressive limb weakness, with predilection for proximal muscles, sensory loss and areflexia and it has a relapsing or progressive course. The physiopathology is still unclear, but it is probably an autoimmune disease involving autoreactive T and B cells. CIDP is well described in association with many diseases but not with leprosy reaction which is an immunologically mediated episode of acute inflammation that occurs in any time of leprosy disease, including after the multidrug therapy. Here we presented a patient who developed CIDP in the context of a leprosy reversal reaction, years after the end of leprosy treatment who recovered for both conditions after corticosteroid therapy. This patient’s CIDP presentation could be attributed to cell injury caused by type 1 reaction that exposed the neural antigens and incited an autoimmune reaction.

Oswaldo Cruz Institute Rio de Janeiro, Avenida Engenheiro Martins Romeo, 78/501 Inga Niterói RJ CEP 24210400, Brazil

*Corresponding author
Andrade LR, Oswaldo Cruz Institute Rio de Janeiro, Avenida Engenheiro Martins Romeo, 78/501 Inga Niterói RJ CEP 24210400, Brazil, E-mail: ligiaandradee@gmail.com.

Submitted: 27 Sep 2017; Accepted: 07 Dec 2017; Published: 12 Dec 2017

Keywords: Leprosy Reactions, Reversal Reaction, Type 1 leprosy Reaction, Chronic Inflammatory Demyelinating Polyneuropathy

Introduction

Chronic inflammatory demyelinating polyneuropathy (CIDP) is a condition affecting the peripheral nervous system [1,2]. The classic form of the disorder is characterized by: (1) progressive limb weakness, usually with a predilection for proximal muscles, sensory loss, and areflexia with a relapsing or progressive course; (2) electrophysiological features of demyelination, including prolonged distal motor and F-wave latencies, reduced conduction velocities, and conduction block and temporal dispersion; (3) laboratory features of albumino-cytological dissociation in the cerebrospinal fluid; and (4) inflammation, demyelination, and remyelination on nerve biopsy. The etiology of CIDP is still unclear, but it is widely accepted that it is an autoimmune disease with underlying immunopathology involving autoreactive T cells and B cells. Because CIDP has an autoimmune basis, it can occur in association with diseases such as HIV infection and hepatitis C, Sjogren’s syndrome, monoclonal gammopathy of unknown significance, melanoma, lymphoma, diabetes and inflammatory bowel diseases [1,3,4].

Leprosy is a complex dermato-neurologic disease caused by Mycobacterium leprae that presents a wide spectrum of clinical manifestations characterized by distinct bacteriologic, immunologic and histopathologic features. One of the main difficulties in the clinical management of leprosy patients is the development of leprosy reactions that can occur anytime during the chronic course of the disease: before diagnosis, during treatment and even years after treatment release [5,6,7]. Leprosy reactions represent immunologically mediated episodes of acute inflammation that if not diagnosed and treated promptly can cause irreversible impairment of nerve function and permanent incapacities [8]. There are two major types of leprosy reactions: type 1 reaction (T1R) or reversal reaction (RR) which is associated with TH1-type immunity and type 2 reaction (T2R) represented mainly by erythema nodosum leprosum (ENL) which is related to TH2-type immune responses. Type 1 reactions typically result in inflammation and pain in preexisting lesions, which may also ulcerate. These reactions may also produce increased neuritis manifested as tenderness and nerve damage [6,9-12]. It typically occurs within the first 6 months after the start of multibacillary therapy, although they can happen at any stage of the disease process [13-15]. We report a case of a patient with reversal leprosy reaction 12 years after Multibacillar treatment associated with chronic demyelinating polyneuropathy.

Case report

A 52-year-old man presented at the year at 2000 a picture of erythematous and scaly skins lesions on limbs, back and feet for four years of evolution. He had no fever, edema, arthralgia or paresthesias. His neurological exam and the nerve conduction study were normal at that time. He had a skin biopsy and was diagnosed as borderline-borderline leprosy disease. He started with multibacillary multidrug therapy for 12 months. By the end of the
Although the CIDP is associated with several disorders, it is not a immunemediated condition, but unlike Guillain-Barré syndrome, damaged are not fully understood. Most evidence suggests that it is mechanisms that trigger CIDP and the way in which the nerves are perineuritis, granuloma with multinucleated giant cells, lymphocytic exacerbation of skin lesions, painful swelling of nerve trunks, and of increased cell-mediated immunity. They lead to swelling and the type I reactions, also known as reversal reaction are the result of Mycobacterium leprae, host immune responses and drug toxicity [16]. The pathogenesis of neuropathy in leprosy is diverse and not well described it’s relation with leprosy reactions. Abundant experimental evidence suggests that CIDP is caused by an aberrant immune response that involves autoreactive T cells, macrophages, and autoantibodies. However, despite extensive research over the last decades, the target antigens of the humoral and cell-mediated immune response are yet poorly defined [19-21,1].

