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MUZZLE AREA INVOLVEMENT – AN UNUSUAL PRESENTATION IN LEPROSY

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ABSTRACT: Lepromatous leprosy is a chronic infectious disease caused by Mycobacterium leprae, characterized by the extensive involvement of skin and peripheral nerves. While it commonly affects the cooler regions of the body, such as the extremities, ears, and face, involvement of the muzzle area is exceedingly rare. We present a unique case of lepromatous leprosy in a 42-year-old male, where the initial manifestation of the disease primarily occurred in the muzzle area. This case report highlights the atypical clinical presentation of lepromatous leprosy, which typically spares the central face and mucous membranes. The patient exhibited disfiguring nodules, erythema, and sensory loss in the muzzle area, leading to a delayed diagnosis due to the unusual site of involvement. A comprehensive clinical evaluation, histopathological examination, and microbiological confirmation were pivotal in establishing the diagnosis. The rarity of muzzle area involvement in lepromatous leprosy underscores the importance of maintaining a high index of suspicion for atypical presentations, especially in regions where leprosy is endemic. Early detection and multidrug therapy remain crucial in preventing deformities and minimizing the spread of the disease. Furthermore, this case serves as a reminder for healthcare professionals to consider leprosy in the differential diagnosis of unusual facial lesions, even in non-endemic regions, as prompt diagnosis and treatment are essential for patient well-being and public health.

INTRODUCTION: The part of the face known as the "muzzle area," which corresponds to the primate's perioral area and the lips on both sides of the cheeks, the face's muzzle area. In the past, only one instance of post-kala-azar dermal leishmaniasis (PKDL), which is characterized by lesions around the lips and perioral area that resemble the monkey muzzle, has been described as involved the muzzle area [1].

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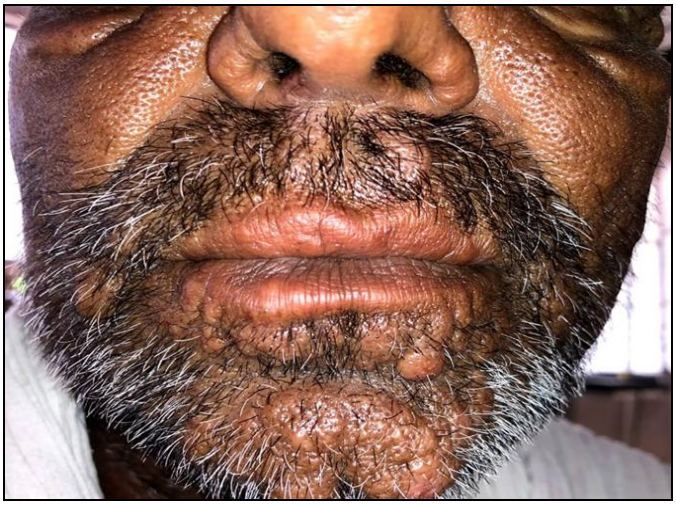
Muzzle area involvement in leprosy has only been documented in sooty mangabey monkeys (Cercocebusatys), which were found to have naturally contracted the disease [2]. Since, there has been only one case report recorded before, our case may help in contributing to the literature of involvement of the muzzle area of the face in lepromatous leprosy.

Here, we discuss an intriguing case with a patient who had lepromatous leprosy and comparable involvement.

Case Report: A 51-year-old male patient was seen with a history of skin-colored elevated lesions that began above the lower lip and spread slowly over time. They gradually expanded to include the top of the lip, chin, and both sides of the cheeks. These

lesions steadily spread over the course of a year, eventually affecting both sides of the face, trunk, and limbs. The patient complained of difficulty in breathing since past 2 months that aggravates on lying down. He described his history of nasal congestion, which was connected to his voice change and epistaxis. There was no past that could have indicated a sensory or motor nerve issue. Lethargy, weight loss, generalized malaise, coughing, chest pain, or palpitations were not present in the past. There was no prior history of nose or lip hardening, nor was there any history of red-colored elevated facial lesions linked to flushing from consuming hot, spicy food. The patient denied having ever had ocular redness, a feeling of a foreign body, photophobia, or blurred vision. The patient had no recent travel history to either Bihar or Jharkhand. Inspecting the lips revealed numerous skin-colored to erythematous, smooth, shiny distinct papules that ranged in size from 2 to 5 millimeters. These papules mostly affected the top lip's vermilion border. Over the face, trunk, buttocks, and both limbs in an even distribution, numerous skin-colored smooth, shiny papules to nodules as well as a few infiltrative plaques of varied sizes, ranging from 0.5 to 3 centimeters, were visible. Additionally seen were nodular ear lobe infiltration and bulbous nose growth **Fig. 1-3.**



