

PURE NEURAL LEPROSY: IMPACT ON DAILY ACTIVITIES

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Abstract: Leprosy is a chronic, granulomatous, infectious disease caused by the acid-fast bacillus *Mycobacterium lepra*, also known as Hansen's bacillus. The nerves are affected in almost all forms of leprosy, and inflammatory infiltrates can be identified around the nerve fibers. There are cases in which there is purely neural involvement and the involvement of the ulnar nerve is one of the most frequent forms of the tuberculoid form of leprosy. The objective of the present study is to report the case of a patient with suspected pure neural involvement by Hansen's bacillus, in the city of Franca-SP.

Keywords: Leprosy, *Mycobacterium leprae*, Ulnar Neuropathies

INTRODUCTION

Leprosy is a chronic, granulomatous, infectious disease caused by the acid-fast bacillus *Mycobacterium lepra*, also known as Hansen's bacillus. This microorganism has tropism for both skin cells and peripheral nerves, and can also affect the eyes and internal organs. However, most of the impediments, deformities and disabilities that leprosy patients have to face are related to neuropathy (WORLD HEALTH ORGANIZATION, 2000).

Leprosy represents, in Brazil, a serious public health problem, being considered a disease of compulsory notification to the Epidemiological Surveillance. In the last 20 years, data show a reduction in the prevalence of the disease, this was due to the contribution of decentralization actions of control campaigns and also to the introduction of free multidrug therapy as a form of treatment. However, in our country, this drop did not impact the issue of transmissibility. In 2012, the case rate in Brazil was 1.51 per 10,000 inhabitants, a high value considering the Ministry of Health's goal of eradicating the disease by 2015. (MINISTÉRIO DA SAÚDE,

2017)

The disease can have an incubation period that varies from two to five years, the course of the disease occurs in an insidious way, however, it can cause a high level of disability. If not treated in the initial form, it evolves, becoming transmissible and can affect people of any sex and age. Hansen's disease is transmitted by the respiratory route when there is close and prolonged contact of a susceptible person with an infected and untreated individual. It is known that most of the population has immunity against the leprosy bacillus and that there is genetic susceptibility involved with the development of the disease. The most common is that this contagion occurs by proximity to a family member who is unaware of the infection. (MINISTRY OF HEALTH, 2008; MINISTRY OF HEALTH, 2009)

Leprosy can be classified in four ways: indeterminate (paucibacillary) leprosy, tuberculoid (paucibacillary) leprosy, borderline (multibacillary) leprosy and virchowian (multibacillary) leprosy. (MINISTRY OF HEALTH, 2017)

All individuals go through the indeterminate form at the beginning of the disease, in some it is noticeable and in others it is not. At this stage, it usually manifests as a single skin lesion, lighter than the surrounding skin, without relief and dry (due to local anhidrosis). There is loss of thermal and/or pain sensation at the site of injury, however, tactile sensitivity is usually preserved. Skin biopsy often does not confirm the diagnosis and smear microscopy is negative, due to the low number of bacilli present in this form of the disease. (MINISTRY OF HEALTH, 2017).

The tuberculoid form is the one in which the individual's immune system has the ability to destroy the bacilli spontaneously, with an average occupancy time of 5 years. It usually manifests as a totally anesthetic

plaque or by plaques with raised edges, well delimited with a clear center. It can also occur as a thickening of a single nerve (peripheral neuritis) with total loss of sensation in its area of innervation. This is because, when the intense reaction of the immune system against the bacillus occurs in the nerve trunk, the formation of the tuberculoid granuloma occurs, responsible for the neural injury. When this occurs, smear microscopy is negative and skin biopsy almost always does not show bacilli and, consequently, does not confirm the diagnosis. Therefore, subsidiary tests are rarely necessary, since there is a total loss of sensitivity, associated or not with changes in motor function, but in a localized way. (MINISTRY OF HEALTH, 2017)

The borderline form is the most common form of disease manifestation (70% of cases), it usually occurs after 10 years or more of incubation. It usually presents several reddish or whitish patches of skin, poorly defined, with raised borders or by multiple well-delimited lesions, but with a poorly defined external border. In these lesions, there is partial or total loss of sensitivity, with decreased sweating and vasoreflexia to histamine. Asymmetric involvement of peripheral nerves is common (more severe and diffuse neuropathy, combining high levels of cellular immunity with diffuse fibrosis, both with intense activity). Smear of the infiltrated border of the lesions is often positive, except in rare cases when the disease is confined to the nerves. (MINISTRY OF HEALTH, 2017)

