Symmetrical Centrofacial Erythematous Plaques and Papules: A Clinicopathological Challenge

Abstract

Facial papular eruptions remain a diagnostic dilemma for the dermatologist with a wide range of inflammatory and infectious conditions manifesting in this manner. Here, we present a case of a 29-year-old, otherwise healthy man from North India with asymptomatic well-defined normoaesthetic symmetrical papules and plaques over the upper and mid-face of 3 months duration. Skin biopsy showed perivascular and periappendageal well-defined collections of foamy macrophages and epithelioid cells in superficial and deep dermis, characteristic of borderline lepromatous leprosy. Though acid-fast stain for lepra bacilli was negative both on lesional biopsy specimen and lesional and ear lobe slit skin smear, a 16s ribosomal ribonucleic acid (16s-r-RNA) polymerase chain reaction on skin biopsy specimen was found to be positive for lepra bacilli. A final diagnosis of borderline lepromatous leprosy in type I reaction was made and the patient received World Health Organization (WHO) multibacillary (MB) multidrug therapy along with oral steroids. This case highlights the unusual localized involvement in a case of MB leprosy lacking all the three cardinal features of leprosy, i.e. sensory loss, peripheral nerve involvement, and acid-fast bacilli positivity on biopsy or slit skin smear but diagnosed on the basis of characteristic histology and positive polymerase chain reaction results.

Keywords: *Centrofacial eruption, foamy macrophages, localized multibacillary leprosy, polymerase chain reaction*

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Introduction

Numerous dermatoses ranging from inflammatory to infectious conditions can present with facial eruption. The final diagnosis depends on the clinicopathological correlation especially in cases with overlapping and atypical clinical manifestations. The dilemma is heightened in a condition like leprosy which itself is known to be a "great imitator" with varied presentations. Multibacillary (MB) forms of leprosy usually present with generalized hypopigmented to skin-colored patches, plaques, or nodules. However, exclusive facial involvement has only rarely been reported.[1,2] Herein, we report an interesting case with symmetrical centrofacial rash, diagnosed as borderline lepromatous leprosy on the basis of histology and polymerase chain reaction (PCR).

Case Report

A 29-year-old, otherwise healthy man presented with asymptomatic persistent erythematous lesions over the face for the past 3 months. The lesions started over the forehead as faint erythema, which gradually progressed to involve the cheeks and lower face over a 1 month period. They also become more elevated and larger over time. There was no history of trauma, photosensitivity, or topical application. He received topical steroids and antifungals from outside with a clinical diagnosis of seborrheic dermatitis with no improvement. He was a resident of Bihar and there was no significant personal or family history. On examination, there was symmetrical involvement of the upper and mid-face in the form of well-defined erythematous shiny succulent plaques and papules [Figure 1a]. The lesions were confluent in the center and smaller discrete papules around the eyelids [Figure 1b]. There was no lesional sensory loss, madarosis, ear lobe infiltration, extrafacial skin lesions, or mucosal involvement. He had incomplete closure of the right eye secondary to some iatrogenic injury in childhood, which was static with no evidence of facial nerve

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palsy. Systemic examination including peripheral nerve examination was normal. Based on history and examination, differential diagnoses of orofacial granulomatosis, granulomatous rosacea, lupus miliaris disseminatus faciei, sebopsoriasis or seborrheic dermatitis, and sarcoidosis were kept. Skin biopsy showed well-defined collections of epithelioid cells, foamy macrophages, lymphocytes, and occasional multinucleate giant cells in a perivascular and peri-appendageal location in the superficial and deep dermis with mild papillary dermal edema [Figure 2a and b]. Peri-neural infiltrate could not be assessed in view of the absence of nerves in the histology sections examined. Modified Ziehl–Neelsen stain, Periodic Acid-Schiff, Giemsa stain, and Warthin–Starry stains were negative. Slit skin smears (SSS), both lesional and bilateral ear lobes, were negative. Polymerase chain reaction (PCR) on biopsy specimen was positive using the template 16s r-RNA and leprae-specific repetitive element (RLEP) antigen. No sensorimotor deficit was detected on nerve conduction studies of limbs. Serology for leishmania and syphilis was negative.

A final diagnosis of Borderline lepromatous (BL) leprosy with type I reaction localized to the face was made. The patient was started on WHO MB multidrug therapy. Initially, aspirin was given in the dose of 300 mg three times a day along with pantoprazole 40 mg once daily before breakfast for the management of leprosy reaction. The patient did not show any change, and hence, prednisolone (40 mg once daily after breakfast) was added on the seventh day. Prednisolone was tapered by 10 mg every month and stopped in the fifth month of follow-up with marked improvement [Figure 3]. Though the patient was on concomitant steroids and aspirin, he did not have any adverse gastrointestinal side effects.

