

Indeterminate leprosy and lepromatous index case: four cases in the same family*

Hanseníase indeterminada e caso índice virchowiano:
4 casos na mesma família

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Abstract: Leprosy is a chronic infectious disease caused by *Mycobacterium leprae*, which primarily affects the skin and peripheral nerves. Brazil remains as the country with the second largest number of cases in the world. We report the case of three patients diagnosed with indeterminate leprosy in the same family. Two patients were HIV positive. An active search led to the discovery of the index case. It was crucial to persist in the search of the index case. This report shows how important it is to teach physicians and the general population about the signs and symptoms of leprosy. Early diagnosis and treatment are necessary to prevent sequelae and to eliminate the disease as a public health problem.

Keywords: HIV; Leprosy; Leprosy, lepromatous; Leprosy, paucibacillary

Resumo: A hanseníase é uma doença infecciosa crônica causada pelo *Mycobacterium leprae*, que afeta principalmente a pele e nervos periféricos. O Brasil continua sendo o segundo país do mundo com maior número de casos. Aqui relatamos três pacientes diagnosticados com hanseníase indeterminada, pertencentes à mesma família, cuja busca ativa levou à descoberta do caso índice. Dois pacientes eram HIV positivos. Foi de imensa importância a insistência na procura pelo caso índice. Este relato ilustra a importância do ensino sobre sinais e sintomas de hanseníase na formação médica e à população. O pronto reconhecimento e tratamento são necessários para prevenir sequelas e eliminar a doença como problema de saúde pública.

Palavras-chave: Hanseníase; Hanseníase Paucibacilar; Hanseníase Virchowiana; HIV

INTRODUCTION

Leprosy is a chronic granulomatous infectious disease caused by the obligate intracellular bacillus *Mycobacterium leprae*. It primarily affects the skin and peripheral nerves.^{1,2} It is endemic in many regions of the world, and Brazil remains the country with the second largest number of cases.³ Over time, the manifestations of the disease cause deformities and mutilations, which are responsible for the stigma of the disease. The gram-positive, acid-fast bacillus⁴ was first described by GH Armauer Hansen in 1873.

We report the case of three patients diagnosed with Indeterminate Leprosy in the same family. An active search led to the discovery of the index case.

CASE REPORT

Patient A was 8 years old, female, and HIV +. She was using no medications and presented a single hypochromic lesion on the left forearm. Biopsy of the lesion was compatible with indeterminate leprosy (Figure 1). During the child's consultation, the mother (patient B), who was 34 years old, had been HIV + for 8 years and on antiretroviral therapy for 7, complained of a macule on the left arm which had appeared about 1 year earlier and which had started to show elevated borders 1 month before the consultation, with reduction of thermal sensitivity (Figure 2). On the next consultation, the mother brought another daughter (patient C), who was 19 years old and pre-

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sented a hypochromic macula with reduced thermal sensitivity in the lateral region of the right knee (Figure 3). Biopsies of lesions from patients B and C were also compatible with indeterminate leprosy. There was no thickening of nerves in patients A, B or C. The three patients were diagnosed with indeterminate leprosy, and paucibacillary-multidrug therapy was started. We did not perform the Mitsuda test because it was not available in our service.

Considering the 3 cases, we sought to identify a possible source of infection among close family members. The patients initially denied knowing about anyone close who had skin lesions. When we showed them a photo of a lepromatous patient in a book, the patients reported on a relative who presented similar lesions. Patient D, who is male, patient B's brother and patients A and C's uncle, was then brought by the family. He presented weakness and numbness in both hands, in addition to nasal obstruction. On examination, he showed nodules and tubercles on the face and pinnae, bilateral madarosis, diffuse infiltration in the trunk and lower limbs (Figure 4). There was cubital and auricular nerve thickening, which occurred bilaterally. There was hypoesthesia in bilateral legs and feet, hypoesthesia of the whole left arm and right arm (Figure 5). Skin smears were positive. Multibacillary multidrug therapy was initiated.