Virchowian leprosy is the most contagious form of the disease. This patient does not have visible stains, the skin is red, dry, infiltrated, and has dilated pores ("orange peel" appearance), usually spares the scalp, armpits and the middle of the lumbar spine (warm areas). With evolution, hansenomas (dark, hardened and asymptomatic lumps) are common. In a more advanced stage, madarosis and loss

of eyelashes and other hairs, except on the scalp, may occur. Generalized anhidrosis may appear, but sweating is more intense in areas spared by the disease, such as the scalp and armpits. Complaints of cramps/tingling in the hands and feet are common, which may suggest a misdiagnosis of rheumatologic disease, since rheumatologic tests (ANA and RF), as well as the VDRL, are often positive. In the nerve trunks, millions of bacilli are scattered around each compartment and layer of the nerve initially without any apparent tissue reaction from the nerve. There is usually some swelling. But over time the proliferation of fibroblasts will create diffuse scarring in the nerve and ultimately complete fibrosis and loss of all nerve function. This process can take many years to complete and is seen in almost all peripheral nerves, from near the spinal cord to the periphery. Peripheral nerves and their superficial branches are symmetrically thickened. Therefore, it is important to look for changes in thermal, painful and tactile sensitivity in the area of innervation of these nerves, and in cold areas of the body, such as elbows, knees, buttocks and legs. In Virchowian leprosy, the diagnosis can be easily confirmed by smear microscopy of the earlobes and elbow. (MINISTRY OF HEALTH, 2017).

The nerves are affected in almost all forms of leprosy, and inflammatory infiltrates can be identified around the nerve fibers. There are cases in which there is purely neural involvement (pure neuritic leprosy) and the involvement of the ulnar nerve is one of the most frequent forms of the tuberculoid form of leprosy. In those cases where the involvement of this nerve is isolated, the patient does not present skin lesions, making the diagnosis difficult. The knowledge of this peculiarity allows the realization of an adequate therapeutic approach, with the attainment of good results and consequent reduction of the

risks of peripheral neurological deterioration. Clinically, this patient has an enlarged nerve, sometimes with drainage of necrotic nerve tissue through a fistula in the skin. It is important to remember that initially only a small part of the nerve is affected, either a fascicle or an area located in the epinerve. When the abscess grows it will compress the healthy fascicles and cause local ischemia and eventually spread to the other fascicles. (MINISTRY OF HEALTH, 2017)

Clinical evaluation is essential for the diagnosis of neuropathy. In history, sensory complaints are usually more frequent and early, and may manifest as pain or dysesthesia. Motor changes usually appear later. Neuropathy can progress insidiously, being slowly progressive, or it can be interspersed with episodes of acute worsening, resulting from leprosy reactions. The topography of the changes (intra-dermal neuropathy, mononeuropathy, multiple mononeuropathy, polyneuropathy) and the type of fibers involved are essential information for diagnosis and follow-up. The patient may complain of a change in sensation (numbness or tingling) on both sides of the little and ring fingers and in the middle of the palm and back of the hand below these fingers, which may be intermittent or constant. In addition, weakness and soreness in the hand and elbow area may occur. On physical examination, the patient with typical ulnar nerve injury presents with paralysis of the interosseous and lumbrical muscles of the fourth and fifth fingers. This establishes an imbalance of forces in the flexo-extensor apparatus of the fingers. Hyperextension of the proximal phalanx occurs and the deep flexors over-flex the distal phalanges, resulting in a claw hand. Atrophied first dorsal interosseous muscle (muscle between the bases of the thumb and index fingers, on the back of the hand) may also be present, presenting as a depression in the region. On palpation, the nerves are swollen,

especially when palpated in the cubital fossa. (MINISTRY OF HEALTH, 2010; CASTRO, 2012; QUEIROZ, 2015).

