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Multibacillary Leprosy With Tumor Like Nasal Deformity

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Abstract

BACKGROUND: Leprosy is a chronic granulomatous infection caused by *Mycobacterium leprae* which affects skin, peripheral nerves, and other body parts including bone, ear, eye, and nose. Nasal manifestation of leprosy may be found in the early stage with non-specific symptoms such as nasal congestion. In advanced stage, destruction of bone structures and cartilages may result in deformities. However, nasal manifestation as the only symptom in early stage can lead to delayed diagnosis.

CASE REPORT: We reported a case of multibacillary leprosy in a 26-year-old male with tumor like nasal deformity for 1 year and lump in the ear lobes since 8 years ago. Initially, there was nasal discharge, unfrequent epistaxis, followed by loss of smelling perception, without associated pain, or numbness since childhood. Lesions on the trunk, palms, and soles were also noted since 2 years ago. The physical examination revealed tumor with crusts and fissures on the nasal tip; accompanied by saddle nose. Slit skin smear revealed acid-fast bacilli with bacteriological index of 4+ in solid form. Hence, he was diagnosed as multibacillary leprosy with Grade 2 disability. The prognosis was poor due to permanent deformities.

CONCLUSION: Early diagnosis and initiation of treatment are crucial in the management of this debilitating and stigmatized disease.

Introduction

Leprosy is a chronic granulomatous infection caused by *Mycobacterium leprae* that affects skin, peripheral nerves, and other body parts such as bone, ear, eye, and nose [1]. In Indonesia, it was reported in 2015 that the prevalence of leprosy was 2.9 out of 10,000 persons. Indonesia rank as the third highest country with new cases of leprosy in the world, after India and Brazil [2]. *M. leprae* is an obligate intracellular acid-fast bacilli, with the length of 1.5–8 μm and diameter of 0.2–0.5 μm. This bacterium lives and proliferates at temperature of 27°C up to 30°C. The incubation period of *M. leprae* varies between several weeks up to 20 years with mean duration of 5–7 years. Transmission of the bacilli occurs in person with close and long contact through bacilli inhalation into nasal area [3], [4]. Ridley-Jopling classification divides leprosy into six spectrums: Tuberculoid polar (TT), borderline tuberculoid (BT), borderline (BB), borderline lepromatous (BL), lepromatosa subpolar (LLs), and lepromatosa polar (LLp) [1]. The lepromatosa spectrum is often presented as manifestations in ear and nose. Early nasal manifestation of leprosy may present with non-specific symptoms such as nasal congestion. However, in advanced stage, destruction of bone structures and cartilages of the nose may take place and followed by nasal deformities [5]. We report a case of leprosy in an adult male with tumor like nasal deformity.

Case

A 26-year-old male consulted to the Department of Dermatology and Venereology at Universitas Sumatera Utara General Hospital, with the chief complaint of mass in the nose since 1 year before consult and lump in the ear lobes since 8 years ago. The patient also complained of non-itchy erythematous patches in the feet, which gradually thickened since 13 years ago. The patient had consulted to general physician and was prescribed unrecalled ointment, without any complete resolution of the symptoms. The lump in the ear presented initially as spots in both earlobes without any pain or itchiness. It was diagnosed as keloid at that time and steroid injection was administered. Since 2 years before consult, the patient was noted to have reddish papules in the body, arm, and legs; which gradually enlarged without any numbness or tingling sensation. The mass in the nose presented initially as red spot on the nose; which gradually enlarged and easily bled. The patient also complained of clear nasal

discharge from the nose, sometimes epistaxis, followed by loss of smelling perception, without associated pain, or numbness. He then noted the appearance of wounds the in hand and feet since 1 month ago. The patient consulted to the ENT departments due to mass in the nose; and then referred to our department of dermatology and venereology. There was no history of fever, joints pain, chronic cough, and weight loss. History of sexual contact and other family member with same symptoms were denied. In the physical examination, the vital signs were within normal limits and nutritional status was normal. Dermatology examination revealed that multiple erythematous to skin-colored nodules were noted on the face and bilateral auricle lobes. There was tumor with crusts and fissures on the nasal tip; accompanied by saddle nose. There were multiple erythematous to hyperpigmented macules, papules, and plaques with xerosis and scales on the sternal, vertebra regions, and bilateral lower arms. On bilateral dorsal hands, there were erythematous plaques with fissures in the fifth fingers and dactylitis in the fourth and fifth fingers. Stiff contractures were found in the fingers of both feet. Bilateral ulnar nerves, bilateral common peroneal nerves, and left great auricular nerve enlargement were found. Sensibility test revealed anesthesia to temperature sensation and hypoesthesia to touch and pain stimulus on the lesions and distribution area of ulnar nerve, median nerve, and common peroneal nerve. Motoric examination revealed Grade 3 weakness in abduction movement of both fifth fingers. The patient was also noted to have decrease of sweat production in the area with skin lesions during activities and area with xerosis.