A spinal fluid exam was performed and showed a protein-cytologic dissociation (2mg/dL of leukocytes and 149 mg/dL of proteins). Hemogram, liver and renal function were normal. A neuroconduction study revealed reduced conduction velocity and prolonged latencies on motor median, ulnar, peroneal and tibial nerves. The Study of the F wave showed response with prolonged latencies in the fibular, ulnar and median nerves. The findings were compatible with motor sensitive polyradiculoneuropathy with demyelinating predominance.

A skin biopsy was performed and the findings were typical of reversal reaction, with the dermis exhibiting perianexial and perivascular edema and granulomatous infiltrate, dissociating erector muscle from the hair and invading nerve fille.

The patient was diagnosed as chronic inflammatory demyelinating polyneuropathy and leprosy reversal reaction. He was treated with pulsoteraphy with methylprednisolone 1g intravenously per day for 3 days in a row, then once a week for 4 weeks and once a month until he completed 6 months of treatment. After 2 months he presented no skin lesions.

The patient gradually improved his neurological condition and by the end of the treatment he was able to walk by his own, only remaining with a mild atrophy in the left quadriceps.

Discussion
The pathogenesis of neuropathy in leprosy is diverse and not fully understood. Mechanisms include Schwann cell invasion by Mycobacterium leprae, host immune responses and drug toxicity [16].

The type I reactions, also known as reversal reaction are the result of increased cell-mediated immunity. They lead to swelling and exacerbation of skin lesions, painful swelling of nerve trunks, and sensory and motor deficits. Nerve biopsies in type I reaction show perineuritis, granuloma with multinucleated giant cells, lymphocytic infiltration and vasculitis [17].

CIDP is a chronic demyelinating condition that causes symmetric weakness of the limbs and paresthesias in its typical presentation. The mechanisms that trigger CIDP and the way in which the nerves are damaged are not fully understood. Most evidence suggests that it is a immunemediated condition, but unlike Guillain-Barré syndrome (GBS), usually there is no antecedent illness [18].

Although the CIDP is associated with several disorders, it is not well described it’s relation with leprosy reactions. Abundant experimental evidence suggests that CIDP is caused by an aberrant immune response that involves autoreactive T cells, macrophages, and autoantibodies. However, despite extensive research over the last decades, the target antigens of the humoral and cell-mediated immune response are yet poorly defined [19-21,1].

Reported a case of a 52-year-old woman with a past history of lepromatous leprosy and 14 years later presented with progressive weakness and severe arm/leg pain, cerebrospinal fluid analysis revealed protein-cytological dissociation, skin and sural nerve biopsy showed no bacilli. Immunomodulatory treatment led to clinical and neurophysiologicalimprovement. The patient was diagnosed with CIDP and type 2 leprosy reaction [22]. They postulated that since in type 2 reactions there is complement activation and deposition of immunoglobulins, probably this altered immunologic state favored the involvement of motor roots leading to increased protein in cerebrospinal fluid and motor impairment.

In our case, the patient presented a classic clinical picture of CIDP in the context of a reversal leprosy reaction, years after the end of treatment of a borderline leprosy disease. The patient demonstrated improvement of both conditions on neurological and nerve conduction grounds after steroid treatment.

The leprosy type 1 reaction damages the nerves most often in a mononeuritis pattern and there was no results founded describing CIDP related to leprosy reverse reaction in the literature. We postulate that this patient’s CIDP presentation could be attributed to cell injury caused by type I reaction that exposed the neural antigens and incited an autoimmune reaction.

References
5. Ridley DS, Jopling WH (1966) Classification of leprosy according to immunity. A five-group system. Int J Lepr Other


