CASE 22

Patient: A 58-year-old Thai female

Chief Complaint: Multiple asymptomatic papules on the face for 2 months

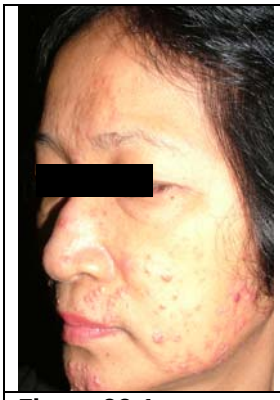
Present Illness: She presented with 2-month history of multiple asymptomatic papules initially existing along the nose. Biopsy was performed and revealed granulation tissue suggesting rupture folliculitis. Topical metronidazole along with isotretenoin 10 mg/day was commenced for 2 months without improvement. The lesions then gradually progressed to involve the entire face and extremities, cosmetically unacceptable to the patient. She was otherwise healthy with no history of fever, weight loss or fatigue.

Past History: She was previously healthy and not taking any medication.

Family History: nil

Physical Examination:
VS: T 37 ^{0}C, RR 20/min, BP 112/67 mmHg, HR 80/min
GA: good consciousness, not pale, no jaundice
Abdomen: no hepatosplenomegaly
LN: not palpable
Ophthalmologic and otolaryngologic examination were normal

Dermatological Examination (Figure 22.1-3)**:** multiple discrete erythematous to yellowish firm dome-shaped papules and nodules on the face predominantly along periorbital and perioral areas and few scattered papules appeared on the upper extremities.



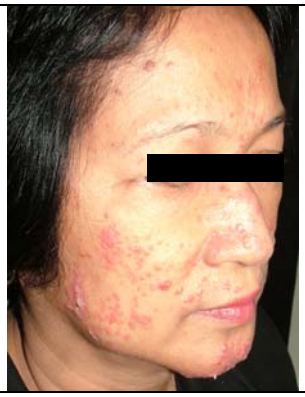


Figure 22.1

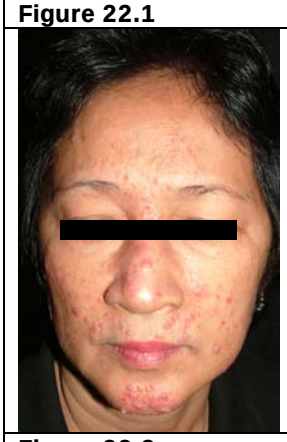


Figure 22.2

Figure 22.3

Investigations:

CBC: Hb 13 g/dL, Hct 40.1%
WBC 7,000/mm^3 (N 61%, L 31%, M 7%, E 1%)
Platelets 230,000/mm^3
ESR 12 mm/hr

LFT: AST14 U/L ALT 38 U/L

Lipid: Chol 285 mg/dL,Triglyceride 186 mg/dL

EBV titer: negative

Chest X ray: no mediastinal mass, no infiltration

Serum electropheresis: normal
Urine analysis: normal
Antinuclear antibody: pending

Histopathology (S09- 7515, S09- 8129) (Figure 22.4-6):

- Diffuse inflammatory-cell infiltrate of lymphocytes, histiocytes neutrophils and plasma cells in the dermis
- Large histiocytes with pale abundant cytoplasm and some phagocytosed showing emperiopolesis of lymphocytes

Immunohistochemistry (Figure 22.7-9):

