Cutaneous Rosai－Dorfman Disease：Clinicopathological profiles，spectrum and evolution of 21 lesions in six patients.

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摘要

Abstract

Background An uncommon histiocytosis primarily involving the lymph nodes, Rosai-Dorfman disease (RDI), originally called sinus histiocytosis with massive lymphadenopathy) involves extranodal sites in 43% of cases; cutaneous RDD (C-RDD) is a rare form of RDD limited to the skin. The clinicopathological diagnosis of C-RDD may sometimes be difficult, with different clinical profiles from those of its nodal counterpart, and occasionally misleading histological pictures. There have been few multipatient studies of C-RDD and documentation of its histological spectrum is rare. Objectives To identify the clinical and histopathological profiles, associated features, and the chronological changes of this rare histiocytosis. Methods From 1991 to 2002, patients diagnosed as having C-RDD were collected in four academic hospitals. Clinical presentations, treatments, and courses of each case were documented. In total, 21 biopsy specimens obtained from these patients were re-evaluated and scored microscopically with attention to the uncommon patterns and chronological evolution both clinically and histologically. Results We examined six patients with C-RDD, three men and three women. The mean age at the first visit was 43·7 years. The clinical presentations were mostly papules, nodules and plaques, varying with the duration and depth of lesions. Although the anatomical distribution was wide, the face was most commonly involved. Evolutional changes were identified clinically, as the lesions typically began with papules or plaques and grew to form nodules with satellite lesions and resolved with fibrotic plaques before complete remission. No patient had lymphadenopathy or extracutaneous lesions during follow-up (mean 50·5 months). At the end of follow-up, the lesions in four patients had completely resolved irrespective of treatment; two patients had persistent lesions. The histopathological pattern of the main infiltrate, the components of cells and the stromal responses showed dynamic changes according to the duration of lesions. The characteristic Rosai-Dorfman cells (RD cells) were found in association with a nodular or diffuse infiltrate in IS lesions (71%). Four lesions (19%) demonstrated a patchy/interstitial pattern.

One lesion (5%) assumed the pattern of a suppurative granuloma. RD cells were less readily found in these atypical patterns. Conspicuous proliferation of histiocytes associated with RD cells was found in three lesions, including xanthoma, localized Langerhans cell histiocytosis and xanthogranuloma. Along with lymphocytes, plasma cells were present in all lesions, often in large numbers with occasional binucleated or trinucleated cells. Variably found in the lesions were neutrophils (nine lesions, 43%) and eosinophils (13 lesions, 62%). The former occasionally formed microabscesses, while the latter were often few in number. Vascular proliferation was a relatively constant feature (90%). Fibrosis was found in 10 lesions (48%).