Differential diagnosis of the patient is multibacillary leprosy, rhinophyma, and tertiary syphilis. Slit skin smear then was performed on both auricle lobes and nasal tip; Ziehl–Neelsen staining revealed acid-fast bacilli with bacteriological Index 4+ in solid form. Serologic examinations of VDRL and TPHA revealed non-reactive results. Hence, the patient was diagnosed as multibacillary leprosy with Grade 2 disability and neuropathic ulcers. He was prescribed with multidrug therapy (MDT), consisted of rifampicin 600 mg, dapsone 100 mg, and clofazimine 300 mg on day 1; followed by dapsone 100 mg and clofazimine 50 mg on day 2–day 28. Vitamins B1, B6, and B12 were given as adjuvant therapy. The wound care was done with wound dressing with NaCl 0.9%, fusidic acid 2% ointment twice a day, and emollient acrylic stearate after bathing. The patient was advised to consume nutritious diet, rest well, and compliance to medications and has regular follow-up every month. He was also advised to use protections while working and always use shoe to prevent new wounds from trauma. This patient has poor prognosis due to the disabilities that have manifested.

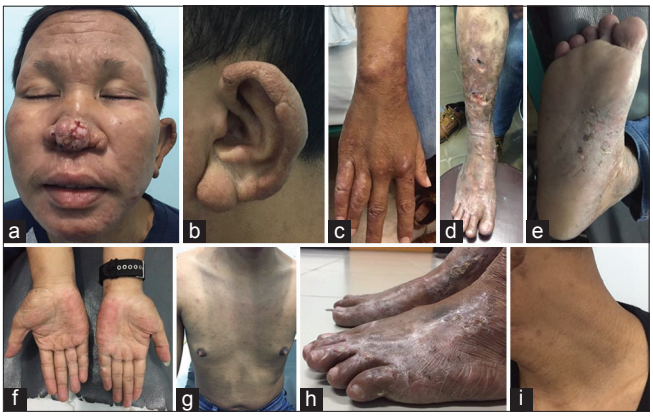

Figure 1: Initial dermatologic examination revealed multiple erythematous to skin-colored nodules on the (a) face with crusted tumor on the nasal tip and saddle nose, (b) ear, (c) dactylitis of 4th and 5th right fingers, multiple erythematous to hyperpigmented macules, papules and plaques with xerosis, scales and some with ulcers on the (d) right lower leg, (e) soles, (f) palms, (g) trunk, (h) with contracture of toes, and (i) enlargement of left great auricular nerve

Discussion

Leprosy or Morbus Hansen is chronic granulomatous infection caused by *M. leprae* with the predilection site on the skin and peripheral nerves. The risk factors for leprosy are followings: Born or living in endemic area, other family member with leprosy, genetic susceptibility, environment exposure, poverty, and other diseases which suppress immune response. This disease is very contagious, but the morbidity is low due to natural immunity in most population with normal immune response [1]. The clinical manifestations of leprosy are variable. The patient can complain of skin lesions (white or reddish patch), numbness in the hand and or feet, weakness in the arm and/or leg, joint pain, nasal congestion, epistaxis, photophobia, and anesthesia on the face and ears. The thickening of peripheral nerves can be found too [1]. Based on the WHO Expert Committee on Leprosy, leprosy is diagnosed if there is at least one of the followings: Definite loss of sensation in a pale (hypopigmented) or reddish skin patch; a thickened or enlarged peripheral nerve, with loss of sensation and/or weakness of the muscles supplied by that nerve; and the presence of acid-fast bacilli in a slit-skin smear [6]. Multibacillary leprosy presents as more than 5 skin lesions with hypoestesia or anesthesia, symmetrical nerve thickening and nerve function deficits, madarosis, leonine facies, and deformities in advanced stage, such as saddle nose, fingers contractures, and ulcers on extremities [1], [7]. In this case, the patient had clinical symptoms of multibacillary leprosy and acid-fast-bacilli with bacteriological Index 4 + in slit skin smear. In some cases, although rare, there is manifestation of dactylitis due to fusiform swelling of fingers, as presented in our patient [8]. Besides, *M. leprae* has the tendency to grow in body regions with temperature < 37°C, such as scalp, axilla, genitalia, and lumbosacral region. Those areas are seldom involved, so-called “immune zones.” Some

literatures reported that palmar and plantar regions are rarely involved due to localized bacilli growth in area with thick epidermal and fat layers as they provide good media of heat insulation. However, it should be emphasized that in fact the bacilli can grow in any skin surface [9]. As shown in this case, the lesions were present in the palm and plantar area of the patient.