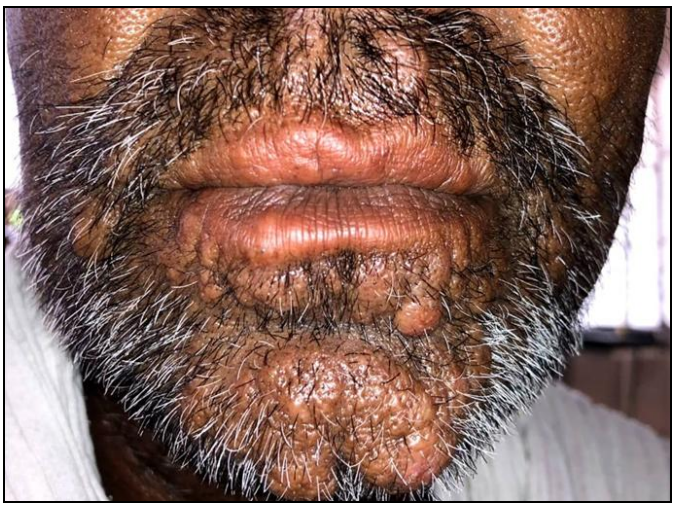


FIG. 1, 2, 3: NUMEROUS SKIN-COLORED SMOOTH, SHINY PAPULES TO NODULES WITH FEW INFILTRATIVE PLAQUES OF VARIED SIZES, RANGING FROM 0.5 TO 3 CENTIMETERS OVER THE FACE, TRUNK, BUTTOCKS, AND BOTH LIMBS IN AN EVEN DISTRIBUTION SEEN. ADDITIONALLY SEEN WERE NODULAR EAR LOBE INFILTRATION AND BULBOUS NOSE GROWTH

All of the peripheral nerves were bilaterally thickened, nontender, firm in consistency, and nodularity-free upon inspection. Temperature, discomfort, and touch perceptions were all present upon sensory assessment. According to the medical research council's (MRC)

assessment of voluntary muscle testing (VMT), there was normal power. The corneal reflex, light reflex, biceps, triceps, brachioradialis, knee jerk, and ankle jerk were all fully functional. The differential diagnoses of lepromatous leprosy, sarcoidosis, post-kala-azar cutaneous leishmaniasis, granulomatous rosacea, and the tumour stage of mycosis fungoides were taken into consideration using the aforementioned characteristics. Six sites were used for slit skin smears. Giemsa and Ziehl-Neelsen (ZN) stains were used to stain the smears. ZN staining revealed a large number of solid-staining, acid-fast bacilli with a morphological index (MI) of 80% and a bacteriological index (BI) of 6+ **Fig. 4**. Giemsa staining did not reveal the presence of Leishmania Donovan (LD) bodies. An acellulargrenz zone in the subepithelium and atrophy of the epidermis were found during a skin biopsy. There were few lymphocytes and sheets of foamy histiocytes in the dermis that extended to the deep dermis **Fig. 5**. Fite Faraco staining of the tissue section was performed to look for acid-fast bacilli. The results showed many pink bacilli that were solidly stained on a blue background **Fig. 6**. No well-formed naked granulomas were present, and the reticulin stain special staining produced negative results. Angiotensin converting enzyme and serum calcium levels were unremarkable. An X-ray of the chest revealed no anomalies.

