A Woman with Nasal Mass and Epistaxis: An Unusual Presentation of Leprosy

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Abstract- Leprosy is an infectious and non-fatal disease that mainly involved skin and peripheral nervous system and caused by *Mycobacterium Lepra*. This case is a 41-year-old woman with intermittent nasal bleeding and a mass (1.5×2.5 cm) in the right side of lateral nasal wall and a (1×1 cm) mass on septum in the left side of the nose. Considering the probability of intranasal tumor, endoscopic biopsy for histopathologic examination had been done and the result confirmed the diagnosis of leprosy. After unexpected proving of pathologic lepreleprom atose, the patient put under 3-drug- treatment.

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Key words: Leprosy; bacillus; nose

Introduction

Leprosy is a chronic and non-fatal disease that caused by *Mycobacterium Lepra*. Its clinical manifestation is largely confined to the skin, peripheral nervous system, upper respiratory tract, eyes and testes.

The clinical manifestation of leper is mainly due to immune mediated peripheral nervous system damage. If the patient is untreated, deformities can cause sociable violent stigma for the patient .With early diagnosis and the considering suitable and effective antimicrobial therapy, patients can have usual life in the community.

In this report, an interesting case of leprosy is introduced only with intranasal masses.

Case Report

The patient was a 41-year-old house wife and from Lahijan (guilan province- Iran). One month prior of admission she had been suffering from intermittent nasal bleeding especially the right side. The patient had not nasal obstruction, anosmia and rhinorea. She did not mention any important disease from the past and did not use especial drug as well. She also did not express any contact with a person who suffered from important infectious disease.

In intranasal examination (Figure 1), was observed a purple lobular soft mass that was about 1.5×2.5 cm in lateral wall, alongside the inferior conchea and a smaller mass that was about 1×1 cm on the left side of septum .Then for studying the mass spreading, CT scan with contrast was requested that confirmed restriction of the damage to interior of the nose. Any bone lesion was not obvious (Figure 2). By considering probability of granulomatous tumoral lesions, nasal endoscopy and biopsy was done, and tissue samples were sent for histopathologic examination.

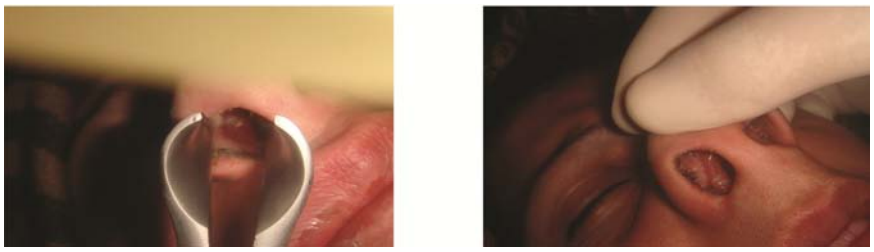

Figure 1. Physical examination: anterior rhinoscopy of the patient reveals a purple lobular soft mass alongside floor of the right nasal passage adhere the septum

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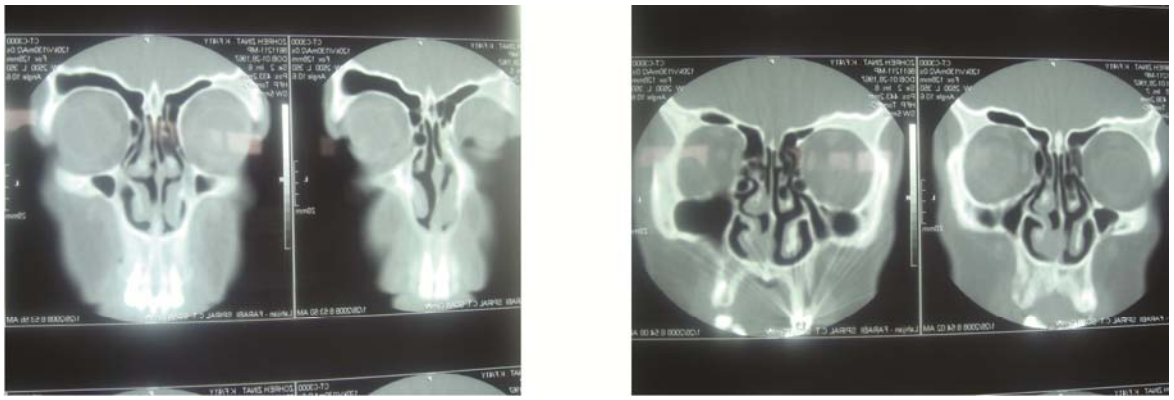

Figure 2. CT scan of paranasal sinuses with contrast: restriction of the damage to anterior nose with no bone erosion

Pathologist report based on especial staining methods was a granolumatose inflammatory process with a great number of bacilli and considered to lepromatose.

