Case 2
A 49-year-old Thai woman.
Chief complaint: multiple asymptomatic rash at face, trunk, and proximal extremities for 3 months.

Present illness

Three months earlier, she had symptoms of eye redness and irritation. She visited the ophthalmologist and was diagnosed with bilateral anterior granulomatous uveitis. Due to the granulomatous nature mycobacterium tuberculosis was suspected.

A few days after initiating antituberculosis drugs, she developed multiple asymptomatic erythematous papules on face, trunk and proximal extremities and was sent to dermatologic clinic for evaluation. She denied fever, systemic symptoms, or previous similar skin lesions.

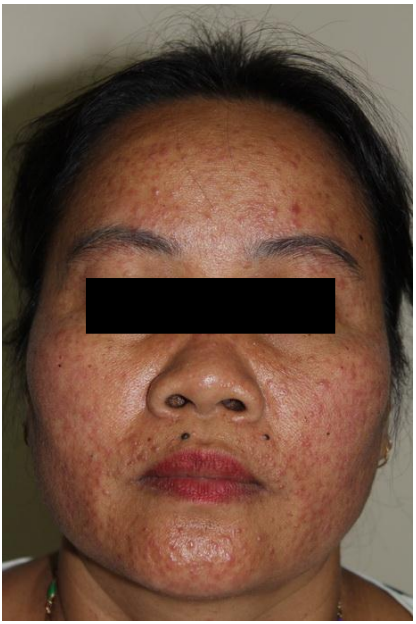
Past history
Underlying type 2 diabetes mellitus, hypertension, dyslipidemia.
She denied history of previously contacted TB.

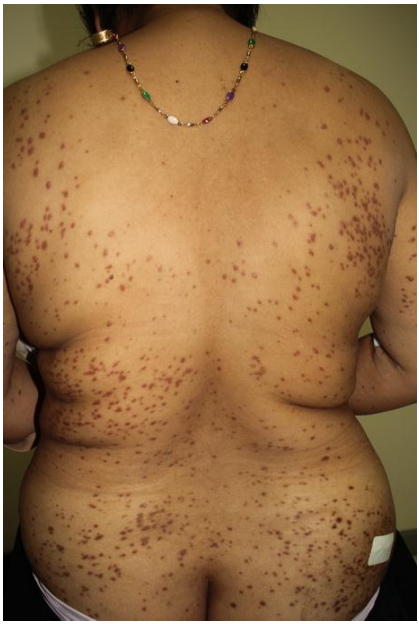
Family history: nil

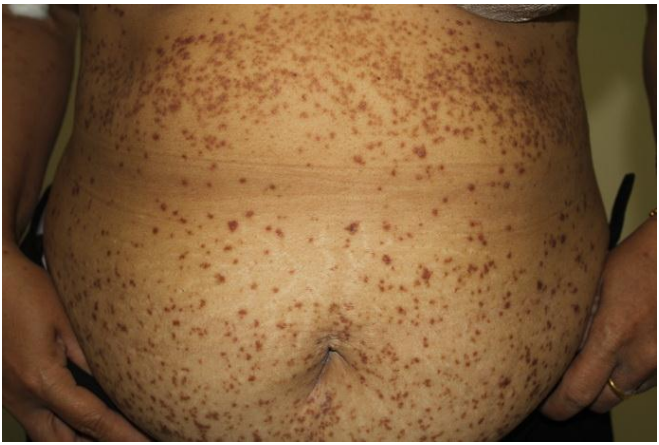
Physical examination
HEENT: not pale, no jaundice, no conjunctival injection
Lymph node: cannot be palpated in all areas
Heart and lung: normal
Abdomen: no hepatosplenomegaly, no mass
Rt buttock: Illed-defined mass size 5*5 cm.
Neurological examination: no deficit
Otolaryngologic examination: waiting for evaluation

Skin examination

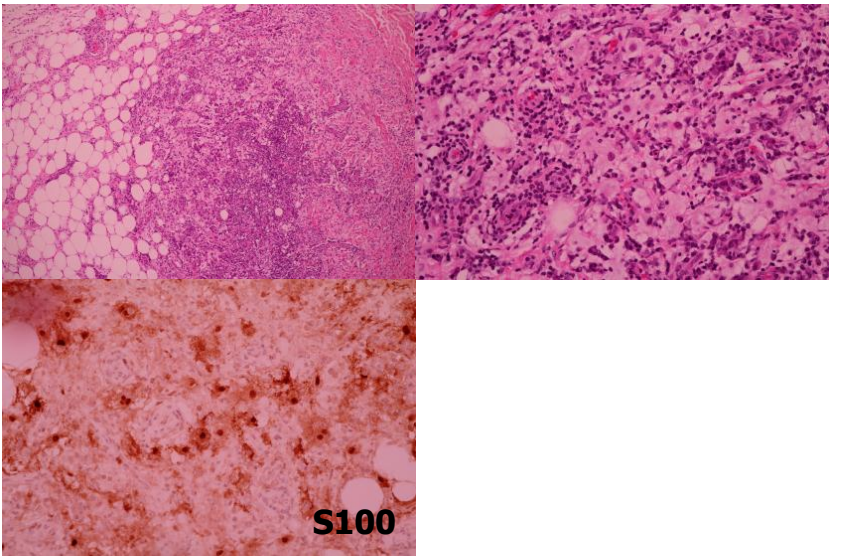
multiple discrete erythematous to yellowish firm dome-shaped papules and nodules, no comedone, no pustule distribute at face, trunk, back and both upper extremities







