DERMATOPATHOLOGY

CUTANEOUS ROSAI-DORFMAN DISEASE:A REPORT OF ELEVEN CASES FROM CHINA AND A REVIEW OF THE LITERATURE

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Background: Rosai-Dorfman disease is a rare benign proliferative disorder of histiocytes, which is characterized by overproduction and accumulation of histiocytes within lymph node sinuses and many other extranodal sites, including skin, oral ,respiratory tract,etc. Cutaneous Rosai-Dorfman disease (CRDD) is rare and sometimes is misdiagnosed as Kimura's disease or pseudolymphoma.

Observation: We analyzed 11 cases diagnosed as CRDD from our department to explore their clinical pathological features of. Of our 11 cases, including 6 male and 5 female, the lesions presented with red plaques, papules or nodules affected face, head, trunk and arms. Histopathological examination of the skin lesions showed that all 11 cases demonstrated diffused mixed infiltration of lymphocytes, histocytes and sparse plasmocytes. Ten cases showed diagnostic feature of emperipolesis with histiocytes phagocytosed inflammatory cells. Two cases demonstrated mild eosinophilic granulocytes infiltration. Positive immunohistological staining for S100 and CD68 and negative for CD1a and Langerin confirmed the diagnosis of CRDD.

Key message:CRDD is rare and difficult to diagnose only based on clinical features,.Histopathological findings of S100 positive histocytes and emperipolesis support the correct diagnosis of CRDD. Differential diagnosis should be identified with infectious granuloma, reticulo-histiocytosis, langerhans cell histiocytosis and malignant histiocytosis. Though CRDD usually develops slowly but has good prognosis. Long-term follow-up is necessary to avoid further system damage.