The four patients are from Laguna, Santa Catarina. Patients A, B and C live in the same house. Patient D lives close, on the same street, and often visits the other patients. Four people were considered close contacts of the patients. These contacts did not want to be examined in our service, so they were advised to seek examination in the health center of their city of origin. Patients A and B completed treatment and were discharged as cured, presenting the characteristics they showed on the first physical examination. Patients C and D received 5 doses and asked to finish the treatment in their hometown. The cases were then transferred.



FIGURE 1: Patient A: HIV-positive, eight year-old girl presenting a hypochromic macula on the left arm



FIGURE 2: Patient B: HIV-positive, 34 year-old woman, patient A's mother, presenting hypochromic macula on the left arm



FIGURE 3: Patient C: HIV-negative, 19-year-old girl, patient A's sister, presenting hypochromic macula on the left thigh



FIGURE 4: Patient D (index case): HIV-negative, 25-year-old man, patients A and C's uncle and patient B's brother, presenting nodules and tubercles, madarosis, and diffuse infiltration in the face



FIGURE 5: Patient D (index case): 25 years old, presenting thickening of the left auricular nerve and nodules and infiltration in the earlobe

DISCUSSION

The transmission of leprosy occurs from individual to individual, between lepromatous and borderline patients and healthy individuals. Higher risks are associated with living in the same place as an untreated patient positive for acid-fast bacilli.¹⁴ The routes of elimination of the microorganism are mainly the upper airway and lesioned areas of the skin.⁴ The bacillus is highly infectious, but presents low pathogenicity and virulence, that is, many people are infected, but few get sick.⁴ The rate at which leprosy spreads depends on the susceptibility of the individual and the opportunity of contact with the microorganism.⁵ The host's innate response is what determines the clinical manifestation of the disease.¹ Contact with infected patients does not always result in transmission of the disease, and different individuals exposed to the same infected case develop different clinical forms.⁵

The World Health Organization classifies patients into paucibacillary, if they have five or fewer skin lesions, and multibacillary, if they have six or more skin lesions or if the skin smear is positive. The Ridley-Jopling classification subdivides leprosy into the following clinical forms: indeterminate, tuberculoid, borderline-tuberculoid, borderline-borderline, borderline-lepromatous and lepromatous.²⁶ In rela-

tion to risk factors for leprosy, close contacts of an infected individual have a greater chance of acquiring the disease in comparison with the general population.⁷ In 2006, Moet *et al.* observed in their work that individuals in contact with leprosy patients aged 5-9 years and older than 30 have a higher risk of acquiring the disease. Those aged between 20-29 years are less likely to acquire it.⁷ Also, those in contact with multibacillary patients are at greater risk of developing the disease. The risk should be higher among relatives, even when they are not close contacts, due to genetic influences.^{7,8}

There is a correlation between cases of multibacillary leprosy in children, the endemic situation of the country, and late diagnosis. The disease is mainly transmitted to children through close family contacts. The younger the child, the greater the number of infected individuals found among the family members.⁹ Barbieri's study shows that the most affected age group among children is between 10 and 15 years, with prevalence increasing as the age increases.⁹

Every hypochromic lesion in children living in an endemic area should raise the suspicion of leprosy.⁹ Most children have lesions on exposed areas such as the trunk and limbs. In Brazil, the high prevalence of leprosy leads children to have contact with the disease. In case of suspicion, a thorough medical history should be done, with an investigation of the clinical and epidemiological history, as well as a detailed dermatoneurological examination.⁹

Regarding a leprosy and HIV coinfection, the difficulty in conducting prospective studies due to the long incubation period of *M. leprae* makes it impossible to confirm whether HIV-positive patients are more likely to develop leprosy than HIV-negative patients.¹⁰ The clinical spectrum seems to remain unchanged, as well as the adverse effects of multidrug therapy.⁴

As demonstrated by the cases here presented, persisting in the search for the index case was of paramount importance. They show how important it is to teach physicians and the general population about the signs and symptoms of leprosy. Prompt recognition and treatment are necessary to prevent sequelae and eliminate the disease as a public health problem. □

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