Atypical Presentation and Dermoscopic Evaluation of Cutaneous Rosai-Dorfman Disease

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Sir,

Sinus histiocytosis with massive lymphadenopathy or Rosai-Dorfman disease (RDD) is a rare benign self-limiting histiocytic disorder, which is confined mainly to cervical, but also other lymph nodes. Extranodal involvement is present in 43% of cases of RDD and the skin is the most commonly affected site (1). A purely cutaneous form of the disease, termed cutaneous RDD, is even more uncommon and has been well defined (2). Histopathological features include the presence of a dense, dermal infiltrate composed mainly of typical, large, polygonal histiocytes with feathery borders, abundant pale eosinophilic cytoplasm, vesicular nuclei and small nucleoli. Intact lymphocytes, plasma cells and neutrophils are usually seen within the cytoplasm of histiocytes, a phenomenon known as emperipolesis (3). There seem to be no differences between cutaneous RDD and the systemic form in terms of pathological findings (4). Immunohistochemistry typically shows positivity for S-100 in the histiophagocytic cells, which often highlights emperipolesis, but they are negative for CD1a.

CASE REPORT

A 25-year-old black man presented with a 5-month painful lesion on the sole of the left foot, which had been treated previously by his podiatrist as a viral wart with topical salicylic acid and curette on different occasions over a period of 10 weeks.

The patient was a native of central Africa and there was no relevant medical history apart from previous filariasis on his right leg and a varicella infection 8 weeks previously. No fever, asthenia, weight loss or other general symptoms were reported. He had been treated with topical salicylic acid and curetage on several attempts, the patient was treated as for neurosyphilis, with intravenous aqueous penicillin G. A 4-month follow-up showed no recurrence of the lesion and no cutaneous or systemic involvement.

On the basis of the clinical, serological, histopathological and immunohistochemical findings a diagnosis of cutaneous RDD and latent syphilis was made.

As it was not possible to perform a lumbar puncture after several attempts, the patient was treated as for neurosyphilis, with intravenous aqueous penicillin G. A 4-month follow-up showed no recurrence of the lesion and no cutaneous or systemic involvement was detected.

DISCUSSION

Only a few cases of purely cutaneous RDD have been reported since the first description of the systemic disease...
A possible aetiopathogenic role of these infections could be speculated. As far as we know, a planar location of a solitary lesion of RDD has not been reported previously, and an abnormal activation of histiocytes after local trauma or past local infection cannot be ruled out.

Milky-white ovoid structures with a cotton-like appearance over a light-red background were observed with dermoscopy. This image is similar to those observed in xanthogranulomas, with yellowish cloud-like structures over an orange-yellow background, being the “clouds” identified as the xanthogranulomatous dermal infiltrate (13) and resembling the yellow nodules present in sebaceous hyperplasias (14), also named “cuminum sign” (15). Similarly to xanthogranulomas, the milky-white globules in our patient would correspond to the dermal histiocytic infiltrate. Hence we can speculate that three types of lesions with common white-yellow globular structures but distinctive features can be recognized with dermoscopy: sebaceous hyperplasia with typical crown vessels, xanthogranulomas with orange background and yellowish hue of globules and cutaneous RDD with red background and whitish hue of globules, lacking the last two entities in blood vessels.

REFERENCES


Fig. 2. Centrally-located histiocytes exhibiting emperipolesis. H&E staining, original magnification ×200.