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Case Report

Muzzle Area Involvement in Lepromatous Leprosy

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In the past, muzzle area involvement has been reported in a case of post-kala-azar dermal leishmaniasis. Here, we report a case of lepromatous leprosy (LL) with unusual muzzle area involvement predominantly with nodular skin lesions. Our case report describing the involvement of the muzzle area of the face in lepromatous leprosy, is very rare and is thus a valuable addition to literature on clinical presentations of leprosy.

Keywords : Muzzle Area, Lepromatous Leprosy, Post Kala-azar Dermal Leishmaniasis

Introduction

"Muzzle area" of the face involves lips and the perioral area extending onto the cheeks on both sides, the equivalent region of the primate muzzle area of the face. In the past, muzzle area involvement has only been reported in a case of post kala-azar dermal leishmaniasis (PKDL), which was characterised by lesions around the lips and the perioral area, giving the appearance of the primate muzzle (Arora et al 2015).

In leprosy, muzzle area involvement has only been described in sooty mangabey monkeys (*Cercocebusatys*) who were discovered to have acquired leprosy naturally (Meyers et al 1994).

Here we report an interesting case with similar involvement in a patient with lepromatous leprosy.

Case Report

A 25-year-old male patient presented with a history of skin-coloured raised lesions, which were insidious in onset and were initially noticed over the lower lip. Slowly, they progressed to involve the upper lip, the chin and extended onto the cheeks on both sides. Over a period of one year, these lesions gradually progressed to involve the face, trunk and extremities bilaterally. The patient complained of bilateral swelling over the legs, which he noticed at the end of the day, which would relieve on resting. He gave the history of nasal stuffiness, which was associated with the

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change in voice and epistaxis. There was no history suggestive of sensory or motor nerve involvement. There was no history of lethargy, weight loss, generalised malaise, dyspnea, cough, chest pain, palpitations. There was no history of red-coloured raised facial lesions associated with flushing on the consumption of hot spicy food, no history of hardening of nose or lips. The patient denied history of redness of eyes, foreign body sensation, photophobia or blurred vision. The patient did not have a history of any recent travel to Bihar or Jharkhand.

On examination, multiple skin coloured to erythematous, smooth, shiny discrete papules, measuring approximately 2 to 5 millimeters in diameter, were present over the lips, predominantly involving the vermilion border of the upper lip. Multiple skin coloured smooth, shiny papules to nodules, along with a few infiltrative plaques, of varying sizes, ranging from 0.5 to 3 centimeters, were present over face, trunk, buttocks and both the extremities in a symmetrical fashion. Also observed were nodular ear lobe infiltration and bulbous enlargement of the nose (Fig. 1).

On peripheral nerve examination, all the peripheral nerves were bilaterally enlarged, non-tender, firm in consistency with no nodularity. On sensory examination, sensations for temperature, pain, touch were all intact. On voluntary muscle testing (VMT), there was normal power according to the grading given by the medical research council (MRC). Corneal reflex, light reflex, biceps reflex, triceps reflex, brachioradialis reflex, knee jerk, ankle jerk were all intact. With the above features, we considered the differential diagnoses of lepromatous leprosy, sarcoidosis, post kala-azar dermal leishmaniasis, granulomatous rosacea and the tumour stage of mycosis fungoides.

Slit skin smears were performed from six sites. Both Ziehl-Neelsen (ZN) staining and Giemsa staining was done on the smears. On ZN staining, multiple solid staining acid-fast bacilli were noticed, wherein the morphological index (MI)

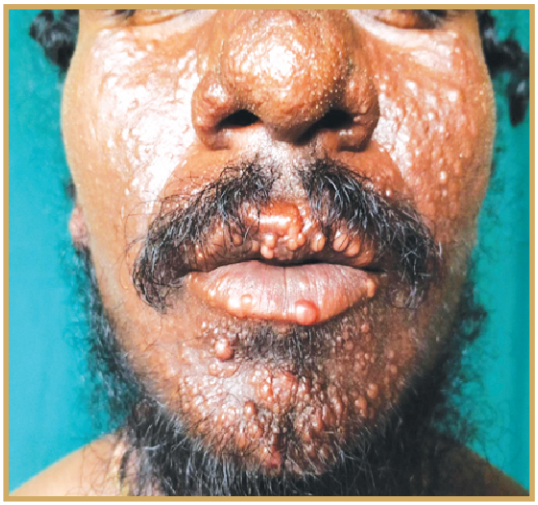

Fig. 1 : Multiple shiny skin coloured to erythematous papules present over the muzzle area (vermilion border of lips, cheeks), chin, nose