Histopathology
S12-25659
• Dense nodular infiltrate of large histiocytes admixed with lymphocytes, neutrophils and plasma cells in the upper dermis
S13-004329A
• Dense nodular infiltrate of large foamy histiocytes, lymphocytes and numerous plasma cells in deep dermis and subcutaneous tissue
• Presence of intact lymphocytes and plasma cells within the cytoplasm large foamy histiocytes (emperipolesis)
• The histiocytes express CD68 and S100 but negative for CD1a



Investigation:
CBC: Hb 13.2 g/dL, Hct 43.92%
WBC 11,570/mm^3 (N 79%, L 15%, M 5%, E 1%)
Platelets 299,000/mm^3
LFT: AST 18 U/L ALT 23 U/L
Chol 162 mg/dL ,Trig 81 mg/dL , HbA1C 7.46

Serum electropheresis : normal
Urine analysis : normal

CT chest: no evidence of pulmonary TB, three tiny nodules (2-3 mm.) in RUL and LLL, too small to characterized causative pathology. No mediastinal lymph node enlargement.
Serum electrophoresis: normal
Ultrasonogram abdomen: normal size and mild increased parenchymal echo liver, no mass, normal size of spleen
MRI hip: A large infiltrative enhancing iso-to intermediate signal T1/hyposignal T2 lesion at the subcutaneous fat of the posterolateral aspect of the right gluteal region, measuring about 6.5*8.9 cm.

Diagnosis: Extranodal Rosai-Dorfman disease
DM, HT, DLP
Treatment: oral isotretinoin 20 mg/day
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Consultant: Kumutnart Chanprapaph, M.D.

Discussion

Rosai-Dorfman disease (RDD), is a non-Langerhans cell histiocytosis. It is first described by Rosai and Dorfman in 1969.[1] It is also called sinus histiocytosis with massive lymphadenopathy(SHML). It is a benign self limiting disease with uncertain etiology. Formerly, RDD is an disorder of the lymph nodes that could involve extranodal sites. However pure cutaneous RDD is increasingly reported independently.[2] This raise the possibility of a distinct entity diease. The cutaneous manifestations are variable. Papulonodular lesions are the most common presentation.[3] Also indurated plaque, tumor, subcutaneous nodule, plaque with satellite papules, and central ulceration have been reported.[4] The most common site of involvement are the limbs followed by the trunk(

back/chest/flank) and face in descending frequency.[3] Variable clinical and indistinct presentation could lead to delay in diagnosis.

Up until now, extranodal and non-cutaneous RDD, which include oral cavity, eye/orbit/ocular, salivary gland, breast, soft tisuue, respiratory tract, bone, genitourinary system and gastrointestinal tract, have been documented.[5-10]

Summarized data from 6 large case series with cutaneous RDD (N=112)[2-4, 11-13] showed that there was slightly female predominate with mean age in the 4th decade(46.25yrs). Multiple site of involvement have been reported in the range of 24-59%. Uveitis was the most frequent extracutaneous involvement (0-14.8%), similar to the finding in this case. Furthermore tuberculosis, lymphoma, systemic lupus erythematosus were reported as associated diseases.

The clinical diagnosis is therefore difficult and relies on histologic findings. The most common characteristic and consistent histologic finding is dense dermal mixed inflammatory infiltrate composing of histiocytes, plasma cells, lymphocytes, and neutrophils. Phagocytosis of inflammatory cells into the cytoplasm of histiocytes, a process called emperipolesis can be highlighted by S-100 protein staining. Further special stains displaying positive response of the histiocytes for CD68 and negative CD 1a exclude LCH.[3]

Treatment depends on clinical manifestation. No standard approach to the treatment of cutaneous RDD has been developed. In many cases the lesions remain asymptomatic and regress spontaneously(range 14 mo-55 mo)[3], however the condition can be chronic and recurrence. Various therapeutic modalities have been approached with different outcomes. The most effective treatment with lowest rate of recurrence was excision. While other treatments such as oral corticosteroid, retinoic acid, thalidomide, dapsone and vibramycin offered incomplete cure.

Due to underlying diabetes mellitus and multiple widespread lesions, we selected oral isotretinoin and careful lipid monitoring for this patient.

References

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