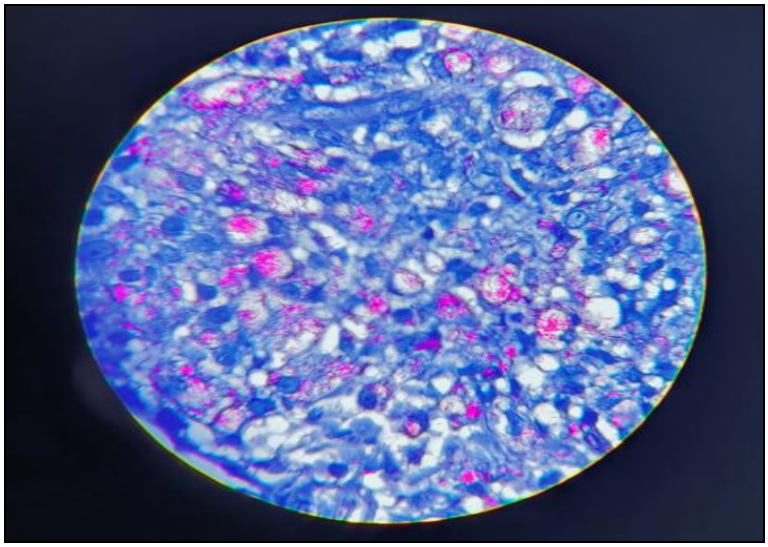


FIG. 4: ONGIEMSA AND ZIEHL-NEELSEN (ZN) STAIN- LARGE NUMBER OF SOLID-STAINING, ACID-FAST BACILLI WITH A MORPHOLOGICAL INDEX (MI) OF 80% AND A BACTERIOLOGICAL INDEX (BI) OF 6+ SEEN

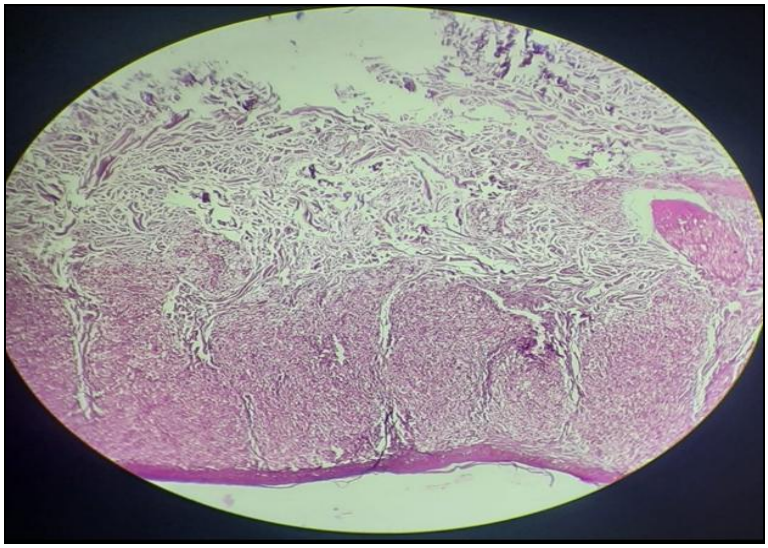


FIG. 5: ON HPE: AN ACELLULARGRENZ ZONE IN THE SUBEPITHELIUM AND ATROPHY OF THE EPIDERMIS WITH FEW LYMPHOCYTES AND SHEETS OF FOAMY HISTIOCYTES IN THE DERMIS THAT EXTENDED TO THE DEEP DERMIS SEEN

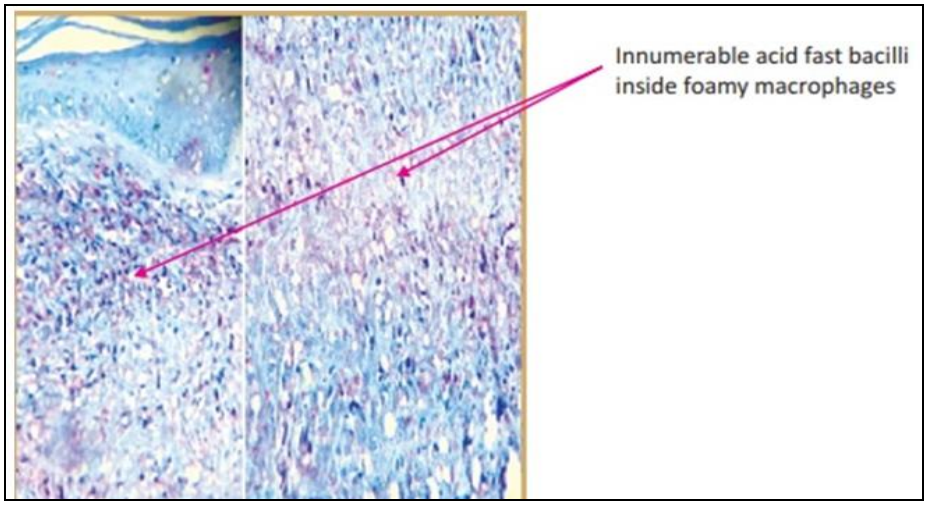


FIG. 6: ON FITE FARACO STAINING PINK SOLIDLY STAINED BACILLI ON A BLUE BACKGROUND SEEN

A diagnosis of lepromatous leprosy was made for the patient as a result of the clinical, bacteriological, and histological results. He began receiving multi-bacillary multidrug therapy (MB-MDT), and monthly follow-up was indicated.

