Dermoscopic and reflectance confocal microscopic findings of cutaneous Rosai-Dorfman disease

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To the Editor: Rosai-Dorfman disease (RDD), also known as sinus histiocytosis with massive lymphadenopathy, is a rare non-Langerhans cell histiocytic proliferative disorder. RDD is characterized by bilateral cervical lymphadenopathy accompanied by extranodal involvement, with the skin being the most commonly affected organ.[1] Cutaneous RDD (CRDD), affecting only the skin without systemic symptoms, is even rarer.[2] Herein, we report the dermoscopic and reflectance confocal miscroscopy (RCM) findings in a patient with CRDD.

A 57-year-old man, presented to the dermatology department with a 6-month history of multiple progressive nodule on his trunk. Physical examination revealed no enlargement of superficial lymph nodes, and no obvious abnormalities on cardiopulmonary and abdominal examinations. Dermatological examination revealed multiple smooth heliotrope plaques or nodules on his chest, abdomen, and back [Figure 1A]. Dermoscopic examination was carried out using an immersion polarized dermoscopy (FotoFinder Medicare 800HD, FotoFinder-Systems GmbH, Birbach Germany) revealed yellow discoloration on a background of erythema, with larger branching blood vessels at the periphery of the lesions and smaller ones around the yellowish structureless foci [Figure 1B].

Further pre-operative evaluation using RCM (VivaScope 1500, Lucid Technologies, Henrietta, NY, USA) revealed preservation of a typical honeycomb pattern in the stratum spinosum, as well as numerous slender high-refractive dendritic cells and some small bright cells [Figure 1E]. Numerous small, round, high-refractive cells corresponding to inflammatory cells clustered in the dermis [Figure 1H]. Large polygonal cells comprising medium-refractive peripheral rings enclosing small, round, high-refractive structures were observed in the superficial dermis [Figure 1H and 1I].

Abdominal lesions biopsy revealed histiocytes and large numbers of lymphocytes, neutrophils, and plasma cells aggregated into nests. Several inflammatory cells were engulfed in the histiocytes [Figure 1G and 1J], referred to as emperipolesis. Immunohistochemical analysis showed that the cells infiltrating the dermis were positive expression of CD68 and S100 [Figure 1C and 1D], and negative of CD1a [Figure 1F]. Ultrasound, computed tomography, and laboratory tests were normal. A diagnosis of CRDD was finally made.

CRDD lesions can be classified as coalesced plaques, single or multiple isolated nodules, multiple papules, or tumor type.[1] The current case was characterized by multiple reddish nodules on the trunk. Dermoscopic findings in previous cases of CRDD showed yellow foci and irregularly branching vessels,[3] as in the present. Yellow foci may thus be related to the accumulation of inflammatory cells in the dermis.[4]

RCM showed a typical honeycomb pattern in the epidermis, but multiple dendritic cells were also observed, corresponding to activated dendritic Langerhans cells (CD1a-positive). Numerous small, round, high-refractive cells corresponding to inflammatory cells were detected in superficial dermis. Xanthogranulomas include giant atypical cells with high-refractile peripheral rings, corresponding to touton cells.[5] However, the large polygonal cells in the current patient differed from the previous description, showing medium-refractive peripheral rings, possibly indicating multinucleated cells, as also observed in CRDD.[6] The small, round, high-refractive structures inside the large cells may represent phagocytized inflammatory cells, possibly correspond to the histological emperipolesis phenomenon.