Ear, nose, and throat examinations are mandatory in the screening of leprosy, due to the involvement of respiratory tract and ear in this disease [5], [10]. The main transmission route in leprosy is through droplets; which can be transmitted during coughing, speaking, or sneezing. The anatomical structures of nose with turbinates are susceptible for entrapment of bacilli in the nose. Besides, abundance of Schwann cells in the nose is the predilection for bacilli growth in this area [11]. Nasal symptoms can precede skin lesions in leprosy for years, depend on the immunity status of the patient, with symptoms such as bloody post nasal discharge, epistaxis, nasal congestion, and hyposmia [12]. Manifestations on nasal and ears area are often found in the lepromatosa leprosy. The clinical presentations are pale-yellowish mucous membrane with nodule or plaque in the anterior tip of the inferior turbinates as the predilection site. The lesion will enlarge and progress to cartilages and bone structures destruction in advanced stage, leading to septal perforation, and saddle nose formation [5]. Localized lesion on the external nose can be found, although rare, in the tuberculoid leprosy. The intranasal manifestations of leprosy are divided into three phase: Initial phase, intermediate phase, and advanced phase. The initial phase is marked by the mucous membrane involvement, with thickening, infiltration, and pale-yellow color that grow into nodule. Dry mucous membrane is caused by disturbance of mucous gland production due to parasympathetic nerve dysfunction. Intermediate phase is marked by nasal obstruction, due to an enlargement of lepromatous infiltration and progressive mucous membrane thickening. The congestion is resistant to any vasoconstrictor medications. The drainage of the mucus will be compromised and increase the risk of repetitive trauma and ulcus formation, as the patients sneeze frequently to clear the nose. Advanced stage is marked by ulceration, secondary infection of the nodule, and lepromatous infiltration. The absence of blood flow to perichondrium causes the septal perforation of nasal cartilages. The reabsorption and perforation process continue until cartilages and bone structures damaged completely; gradually become atrophy tissue which damage turbinates and endonasal bone structures [10].

The patient had symptoms of frequent sneezing and congested nose since childhood, which can be the early manifestations of leprosy. During adolescent, there were lesions on both earlobes, which were misdiagnosed as keloid. The patient just experienced the symptoms of nasal obstruction, blood-tinged nasal discharge, voice change, and smelling disturbance on his early adult age. On the physical examination, the patient had nodule infiltrates in the septum and turbinates with dry mucous membrane, accompanied by crusting, ulceration, and septal perforation. The nasal manifestations in this patient were in the advanced stage and the deformities were permanent. Rhinophyma is the variant of rosacea, which marked by the nodules on the nose with rough surface, skin thickening, and skin follicles enlargement. This condition is found only in male [13]. Tertiary syphilis is marked by destructive granulomatous lesion affecting the nasal area, and described as the great imitator [14]. In this case, the patient did not have history of sexual contact and lesions in the genital area. The negative serologic examination of VDRL and TPHA ruled out the diagnosis of syphilis. On both conditions above, there should be absence of cardinal signs of leprosy (skin lesions with hypoesthesia or anesthesia, peripheral nerve thickening or dysfunctions, and positive acid-fast bacilli in the slit skin smear) in contrast to the findings in our patient. The patient received multi-drug therapy as recommended by the WHO.

Leprosy patients often suffer from dry skin due to autonomic nerve dysfunction, so the patient was given emollient. Based on the WHO classification, this patient was classified to have disability Grade 2 due to nasal deformities and finger contractures on hand and feet [2]. The prognosis of the patient was poor due to delayed diagnosis and treatments were given after the deformities had emerged. However, compliance to therapy and avoidance of new wounds are mandatory to prevent the progression of disease and the occurrence of another disability.

Conclusion

Leprosy is still a global health burden with high numbers of disability manifestations. As the great imitator, rare clinical manifestations in leprosy such as nasal tumor, skin lesions on palm and sole, and dactylitis can lead to delayed and or misdiagnosis in leprosy patients. However, the physician should be aware of leprosy diagnosis if there is finding of at least one out of three cardinals' signs in the leprosy.

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