Poster ID 36

Cutaneous Rosai-Dorfman disease: A rare histiocytic disorder

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Introduction

- Rosai-Dorfman Disease (RDD) is also known as sinus histiocytosis with massive lymphadenopathy.
- Benign multi-organ non-Langerhans cell histiocytic proliferation.
- Most common site is lymph nodes; purely cutaneous RDD is rare (3% of cases).

Discussion

- Classic cutaneous RDD presents as firm red to red-brown or xanthomatous macules, papules, nodules or plaques.
- Clinical lesions of RDD are relatively non-specific, & other histiocytoses, infectious processes, sarcoidosis, & infiltrative disorders are the differential diagnosis.
- Cutaneous histopathology may be very non-specific, and immunohistochemistry might help.
- Disease may be self-limited due to spontaneous resolution.
- In persistent disease- surgical excision, corticosteroids, thalidomide, cryosurgery, radiotherapy, dapsone, isotretinoin, interferon α, acitretin, and pulsed dye laser have been used.

Conclusion

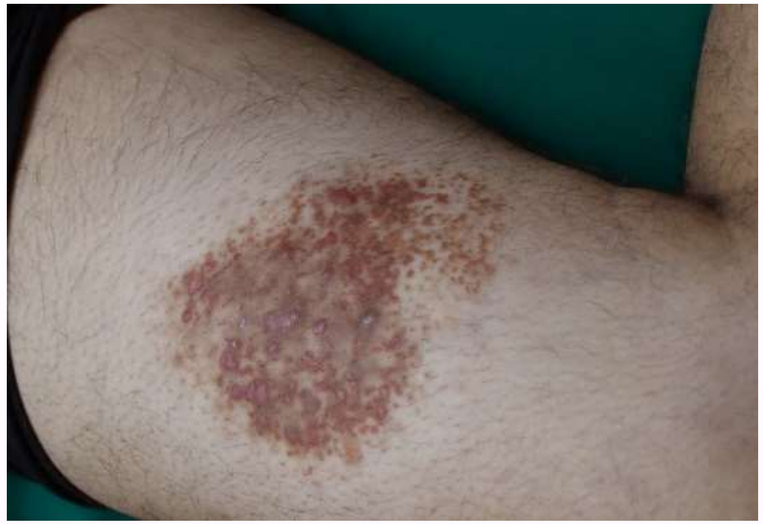
- This rare histiocytic disorder, purely cutaneous RDD, is characterized by non-specific clinical features, but more specific histopathology and immunohistochemistry findings.

References

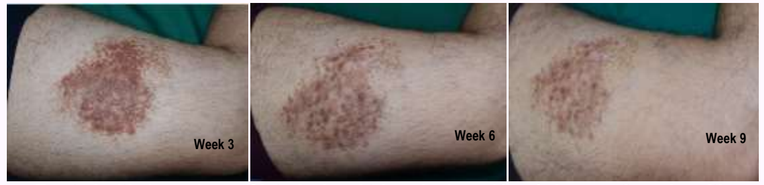
- Ohnishi A, Yoshida Y, Yamamoto O. Cutaneous Rosai-Dorfman disease. J Dermatol 2008;35: 604-5.
- Lu CI, Kuo TT, Wong WR, Hong HS. Clinical and histopathologic spectrum of cutaneous Rosai-Dorfman disease in Taiwan. J Am Acad Dermatol 2004;51:931-9.
- Cook-Norris RH, Hayes BB, Robb C, Boyd AS, Zic JA. Purely cutaneous Rosai-Dorfman disease. Int J Dermatol 2009;48:439-40.

Case Report

Clinical



- A 33-year old man presented with an asymptomatic, erythematous to hyperpigmented, gradually progressive, painless plaque on his right thigh for four months.
- No lymphadenopathy, fever or weight loss.
- On examination, there was an ill-defined, light-brown to skin colored swelling of size 10 X 12 cm over medial aspect of his thigh with overlying multiple, closely-aggregated, dark-brown hyperpigmented papules and nodules

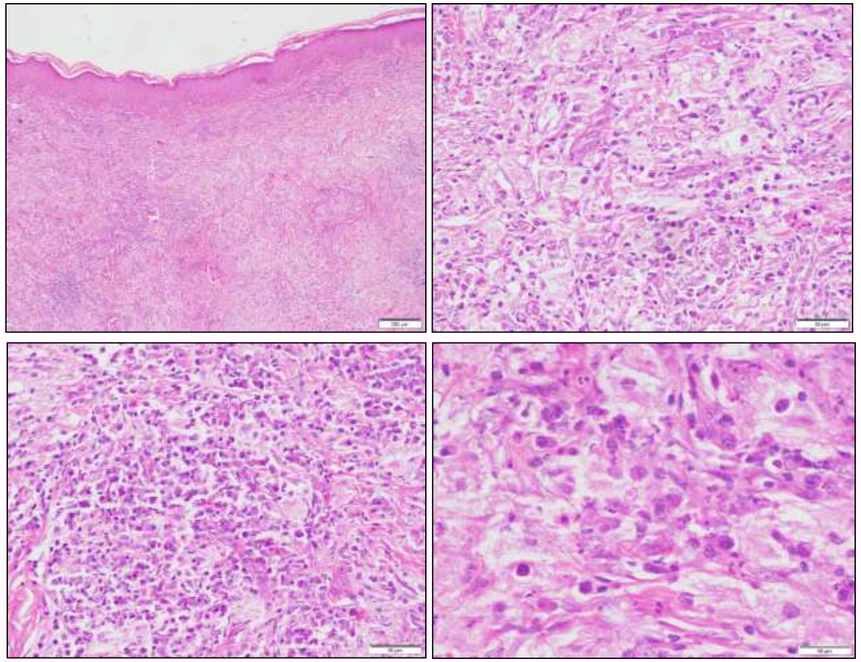


Treated with 4 sessions of intralesional triamcinolone 10mg/ml, every 3 weeks with partial (75%) improvement

Laboratory Investigations

- CBC, LFT, RFT- normal
- Chest X-ray - normal
- MRI- Ill-defined homogenously enhancing subcutaneous swelling

Histopathology



Dense nodular collections of foamy histiocytes, lymphocytes, histiocytes, plasma cells, neutrophils and few eosinophils in the dermis, with evidence of emperipolesis