DISCUSSION: In their case report of PKDL in a 50-year-old male who presented with diffuse lip enlargement and the perioral area extending onto the cheeks on both sides, which corresponds to the muzzle area, the equivalent region of the primate muzzle area of the face, Arora *et al.* (2015) provided the first instance of the muzzle area being involved. A granulomatous infiltrate was discovered during a skin biopsy, and tests for LD bodies and rK39 were both negative. A tissue sample's polymerase chain reaction (PCR) for leishmania also revealed a repeatedly positive result for rK39. Based on the aforementioned facts, he was identified as having PKDL and was given the appropriate treatment [1].

Similar lesions including the lips, chin, and lower aspect of the cheeks on both sides are present in our patient, a male 51-year-old. Multiple solid staining, acid-fast bacilli with a morphological index of 80%, a bacteriological index of 6+, and this observation were found in slit skin smears with ZN staining. Fite-Faraco staining also supported this finding. It was determined by a skin biopsy that the patient had lepromatous leprosy. This finding in our case is more of a morphological entity, and similar muzzle area involvement has been reported in other diseases such as granulomatous rosacea, sarcoidosis, the papular stage of mycosis fungoides, tumors of the salivary glands such as pleomorphic adenomas, and mucoepidermoid carcinomas of the oral cavity. Shiny papules that are skin-colored and across the upper lip can also be caused by granular cell tumors, angioleiomyomas, mucoceles, schwannomas, oral focal mucinosis, *etc.*

However, the presence of comparable lesions or infiltrative papules involving the trunk and extremities with bilaterally enlarged nerves and skin biopsy findings eliminates the possibility of other diseases and confirms our diagnosis of lepromatous leprosy while also emphasizing the fact that this appearance is not unique to PKDL [3]. Till date, only one case report of the muzzle area involvement in lepromatous leprosy in a male patient by KH Reganti and her colleagues has been documented. Mycobacterium leprae multiplies uncheckedly and spreads across the entire body in lepromatous leprosy, with the host mounting very little resistance (CMI).

The patients may exhibit many macules, papules, nodules, or all three of these skin abnormalities at once. These many cutaneous lesions are symmetrically and bilaterally distributed. Nasal symptoms and edema in the legs and ankles may appear months or years before the typical skin lesions [4]. A temperature of less than 37 degree Celsius is preferred by M. leprae for optimum growth. Skin, nasal mucosa, and peripheral nerves areas where the temperature is lower than that of the core body are the most commonly affected by leprosy. The unique localization of lesions in the perioral region as found in our patient may be caused by the organisms' preference for cooler locations [5].

There are many clinical, bacteriological, and immunological parallels between leishmaniasis and leprosy. Organisms that are obligatory intracellular are the root of both diseases. Clinical and pathological manifestations are influenced by the host reaction, most likely as a result of genetic predisposition and environmental factors. The patient exhibits localized lesions with well-formed granulomas and few to no organisms at the hyperergic pole, but at the anergic pole, the lesions are broad, there is no epithelioid granulomatous reaction, and there are many parasites [6]. We therefore suggest that M. leprae can also influence the muzzle area of the face, as seen in our case imitating PKDL, given the similarity of the immuno-pathophysiological involvement caused by both *M. leprae* and *L. donovani* and the preferential involvement of the colder portions of the body.

CONCLUSION: It's unusual for this patient to have skin-colored papules covering the lips and the perioral area that extends across both cheeks, creating the appearance of a monkey snout. Since there has been only one case report recorded before, to the best of our knowledge and understanding of the literature, our case may help in contributing to the literature of involvement of the snout area of the face in lepromatous leprosy.

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CONFLICTS OF INTEREST: There are no conflicts of interest.

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