Lepromatous leprosy, in the usual macular or infiltrative-nodula lesions,exhibits an extensive cellular infiltrate that is almost invariably separated from the flattened epidermis by narrow gram zone of collagen.Infiltration causes the destruction of the cutaneous appendages and extends into the subcutaneous fat.In florid early lesions, the macrophages have abundant eosinophilic cytoplasm and contain a mixed population of solid and fragmented bacilli (BI=4 or 5). Bacilli are commonly observed also in endothelial cells. There is no macrophage activation to form epithelioid cell granulomatose. Lymphocyte infiltration is not prominent, but may be many plasma cells (1).

After unexpected pathology result, more studies were conducted. The patient did not have any familial background of the disease and did not mention any contact with any patient who suffered from leper. She mentioned sensory loss in feet from the past few months that confirmed in examination. There was no obvious nerve bundle involvement or enlargement. There were some pinky patchy lesions on the back of the patient that was negative in biopsy for leper. Some biopsies were taken from auricle, eyebrow and finger. Finger was not involved, however auricle contains 30% free bacilli, 50% fragmented bacilli and 20% dead bacilli. In eyebrow region 20% free bacilli, 60% fragmented bacilli and 20% dead bacilli were observed in microscopic staining.

The patient with leper diagnosis put under 3-drug-treatment with Rifampin, Dapsone and Clofazimine.

Discussion

Leper is an ancient word that used to recognize patients who were infected with *M. lepra* and severely deformed as a sequence of the infection.

Peripheral nerve damage is the most common complication of leprosy. Recognition and rapid treatment can prevent obvious disability. *M. lepra* was the first bacterium recognized in relation to illness. Armauer Hansen, a Norwegian scientist, discovered the microbe in 1874 exactly before discovering *M. tuberculosis* by Koch, however he could not cultured it. This problem has not solved yet.

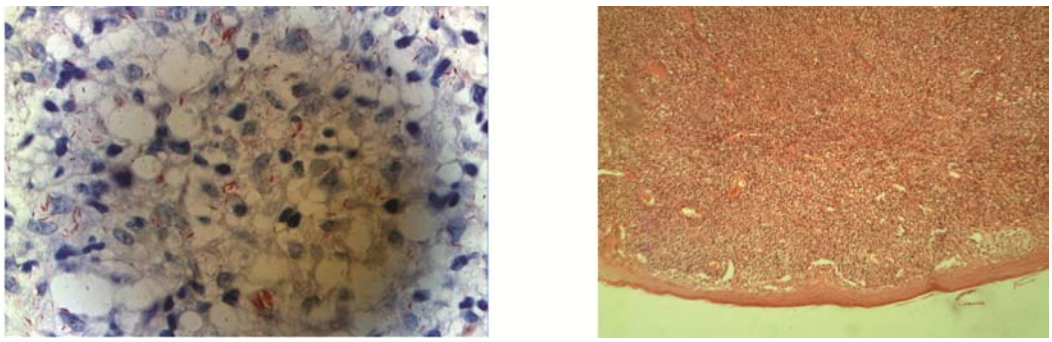

Figure 3. Acid fast staining and H&E Staining: a granolumatose inflammatory process with a great number of bacilli like lepromatose

In spite of improving treatments that has attained nowadays, this illness in some countries such as Brazil and India, has high prevalence (2-3); and also, it can be observed in developed countries like England too (4).

M. lepra is an obligatory internal cell bacillus (0.3 to 1 micron width and 1 to 8 micron lengths) that is acid fast and via microscopic can be distinguished from other mycobacteria. The primary diagnosis of disease is made by history taking and physical examination and confirmed by cutaneous smear and biopsy of lesions (2,3,5). This microbe is detected obviously in tissue sections by modified staining of Fite methods. *M. lepra* adapts well to penetrate and reside within macrophage. However it can be alive outside the body for 7-10 days. (6). Morphologic index or the number of acid fast bacilli in skin samples-that are colorized monotonously- is related to viability of bacilli. Newer recognition methods include finding antibody in glycopeptides fenolic (PGL.1) and reinforcement of M.lepra gene sequence with PCR (6). Bactriologic index or the rate of logarithmic increasing of *M. Lepra* density in derm can be about +4 and +6 in untreated patients that decrease one unit /year during effective treatment (7). Increasing bacteriologic index or morphology, means recurrence and if the patients are being treated,the infection may be resistance toantimicrobials. The standard enumeration of leprosy bacilli in lesions- the bacterial index (BI)- follows Raidly's logarithmic scale (which applies to both skin biopsies and slit skin smears) (1).

