

## Introduction

Multicentric reticulohistiocytosis (MRH) is an insidious onset, non-Langerhans cell histiocytosis affecting joints and skin. Early diagnosis is important to prevent destructive arthritis and disease related complications. Reflectance confocal microscopy (RCM) is a non-invasive imaging technique to visualize the epidermis and superficial dermis on a horizontal plane and provides quasi-histologic view of the skin. RCM features of non-Langerhans cell histiocytosis including juvenile xanthogranuloma (JXG), adult xanthogranuloma (AXG) and Rosai-Dorfman disease have been reported in few cases. To date, reflectance confocal microscopy (RCM) features have not been described for MRH. Herein, we report a case of MRH and describe the reflectance confocal microscopy features and distinctive clinical features.

## Case History

A 59-year-old female was referred to our clinic for multiple erythematous lesions on the head, neck, and extremities (Fig 1). Dermoscopic and reflectance confocal microscopic images were acquired. RCM images showed bright ringed-edged papillae with infiltration of small-bright cells around dilated round blood vessels at the level of DEJ. Round, well-demarcated, hypo-refractile cells were observed within the dermal papilla (Fig 2). Three lesions were sent for biopsy. Biopsy revealed abundant lymphohistiocytic infiltrate of the dermis with multinucleated giant cells with oncocyctic cytoplasm (Fig 3). The patient was diagnosed with MRH.

## Discussion

The diagnosis of MRH can be challenging due to overlap of clinical features with rheumatologic disorders.<sup>1-7</sup> MRH may present with dermatomyositis or lupus-like like features.<sup>3,6,7</sup> Histology can provide definitive diagnosis; however, skin biopsies are not performed routinely in these patients. Thus, RCM can be a useful tool to diagnose these patients.

RCM features of non-Langerhans cell histiocytosis (AXG, JXG) have been previously described as dilated dermal papillae filled with clusters of round giant cells with conspicuous nuclei and bright cytoplasm.<sup>1,14-17</sup> RCM features of Rosai-Dorfman disease have been reported as medium-refractile large polygonal cells and edged papillae with small bright cells.<sup>18</sup> In our case, we observed foci of disarranged keratinocytes with small bright cells at the level of DEJ. Most importantly we were able to identify oncocyctic cells within papillary structures with RCM.

RCM can diagnose subtle cases of MRH by showing distinctive RCM features, and can enable physicians to diagnose this rare disease in its earliest stages, preventing MRH-related morbidity.

## Images

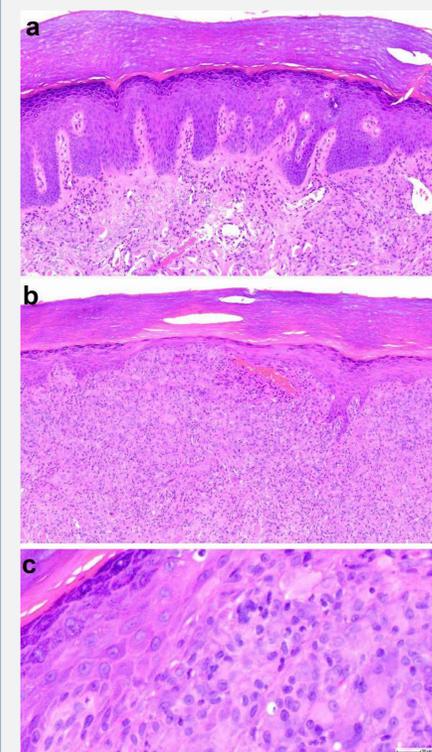
**Figure 1: Clinical and dermatoscopic images of the patient.**



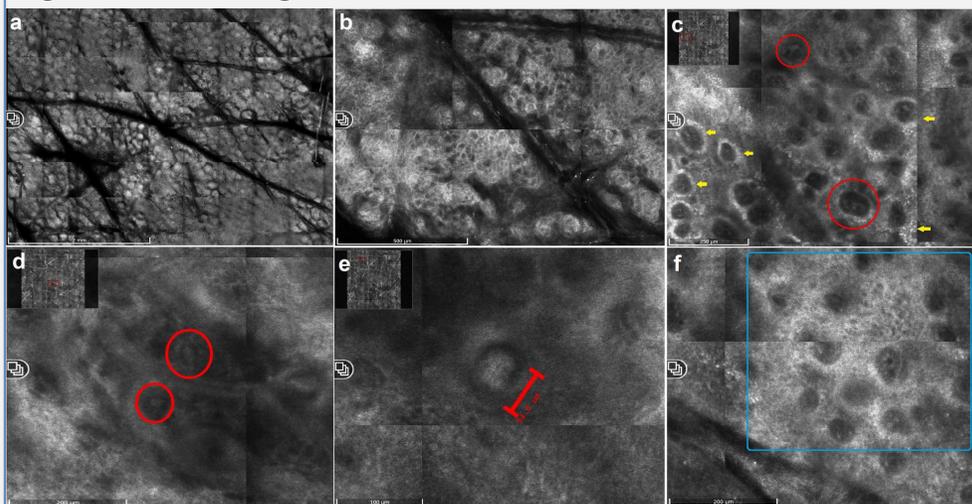
- Periorbital and malar erythema with multiple erythematous papules on frontal, nasal, perinasal and periorbital skin, 0.2 to 2 cm in diameter.
- Pink, clustered papulonodular lesions; left elbow.
- Periungual papular lesion resembling 'coral bead' appearance with pink nodules on the dorsal phalanges and erythema on the metacarpophalangeal joints resembling Gottron's papules.
- A pink papule on the lateral aspect of the left index finger.
- Erythematous rash on the head and neck extending to nape, 'V-neck erythema'.
- Exophytic, soft, dome-shaped nodules; right buccal mucosa.
- Dermatoscopic image of the lesion on the lateral aspect of the index finger showing a raised lesion with central yellow structure with dotted vessels and surrounding erythema.

- Epidermal hyperplasia, papillary dermal fibrosis, and dilated blood vessels in the superficial dermis.
- Abundant lymphohistiocytic infiltrate of the dermis with multinucleated giant cells.
- A close view of multinucleated giant cells with 'ground-glass' appearance.

**Figure 3: Histologic images of a palmar lesion.**



**Figure 2: RCM images of the lesion located on elbow.**



- RCM at the spinous layer revealing foci of atypical honeycomb pattern
- Irregular size of nuclei and thickness of cellular outlines.
- Bright ringed-edged papillae with infiltration of small-bright cells around dilated round blood vessels at the level of DEJ (yellow arrows).
- Round well-demarcated hypo-refractile cells within dermal papilla (red circles).
- Moderately refractile, round-shaped histiocytes 54  $\mu$ m in diameter in the papillary dermis
- Moderately refractile, lace-like collagen bundles at the level of superficial dermis (blue box).

## References

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