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 curettage 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. No relevant personal or family history was obtained. No relevant social history apart from daily consumption of alcohol was elicited. The patient was married and there were no children. No relevant travel or recent immigration history was elicited.

An excisional biopsy was performed and histopathological examination revealed a diffuse dermal infiltrate of lymphocytes, plasma cells and many large histiocytes, which extended close to the epidermis but did not infiltrate it. The histiocytes had abundant, pale pink, cytoplasm and vesicular nuclei. They showed emperipolesis of lymphocytes and neutrophils (Fig. 2). Immunohistochemistry showed diffuse staining in the histiocytes for protein S-100 and, to a lesser degree, for CD68, while CD1a was negative.

Cell blood count, liver and kidney function test, chest X-ray and abdominal ultrasonography were all normal and a discrete polycythaemic hypergamma globulinaemia was detected. HIV and hepatitis C antibody tests were negative and Epstein Barr virus (EBV), human herpesvirus (HHV)-1 and -2 tests were positive for immunoglobulin (Ig)G but not for IgM, being consistent with past infections. Anti-hepatitis B core antigen (anti-HBc) was positive without HBsAg and anti-HBc IgM positivity. Syphilis serology showed positive treponenic and reaginic tests (fluorescent treponemal antibody absorption ++, Treponema pallidum haemagglutination assay + 1/640, Venereal Disease Research Laboratory + 1/8, rapid plasma reagin ++) Ophthalmological examination did not reveal any alterations.

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

Fig. 1. (A) A 10-mm ulcerated skin lesion located on the central anterior part of the left sole with a clean bed and a hemorrhagic hyperkeratotic border. (B) Videodermoscopy of the lesion at 30-fold magnification showed cotton-like white irregularly distributed globular structures over a light-red background and a small ulceration.
by Destombes in 1965 (5) and the subsequent definition as a distinct clinicopathological entity by Rosai & Dorfman in 1969 (6). The epidemiological distribution of cutaneous RDD is different from the systemic form, meaning a predilection for young adults (mean age 20.6 years), males (1.4:1) and blacks and whites in the latter (1), and a predominance of Asian or white middle-aged (fifth decade of life) females in the former (2). Interestingly, the epidemiological characteristics of our patient match the profile of the systemic disease.

The clinical features of cutaneous RDD are heterogeneous and include single or, more commonly, multiple, indurated papules, plaques or nodules of different sizes with no anatomical predilection site. A 3-type classification based on the clinical morphology of 39 skin lesions has been made, the “papulonodular” type being the most common (79.5%), followed by the “indurated plaque” type (12.5%) and the “tumour” type (7.7%) in which our patient can be included (7). Other reported clinical cutaneous manifestations are erythema, fistulas, ulcers, bilateral cauliflower ear deformity and lesions resembling granuloma annulare, xanthomas, Peyronie’s disease, bilateral cauliflower ear deformity and lesions resembling granuloma annulare, xanthomas, Peyronie’s disease, bilaterally. Different types of lesions with common white-yellow globular structures but distinctive features can be recognized with dermatoscopy: sebaceous hyperplasias, 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 “cumulus 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 dermatoscopy: 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.