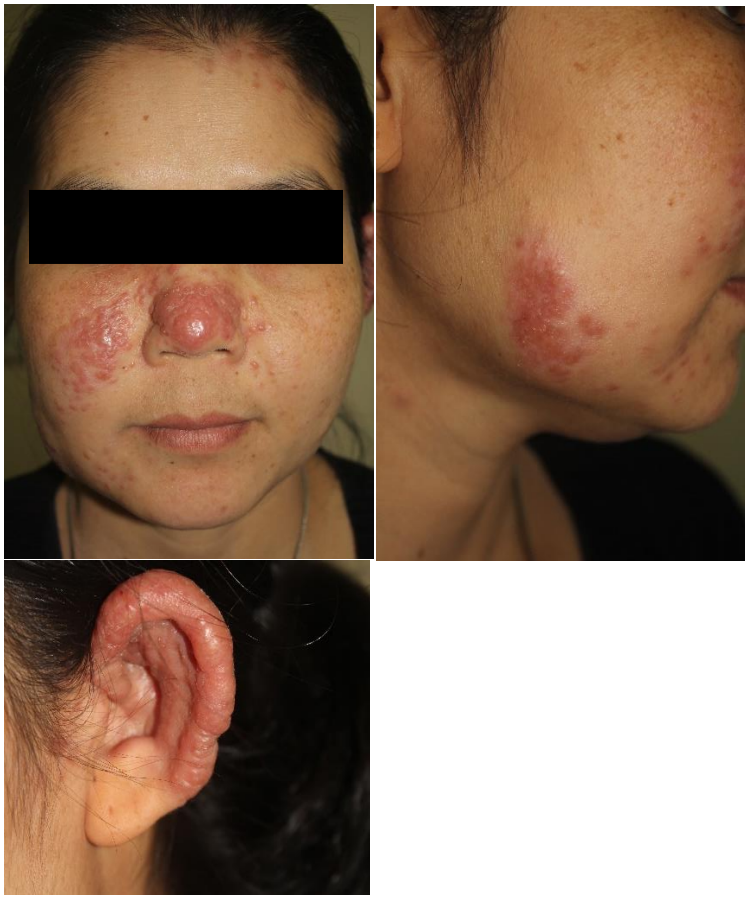
Case 16
A 34-year-old Thai woman from Sisaket province
Chief complaint: Rash on face and ear for 1 year.



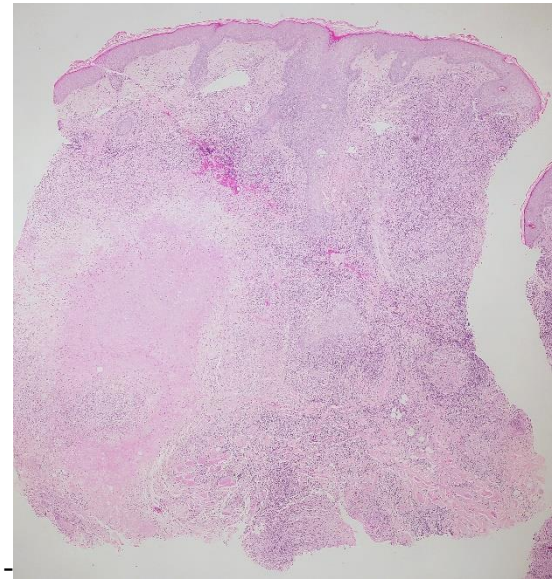
Present illness: The patient presented with a 1-year history of skin color to red-brown papules, which some lesions developed into pustules, on central face, left ear and right lower cheek area. Mild itching and tenderness was observed. She was otherwise well, and denied fever or significant weight loss. She denied history of cosmetic treatment or injection on her face.