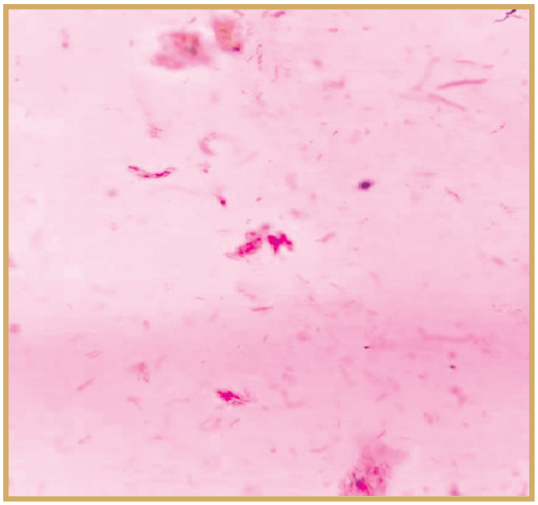

Fig. 2 : Slit skin smear showing multiple solid staining acid fast bacilli with a Bacillary Index 6+ (Modified Ziehl-Neelsen stain, x 1000)

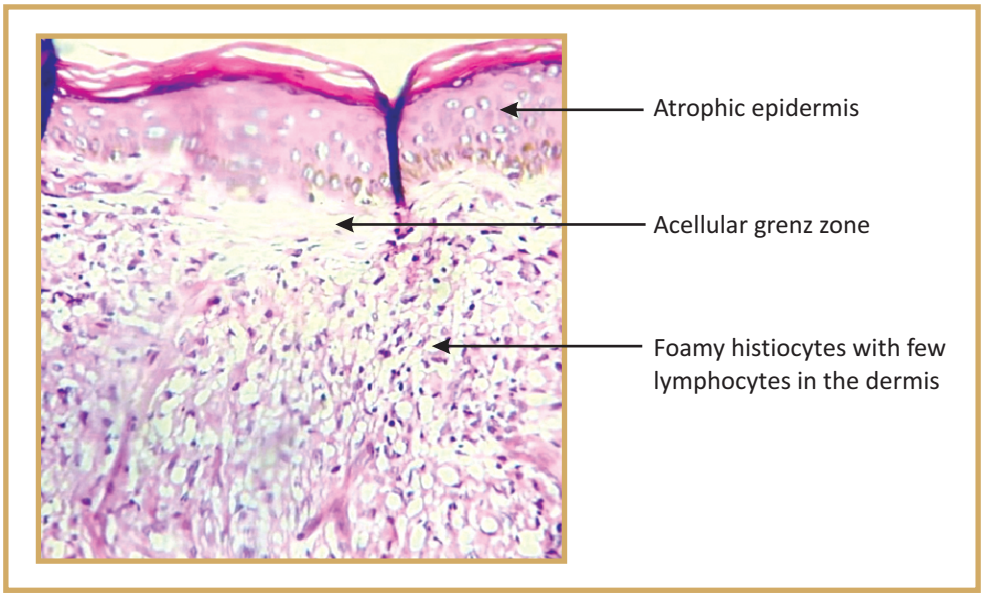

Fig. 3 : Photomicrograph showing atrophic epidermis, with an acellular Grenz zone in the subepithelium. Dermis showing sheets of foamy histiocytes with few lymphocytes. (H & E, x 40)

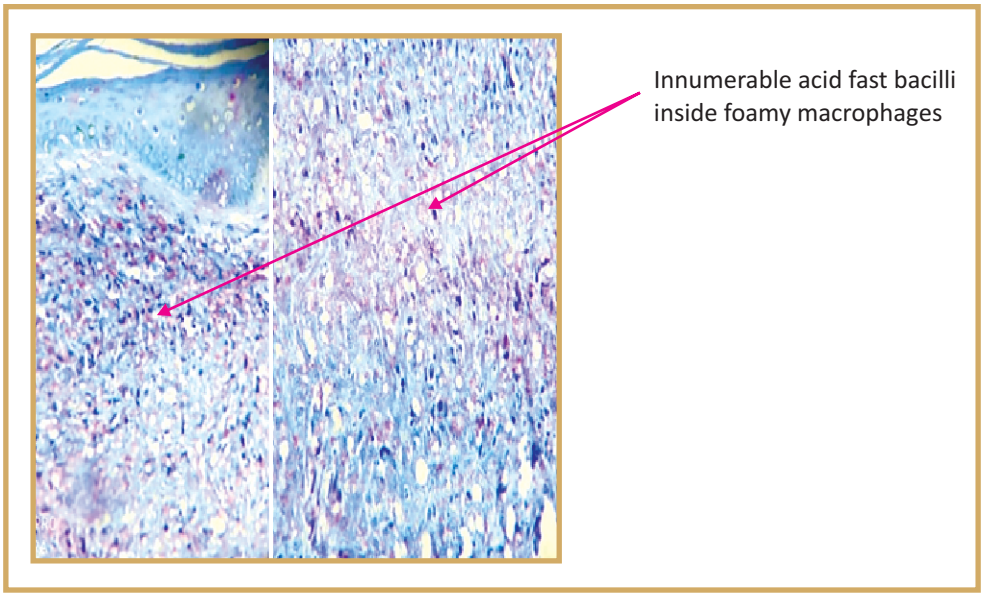

Fig. 4 : Innumerable solid staining acid fast bacilli in biopsy sample (Fite Faraco, x 40)