1- 1-10 bacilli per 10-100 HPF
2- 1-10 bacilli per 1-10 HPF
3- 1-10 bacilli per HPF
4- 10-100 bacilli per HPF
5- 100-1000 bacilli per HPF
6- More than 1000 bacilli per HPF

M.lepra genome is only containing 3 million double base that is two third of *M. tuberculosis* genome. Bacterial cell wall, has a peptidoglycon skeleton that is linked to arabinoglycan and micolic acid (6). External capsule contains a lot of pictures of *M. lepra* especial phenolic glycolipid that is recognized in serologic tests. *M. lepra* grows up perfectly in cooler tissues (the skin, peripheral nerves, anterior champer of the eye and testes), and usually doesn't involve warmer areas of the skin (the axilla, grain, scalp and midline of the back) (6). Transmission of illness is usually done from the nasorespiratory route, but sometimes infectious transmission can be from transplacental route (8).

The incubation period before appearing of clinical disease is variable from 2 to 40 years, although it usually takes 5-7 years (6). Leprosy according to Ridley-Jopling classification is divided into 5 groups which are based primarily on histologic changes. These groups contain: TT (tuberculoid), BT (borderline tuberculoid), BB (borderline), BL(borderline lepromatous) and LL (lepromatous). In this classification, epithelioid cells and lymphocytes in tuberculoid pole go into macrophage interior that appear totally foamy at lepromatose pole (9,10). The main manifestations of leprosy are,infiltrative skin lesions, hypoesthesia and peripheral neuropathy and these manifestations are closely

related to polarity of illness. Many patients with borderline tuberculoid or stable tuberculoid may present with skin lesions, but do not have other symptoms.On the other hand, those patients with developing tuberculoid leprosy may present with peripheral neuropathy, that is usually asymmetric (11). Patients with stable L. leprosy or borderline lepromatous may have with widespread infiltrative skin lesions or severe and obvious peripheral neuropathies with secondary deformities (like Claw Hand) or nonhealing painful ulcers (11). There are 3 clinical types: macular, infiltrative nodular and diffuse (1). Lepromatous form initially has cutaneous and mucosal lesions and neural changing occurs later. Lesions are usually numerous and symmetric. In macular type, a lot of hypopigmented or erythematous macules with unclear margin, are observed (1). The infiltrative-nodular type that is the classical and most common variety, may develop from macular type or arise independently. It is characterized by papules, nodules and diffuse infiltrates that are often dull red (1).

In diffuse type of leprosy, called Lucio leprosy, the patient shows diffuse infiltration of the skin without nodules (1).

It is noteworthy that *M. lepra* usually attack intranasal structures without any relationship to clinical form of disease and this can be happened even before skin lesions or lesions in other parts of the body. There are some cases of nasal leprosy in the literature without having any symptom in other parts of the body (12).

Since 1995, WHO founded free multi-drug therapy(MOT) for all patients around the world (13). Effective antimicrobial agents include dapsone, rifampin and clofazimine. Minocycline is effective especially for those patients who can not tolerate those 3 drugs or have allergic reaction to them (11). With this treatment, patients with fewer bacilli are treated in 6 months and patients with more bacilli are treated in 12 months (13). In 1991, WHO in official deceleration set a goal to omit leprosy as a general problem until 2000 that means the prevalence of the disease reach to less than one person in 1000 persons (13).

It seems that nasal mucosa is attacked by *M. lepra* and this can be the main way of entrance of infection. Recent studies confirm wide spreading of *M. lepra* DNA in healthy individuals (14,15).

In 95% of patients who suffer from lepromatous, nasal mucosa is involved (7,14,15). In tuberculoid type only those that have external nasal lesions-especially during active phase- show spreading of infection to nasal mucosa (7,14-16).

Because of involving nasal vascular network by leprosy, epistaxis can be the first manifestation of illness. This problem might be happened because of nasal obstruction, thick secretions, ulcers and also septum perforation (7,14-17). Anosmia and hyposmia might be occurred because of nasal obstruction due to infiltration or crust or involvement of receptors of olfactory mucosa (7,14-18). Another symptom is continuous rhinorea that is due to incorrect renervation (7,14-19).

It is interesting that despite high prevalence of infiltrative nasal mucosa by bacteria, occurrence of leprosy, with intranasal manifestations as primary symptoms of disease is so rare and usually a group of symptoms and related signs are seen especially in skin and peripheral nervous system. By a thorough review of literature, there were few reports of primary manifestations leprosy with nasal symptoms. For example S.Yasushi *et al.* (2002) reported a leprosy case with a submucosal lesion (1.5 cm) on nasal mucosa in a 58 year-old-brazil man (20) or in 2005,Kim Y.R and colleagues reported a leprosy case who was a 75-year-old Korean man suffering from leper lepromatous that had chronic nasal obstruction with epistaxis. Intranasal examination showed lots of crusts on inferior conchea and air passage narrowing (21,22). However another study revealed two intranasal lobulated masses which occurred during one month. It was a rare manifestation of leprosy with extensive symptoms and required more care to recognize in spite of rare and unusual manifestations.

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