Past history
She had *Mycobacterium abscessus* infection, presented with lymphadenopathy, which was completely treated for 3 years. Her underlying disease is thalassemia trait Hb AE

Physical examination
General appearance: no anemia, no icteric sclera
Lymph node: axillary and supraclavicular lymph node cannot be palpated
Lung: equal breath sound, no adventitious sound
Abdomen: liver and spleen cannot be palpated

Skin examination
-Multiple red-brown papules coalescing to form plaques on nose, right lower cheek and left ear pinna.
-No telangiectasia or erythematous background on face

Histopathology (S15-019478, face)
- Dense diffuse inflammatory cells infiltrate of lymphocytes admixed with histiocytes and a few multinucleated giant cells
- Large area of caseous necrosis within the infiltrate
- Microscopic diagnosis: Tuberculoid granulomatous dermatitis with caseous necrosis



Investigation:
Special stain - GMS, Brown&Benn, PAS, Fite – failed to demonstrate the organism
Tissue culture (aerobe, mycobacteria, fungus) – negative
Tissue PCR for mycobacteria – negative

Chest X-ray – normal

Diagnosis: Lupus miliaris disseminatus faciei

Treatment: Single dose of intramuscular triamcinolone acetonide 40mg, oral isotretinoin 20 mg per day

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Discussion:

Lupus miliaris disseminatus faciei (LMDF), also known as acne agminata[1], acnitis, micropapular tuberculid[2], FIGURE[3], is a rare dermatosis with characteristic clinicopathologic features. First described by Fox et al in 1878[4]. The condition is most often seen in young adults of both sexes between the second and the fourth decade of life. The precise etiology of LMDF is still currently unknown. Previously it is believed to be a "tuberculid", reaction from Mycobacterium tuberculosis infection, yet most studied have failed to demonstrate the organism by several methods, and most of antituberculosis treatment did not improved the cutaneous eruptions. Therefore, the tuberculosis origin of LMDF is no longer accepted[5]. On contrary, due to the granulomatous infiltrate in histology of LMDF which is associated with pilosebaceous unit, it is speculated that the granulomatous reaction to hair follicle destruction or ruptured epidermal cyst may be a pathogenesis of the disease[6].

The usual clinical features include multiple, discrete, smooth 1-3 mm red brown or brown to yellowish dome shape papules. They are usually distributed over the central and lateral side of face, infrequently extending to involve the neck, eyebrows and eyelids[5]. Extrafacial areas can be rarely affected, including axilla[7], neck, scalp, leg, trunk and genitalia. Interestingly, to our knowledge, our patient is the first case that has involvement of the ear.

Histopathologically, LMDF in fully developed lesion is characterized by a dermal granulomatous infiltrate, epithelioid cell granuloma with central necrosis which was the hallmark of this disease[8].

The differential diagnosis in this patient is shown in Table 1[9]. The infectious causes are completely excluded in our patient by special stains and tissue culture results.

Table1.

	LMDF / Acne acminata	Granulomatous rosacea	Sarcoidosis
Age	Younger age		
Clinical	No background erythema or telangiectasias Extra facial	Rare extrafacial Spare eyelid/ upper lip	Multi-system involve
Suggested causes	Granulomatous reaction to hair follicle destruction	Multifactorial	Unknown
Histopathology	Round granulomas with central caseation necrosis	Sarcoidal granuloma	Granulomatous infiltrate without necrosis
Prognosis	Tend to spontaneously resolve in 1 to 3 years result in permanent scarring	More chronic course than LMDF contain Demodex mites	Remission often in 6 mo.
Comment	Extrafacial involement may occur more resistant to treatment		Serum ACE Systemic involvement

The natural history of LMDF is one of gradual, but spontaneous regression in 2-4 years leaving pock-like scars is the main course. However, various treatment modalities have been tried including systemic and topical corticosteroids[10], oral tetracycline and isotretinoin. Dapsone was reported an excellent response in treatment of the patient defined by complete clearance, no scar and relapse after follow up 1 year[11]. Because of underlying hemoglobinopathy of this patient, dapsone is not suitable for her treatment. Therefore we decided to use oral isotretinoin 20 mg per day and systemic corticosteroid 40 mg once a month. Unfortunately, the patient lost to follow up after started the treatment for 1 month. We attempted to contact her by telephone call. She reported moderately improvement with continue treatment with this regimen.

References

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