was 80% and the bacteriological index (BI) was 6+ (Fig. 2). Giemsa staining was negative for Leishmania Donovan (LD) bodies. Skin biopsy showed atrophy of epidermis with the subepithelium showing an acellular grenz zone. There were sheets of foamy histiocytes within the dermis that extended upto the deep dermis, with few lymphocytes (Fig. 3). Fite Faraco staining for acid-fast bacilli in the tissue section was done, which showed multiple solid staining red coloured bacilli on a blue background (Fig 4). Special staining with reticulin stain was negative and there were no well-formed naked granulomas. Serum calcium and angiotensin converting enzyme levels were unremarkable. Chest X-ray, showed no abnormalities.

Thus, the patient was diagnosed as a case of lepromatous leprosy based on the clinical, bacteriological (SSSE) and histopathological findings. He was started on multi-bacillary multidrug therapy (MB- MDT) and was advised for monthly follow up.

Discussion

The first instance of involvement of muzzle area was given by Arora et al (2015), in their case report of PKDL occurring in a 50-year-old male who presented with diffuse enlargement of lips 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. On skin biopsy, a granulomatous infiltration negative for acid-fast bacilli as well as LD bodies was revealed, immunochromatography test for rK39 was persistently positive, and polymerase chain reaction (PCR) for leishmania from the tissue sample was also positive. He was diagnosed as a case of PKDL based on the above findings and was treated accordingly.

Our patient, who is a 25-year-old male, also has similar lesions with involvement of lips, chin extending onto the cheeks on both sides. Slit skin smears with ZN staining revealed multiple solid staining acid-fast bacilli with a morphological index of 80%, a bacteriological index of 6+ and this observation was confirmed by Fite-Faraco staining. On skin biopsy, it was confirmed as a case of lepromatous leprosy.

This observation made in our case is more of a morphological entity and a similar involvement of muzzle area can also be seen in other conditions like granulomatous rosacea, sarcoidosis, papular stage of mycosis fungoides, salivary gland tumors, like pleomorphic adenomas, mucoepidermoid carcinomas of the oral cavity. Angioleiomyomas, mucoceles, schwannomas, oral focal mucinosis, granular cell tumors, also present as skin ϵoloured shiny papules over the upper lip. However, the presence of similar lesions or infiltrative papules involving the trunk and extremities with bilaterally enlarged nerves and findings of skin biopsy rules out the possibility of other conditions and confirms our diagnosis of lepromatous leprosy and also emphasizes the fact that this appearance is not limited only to PKDL (Bhandarkar & Shetty 2017).

In lepromatous leprosy, there is unchecked multiplication and universal spread of *Mycobacterium leprae*, with almost no resistance (CMI) mounted by the host. The patients may present with innumerable macules, papules, nodules or with all three types of skin lesions. These innumerable skin lesions have a bilateral and symmetrical distribution. Nasal symptoms and edema of legs and ankles may precede the classical skin lesions by months or years (Jopling & McDougall 1996).

M. leprae has a preference for a temperature less than 37°C for its optimal growth. Leprosy predominantly involves skin, nasal mucosa, peripheral nerves, where the temperature is less than the core body temperature. The specific localisation of lesions in the perioral area as seen

in our patient may be due to the preferential selection of the organisms for cooler areas (Sekar 2015).

Leishmaniasis and leprosy share a lot of clinical, bacteriological and immunological similarities. Both diseases are caused by obligate intracellular organisms. The clinical and pathological expressions depend on the host response, probably due to genetic determination and environmental influences. At the hyperergic pole, the patient shows localised lesions with well-formed granulomas with few or absent organisms, whereas at the anergic pole, the lesions are widespread, there is no epithelioid granulomatous reaction, and there are numerous parasites (Trindade et al 2015).

Hence we propose that in view of the similarity of the immuno-pathophysiological involvement induced by both *M. leprae* and *L. donovani* and the preferential involvement of the cooler areas of the body, *M. leprae* can also affect muzzle area of the face as shown in our case mimicking PKDL.

Conclusion

In this patient, the presence of skin-coloured papules over the lips and the perioral area extending onto cheeks on both sides, giving an appearance of the primate muzzle, is a rarity. Our case may help in contributing to the literature of involvement of muzzle area of the face in lepromatous leprosy since this has not been reported before to the best of our knowledge and literature search.

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