Thus, knowing the prevalence of leprosy and the implications of the limitations caused by this disease in daily life, the development of public policies to combat the disease is very important, since knowing the epidemiological data collected on a particular location, Through the notified cases, there are mechanisms that contribute to carrying out disease control programs, this way the chain of transmission can be broken, infected patients can be treated and clinical situations of physical disability can be prevented.

The objective of this article is to report the case of a patient with suspected pure neural involvement by Hansen's bacillus, in the city of Franca-SP, correlating the epidemiology and clinical manifestations found in the patient with those present in the literature.

CASE DESCRIPTION

The work in question is a case study carried out in February 2020, from a consultation at the Outpatient Management Center in the city of Franca-SP, with a male patient, 51 years old, black, worker in the footwear production.

In the anamnesis, he complained of weakness in the right upper limb and tingling in the territory of the right ulnar nerve, in addition to not being able to carry out an extension movement with the fourth and fifth fingers of the right hand, with an impact on the performance of daily activities. He reported that he had already been through other doctors, without a definitive diagnosis.

Palpation of the ulnar nerve showed very significant thickening of this nerve in the cubital region, and percussion showed pain along the path of the nerve (positive tine). The neurological examination showed a reduction in muscle strength in the abduction of the little finger, ulnar claw, thermo-painful anesthesia

in the territory of the cutaneous distribution of the nerve and neural thickening at the level of the elbow on the side of the lesion. Dermatological examination did not identify a skin lesion. Electroneuromyography showed a reduction in sensory and motor action potentials in the ulnar nerve.

Due to the local epidemiology, where there is a high prevalence of leprosy, Hansen's disease was raised as the main diagnostic hypothesis. As a conduct, we refer the patient to the Specialized Leprosy Service, for diagnostic confirmation and follow-up.

DISCUSSION

Since the dawn of civilizations, leprosy is reported as a disease that represents a paradigm in societies, also known as leprosy. Biblical accounts characterize lepers as individuals who lived on the margins of society because they were considered unclean. Even today, an important barrier to be overcome is that of prejudice, rooted in many people when it comes to leprosy.

Pure neural leprosy is a form of leprosy in which the patient has no dermatological manifestations, manifesting the disease as pure neurological impairment. It arises from the invasion of peripheral nerves by the bacillus and results from immunoinflammatory responses to the pathogen. Clinically, the patient presents signs and symptoms of neural impairment of sensitivity with localized paresthesia, hypoesthesia, thermal and painful anesthesia, pain on palpation and thickening of the affected nerve with evolution to motor alteration. The reported patient arrived at the service with the presence of pain on palpation and neural thickening, in addition to paresthesia of the limb innervated by the affected nerve, as previously described in the literature. (SKACEL, 2000)

The diagnosis in these cases is a challenge, since the standardized and unmistakable

picture of leprosy is not present and many of these patients are in the paucibacillary form, that is, the bacilli are not evidenced in smear microscopy because they have a low bacterial load. No electroneuromyographic finding is specific to leprosy neural injury, which reduces the importance of this test in the etiological diagnosis of isolated ulnar nerve palsy. On the other hand, the possibility that the biopsy of the dorsal cutaneous branch of the ulnar nerve demonstrates the bacillus, in addition to the nonspecific findings already mentioned, makes this test important in the diagnosis of forms of leprosy that progress with ulnar nerve injury, without affecting the skin. The case reported is illustrative of a rare type of leprosy involvement, but which must be considered during the evaluation of patients with increased nerve volume, even if isolated and without cutaneous manifestations. (CAMPOS, 2016)

Knowing that leprosy is a very prevalent disease in Brazil, especially in some regions, it is necessary to create more effective measures for the early detection of the disease. of involvement. In addition, it is important to alert the population to the signs of the disease.

FINAL CONSIDERATIONS

Considering the present report, in which the patient was previously treated by several professionals and none of them raised the suspicion of leprosy even knowing the prevailing scenario of the disease in the country.

Taking into account the biopsychosocial impact of the limitations caused by leprosy, especially with regard to neurological manifestations, it is extremely important that these patients receive individualized care, in order to minimize the damage of the disease. Therefore, multidrug therapy is efficient, not only in minimizing the effects of the disease, but also in preventing the contamination of

close contacts.

It is important to fight not only the spread of the disease, but also prejudice against patients, so that they can be treated and live normally in the community.

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