- Positive S100 and CD68
- Negative CD1a

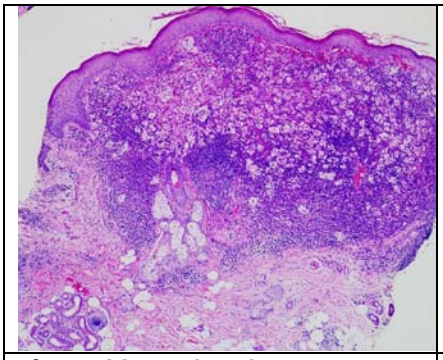


Figure 22.4, H&E 40x

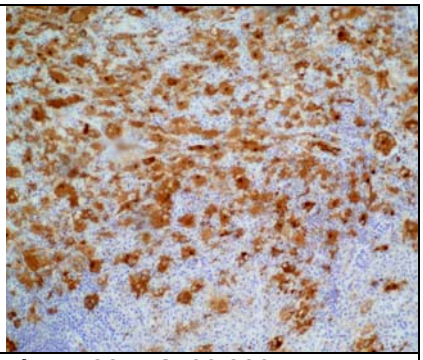


Figure 22.7, S100 200x

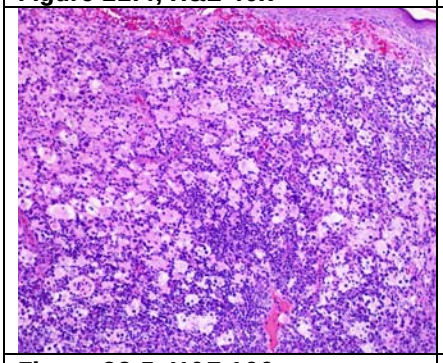


Figure 22.5, H&E 100x

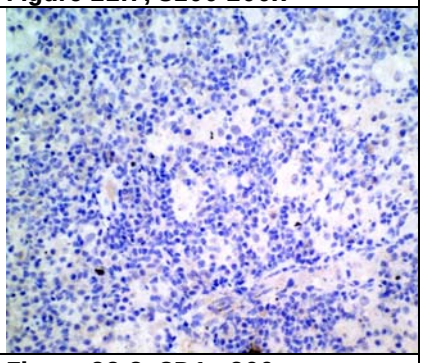


Figure 22.8, CD1a 200x

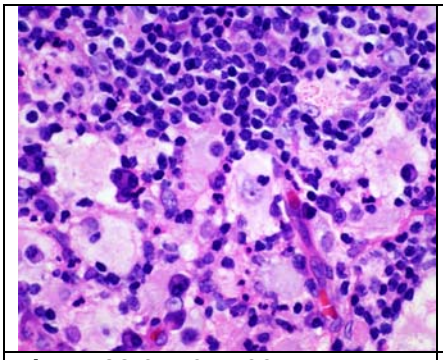


Figure 22.6, H&E 400x

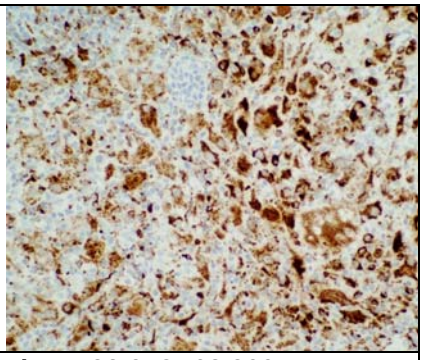


Figure 22.9, CD68 200x

Diagnosis: Cutaneous Rosai-Dorfman disease

Treatment: Prednisolone 60 mg/d then tapering to 10 mg/d

Carbon dioxide laser

Presenter: Silada Kanokrungsee

Consultant: Kumutnart Chanprapaph

Discussion:

Rosai-Dorfman disease (RDD), also called sinus histiocytosis with massive lymphadenopathy, is a non-Langerhans cell histiocytosis first described by Rosai and Dorfman in 1969.[1] It is a benign self limiting disease characterized by painless enlarged lymph nodes especially the cervical area caused by increase numbers of histiocytes within the lymph nodes sinus. Extranodal involvement occurs in approximately 43%, of which the skin is the most common site.[2] Purely cutaneous disease, a distinct clinical entity without involvement of lymph nodes or other organs is rare, accounting for only 3 % of reported RDD cases.[2] Clinically, cutaneous RDD can be divided into 3 main types: papulonodular type (79.5%), plaque type (12.8%), and tumor type (7.7%), the former being the appearance of our patient.[3] Most lesions are located on the

face, followed by the back, chest, thigh, flanks, and shoulder. Documents with demographic data and geographic distribution demonstrated that most cases of cutaneous RDD occur in Asians and middle- aged females are most frequently affected.[3-5]

Cutaneous RDD has variable clinical presentation indistinguishable from other non-Langerhans cells histiocytosis (LCH) such as xanthrogranuloma, reticulohistiocytosis, generalized eruptive histiocytosis and xanthoma. Moreover, cutaneous RDD can also mimic granulomatous rosacea, sarcoidosis and rupture folliculitis leading to diagnosis delay as in this case. The clinical diagnosis is therefore difficult and relies on histologic findings. The most 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 displayed positive response of the histiocytes for CD68 and negative CD 1a excluding LCH. [3,4]

Cutaneous RDD generally follows a benign clinical course but can be associated with other organ involvement such as bilateral uveitis, antinuclear antibody positive lupus erythematosus, glomerulonephritis, rheumatoid arthritis, hypothyroidism, lymphoma, leukemia and hypergammaglobulinemia.[4, 6] Uveal involvement being the most common association has no impact on disease prognosis but definitely increases morbidity. [4]

Spontaneous regression tends to occur over months to years regardless of different treatment, ranging from 6 to 55 months.[5] No standard approach to the treatment of cutaneous RDD has been developed. Surgical excisions are preferred for

solitary or localized lesions. Some patients have been reported to respond to radiotherapy, cryotherapy, and carbon dioxide laser therapy. [3,4,7] Several documents of topical and systemic therapy have been reported with variable response including intralesional, topical and systemic corticosteroid, alkylating agents, thalidomide, retinoid and imatinib. [8,9,10-12] Regarding the original diagnosis of extensive rupture folliculitis, isotretinoin 10mg/d was administered for 2 months without improvement. Moreover, the patient developed significant elevation of LDL and triglyceride causing discontinuation of therapy. Our patient was successfully treated with systemic corticosteroid 1 mg/kg/d with prompt tapering to 10 mg/d in 3 months. Few perioral papules were left and removed with carbon dioxide laser therapy. Remaining facial erythematous dusk red patches were still cosmetically disturbing to the patient, hence, pulse dye laser therapy was applied monthly for 3 consecutive months showed partial improvement.

References

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