Discussion

Facial papular eruptions remain a diagnostic dilemma for the dermatologist with a wide range of inflammatory and infectious conditions manifesting in this manner. Correct diagnosis is often established only after clinicopathological correlation. MB leprosy classically presents with multiple hypo to normoaesthetic plaques, nodules, or diffuse infiltration of skin in a generalized distribution along with symmetric peripheral nerve involvement in the form of sensory or motor deficit and nerve thickening. Nevertheless, given its enigmatic nature, it can rarely present with atypical features like localized lesions with or without sensory loss and peripheral nerve involvement. Ranjan *et al*.[1] and Gupta *et al*.[2] have reported MB leprosy with exclusive facial involvement. In the present case, the exclusive facial involvement without sensory loss and peripheral nerve involvement made leprosy an unlikely clinical differential.

However, the histological pattern of the perivascular, periappendageal, and/or perineural infiltrate of foamy macrophages, lymphocytes with or without epithelioid cells as seen in the present case is considered pathognomonic of MB-leprosy.[3] Besides leprosy, other clinical differentials kept like orofacial granulomatosis (OFG), granulomatous rosacea (GR), lupus miliaris disseminatus faciei (LMDF), seborrheic dermatitis, and sarcoidosis could not explain the histological pattern, especially the presence of foamy macrophages. OFG shows loose epithelioid cell granuloma without any specific predilection for perivascular and periappendageal location. GR is characterized by dilated capillaries in the upper dermis along with a few epithelioid cell granulomas in the upper and mid-dermis, while compact perifollicular granulomas with caseous necrosis are diagnostic of LMDF. Seborrheic dermatitis shows spongiotic dermatitis, while sarcoidosis shows compact closely spaced yet discrete epithelioid cell granulomas. On the other hand, conditions which show foamy macrophages on histology like xanthoma, xanthogranuloma, post kala-azar dermal leishmaniasis (PKDL), syphilis, fungal and atypical mycobacterial infections, also could not justify the clinicopathological presentation. Xanthoma and xanthogranuloma were unlikely due to a lack of yellowish hue clinically and so were PKDL and syphilis in view of the absence of plasma cell-rich infiltrate, negative serology, and absence of organisms on special stains. Fungal and

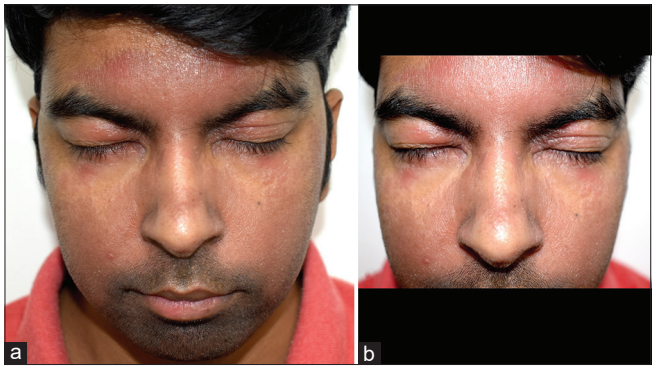

Figure 1: (a) Symmetrical erythematous plaques and papules over the upper and mid-face (b) Discrete small papules over the periocular region

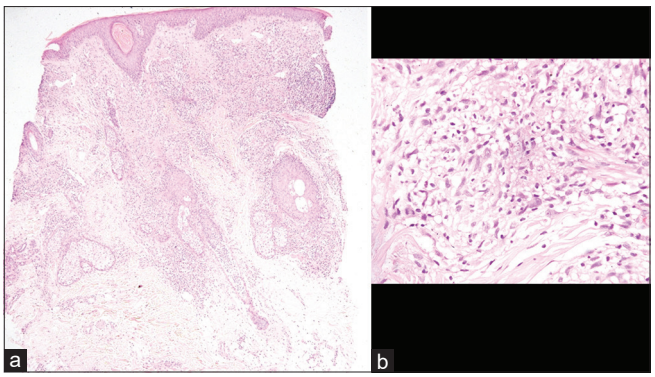

Figure 2: (a) Perivascular and periappendageal nodular collections of foamy macrophages, epithelioid cells, and lymphocytes in the superficial and deep dermis (H and E, 40X). (b) Foamy macrophages with epithelioid cell collections (H and E, 400X)

Figure 3: Flattening of facial skin lesions at 5 months follow-up

atypical mycobacterial infections were excluded due to the lack of eosinophils and no organisms detected on the special stain.

Nevertheless, despite the typical histological features of MB-leprosy, Acid fast bacilli (AFB) stain was negative in our case which added to the diagnostic dilemma. AFB stain is usually positive in MB-leprosy. However, AFB can rarely be negative.[4] In such cases, clinicopathological correlation alone can help in making the correct diagnosis. Another clue toward the diagnosis of leprosy in our case was PCR positivity. The sensitivity of PCR ranges from 80 to 100% in MB cases with 100% specificity.[5] PCR of skin biopsy specimen has been found to be more sensitive than AFB staining on SSS samples.[6] The good response to anti-leprosy therapy is another crucial evidence in favor of the diagnosis.

In conclusion, MB-leprosy can present with exclusive facial involvement without cardinal features of sensory loss, peripheral nerve involvement, and AFB positivity. Characteristic histology and positive PCR results along with the exclusion of other mimickers can confirm the diagnosis.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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