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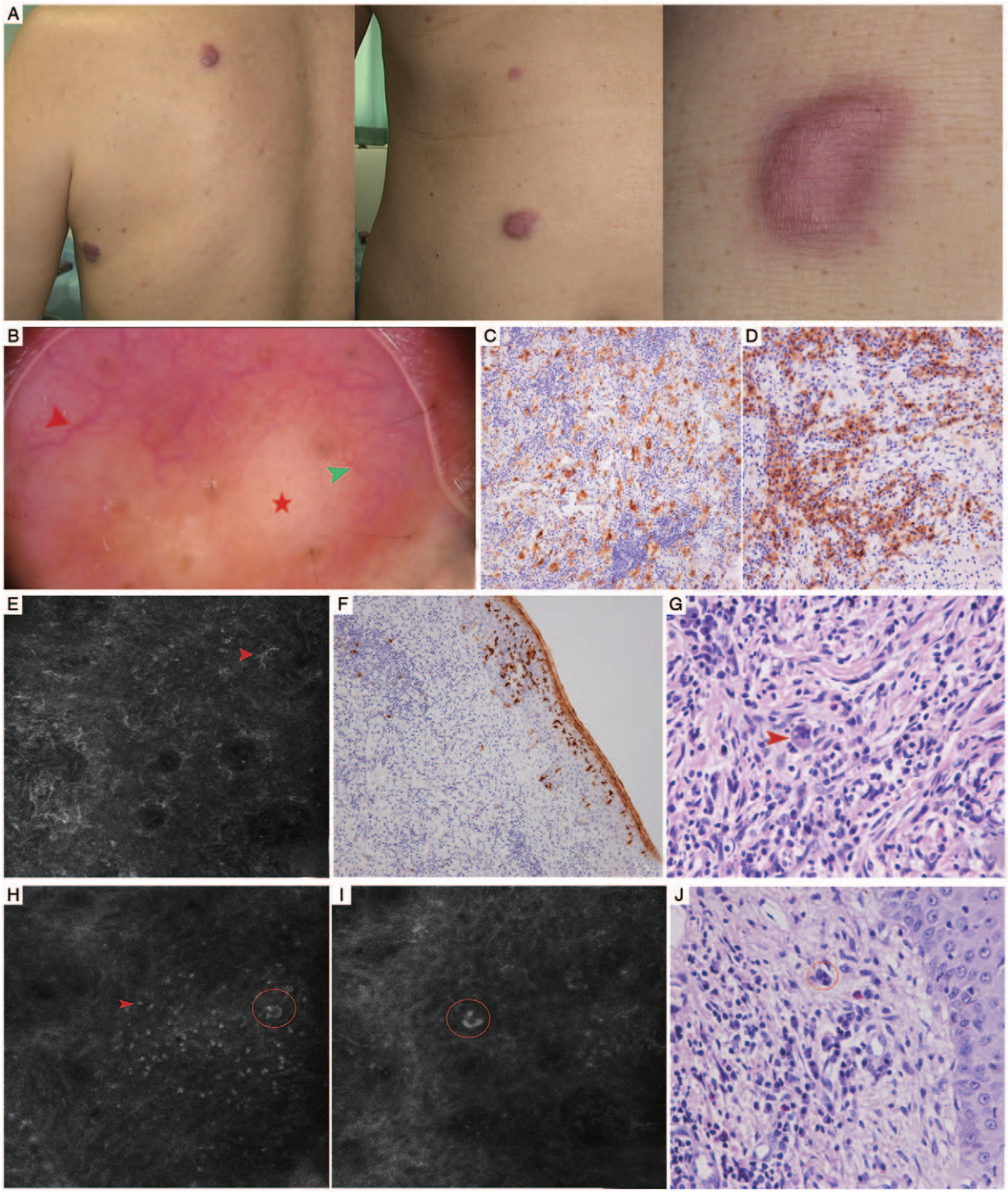

Figure 1: Dermoscopic and RCM features of CRDD. (A) Clinical features: multiple isolated nodule on the trunk; (B) Immersion polarized dermoscopy revealed yellow discoloration (pentacle) and larger branching vessels (read arrowhead) and smaller branching vessels (green arrowhead); (C and D) Immunohistopathology (HE, 10×) showed positive expression of CD68 and S-100. (E) RCM image (0.5 mm × 0.5 mm) taken at the level of epidermis, and showed multiple dendritic cells (read arrowhead); (F) Immunohistopathology (HE, 10×) showed positive expression of CD1a in epidermis and negative in dermis, and revealed activated langerhans cells. (G and J) Histology (HE, 40×) showed large numbers of lymphocytes, neutrophils, plasma cells aggregation, and emperipolesis phenomenon (circles); (H and I) RCM revealed numerous small round high-refractive cells (read arrowhead) and larger polygonal cells with medium refractive peripheral semicircle and small, round, high-refractive cells (red circles). CRDD: Cutaneous RDD; RCM: Reflectance confocal miscroscopy.

In summary, the yellowish foci and irregularly branching vessels may be key clues for the dermoscopic diagnosis of CRDD, while large polygonal cells with medium-refractive peripheral rings and small, round, high-refractive centers might be characteristic RCM structures. The diversity of skin lesions increases the difficulty of diagnosing CRDD. The combination of dermoscopy and RCM may provide additional clinical information to improve the accuracy of diagnosis. However, more cases are need to be investigated, and the detection depth of RCM is still an issue to be considered.

Conflicts of interest

None.

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