A 56-year-old woman with a 6-month, pruritic, tender facial rash sought clinical evaluation and treatment. She had previously been treated with oral minocycline and showed no signs of improvement. Physical exam revealed many hyperpigmented and flesh-colored papules and nodules, some of which were located on the upper cheeks, nasal bridge, medial canthus, and lower cheeks and had central excoriation and umbilication. She had no flushing or telangiectasias (Figs 1 and 2). The patient had no systemic symptoms and did not take any medications. A punch biopsy was taken and sent for routine pathology (Fig 3).
Question 1. What is the most likely diagnosis?

A. Cystic hormonal acne
B. Eruptive syringomas
C. Lupus miliaris disseminatus faciei
D. Lupus vulgaris
E. Sarcoidosis

Answers:

A. Cystic hormonal acne — Incorrect. Acne is a multifactorial disorder of the pilosebaceous unit clinically characterized by comedones, papules, pustules, cysts, and sometimes scarring. Hormonal effects on sebum secretion are key to the pathogenesis of acne. Cystic acne lesions are deeper and filled with a combination of pus and serosanguineous fluid. Biopsy demonstrates an inflamed hair follicle dilated by keratin, sebaceous, and bacterial debris.

B. Eruptive syringomas — Incorrect. Syringomas are asymptomatic benign adnexal tumors that present as multiple discrete, flesh-colored papules, 2-4 mm in diameter. They are usually more common on the lower lid. They likely arise from luminal cells of eccrine sweat ducts. In eruptive syringomas, multiple lesions are apparent in childhood or early adulthood on the neck, chest, shoulders, abdomen, and pubic area.

C. Lupus miliaris disseminatus faciei — Correct. Lupus miliaris disseminatus faciei (LMDF) is an uncommon granulomatous inflammatory disease that typically affects the central face and eyelids but can rarely affect extrafacial sites, such as the axillae and genitalia. It has a sudden onset with spontaneous resolution in 1-4 years. LMDF presents as yellow-brown, dome-shaped papules and nodules with an apple-jelly-like color on diascopy. Lesions heal with scarring. Biopsy demonstrates epithelioid granulomas with central caseous necrosis.

D. Lupus vulgaris — Incorrect. Lupus vulgaris is a form of cutaneous tuberculosis that occurs in previously sensitized hosts that presents with an asymptomatic gelatinous reddish-brown plaque on the face or neck. A brown-yellow (apple-jelly-like) color can be seen on diascopy. On histology, granulomas with central caseation necrosis and dense collections of peripheral lymphocytes are apparent.

E. Sarcoidosis — Incorrect. Sarcoidosis is a multisystem inflammatory disease characterized by non-caseating granulomas of unknown etiology. Approximately 25% of patients will have cutaneous involvement. Sarcoidosis presents with asymptomatic nonscaly, skin-colored to red-brown infiltrated papules and plaques on face, lips, neck, trunk, or extremities. On histology, epithelioid histiocytes aggregate to form naked granulomas with minimal surrounding lymphocytic inflammation and no necrosis.

Question 2. What is the recommended treatment?

A. Isotretinoin
B. Surgical excision
C. Topical antifungal
D. Topical steroids
E. Topical tretinoin

Answers:

A. Isotretinoin — Correct. Successful treatment with isotretinoin has been documented in case reports for LMDF. Doses used ranged from 0.4-1 mg/kg/day. Al Mutari et al reported success with oral isotretinoin therapy in the treatment of LMDF. Schaarschmidt et al subsequently reported a case in which complete clearance of facial lesions was achieved with oral isotretinoin therapy at a dose of 0.4 mg/kg/day but no improvement in axillary lesions. In the case presented here, the patient reported 80% clearance after 2 months of isotretinoin therapy with no side effects reported. Other reported treatment options that have been used with variable success include intralesional or systemic corticosteroids, topical tacrolimus, tetracyclines, metronidazole, erythromycin, and dapsone.

B. Surgical excision — Incorrect. Surgical excision is not recommended as initial therapy for LMDF. Typically, multiple lesions are present at one time making surgical excision impractical in addition to the risk of scarring from the procedure. However, there are reports of success with laser therapy with a 1450-nm diode laser used in 3 monthly treatments.

C. Topical antifungal — Incorrect. Topical antifungal agents like ketoconazole or terbinafine are used to treat superficial fungal infections. These therapies would be ineffective in LMDF because there is no evidence that any fungal process is involved in the pathogenesis of this disorder.
D. Topical steroids — Incorrect. The efficacy of treatment is difficult to assess in LMDF because it is a rare and self-limiting condition. There are no randomized controlled trial data, but case reports and case series show that topical steroids are ineffective. 

E. Topical tretinoin — Incorrect. Topical tretinoin would be an appropriate first-line therapy for acne but would be ineffective as treatment for LMDF.

**Question 3. What would one expect to see in a biopsy from this patient?**

A. Dermal granulomatous reaction with central caseous necrosis — Correct. LMDF exhibits a small pea-like palisaded granuloma with central caseous necrosis and occasionally degenerated hair follicles on pathology. Although the disease resembles miliary tuberculosis on histology, the two can be distinguished by the acid-fast staining, with cases of LMDF showing negative results. In addition, if one is concerned about tuberculosis, an interferon-γ release assay or tuberculin skin test can be used to evaluate for tuberculosis. Histochemical stains, culturing for bacilli, and PCR for *Mycobacterium tuberculosis* DNA are other methods that can be used to work up a suspected case of tuberculosis infection.

B. Dilated hair follicles containing keratin, sebaceous, and bacterial debris — Incorrect. This would be the expected histology of an acne lesion.

C. Noncaseating naked granulomas — Incorrect. Noncaseating naked granulomas are characteristic of sarcoidosis. The lack of necrosis helps to distinguish this entity from LMDF. In addition to the histologic differences between these diagnoses, additional studies such as chest x-ray, serum calcium, and serum angiotensin-converting enzyme levels are helpful in further excluding sarcoidosis.

D. Paisley-tie pattern of tadpole-shaped ducts — Incorrect. This histologic description is consistent with a syringoma. Although syringomas can be found in a similar distribution around the eyelids as LMDF, they are easily distinguished on pathology.

E. Suppurative granulomas containing Kinyoun-positive bacilli — Incorrect. Lupus vulgaris typically demonstrates a granulomatous reaction with central necrosis and mantle of lymphocytes. Although similar to LMDF histologically, an interferon-γ release assay or tuberculin skin test can be used to evaluate for tuberculosis.

**Answers:**

A. Dermal granulomatous reaction with central caseous necrosis

B. Dilated hair follicles containing keratin, sebaceous, and bacterial debris

C. Noncaseating naked granulomas

D. Paisley-tie pattern of tadpole-shaped ducts

E. Suppurative granulomas containing Kinyoun-positive bacilli

Abbreviation used:

**LMDF:** Lupus miliaris disseminatus faciei

REFERENCES

1. Al-Mutairi N. Nosology and therapeutic options for lupus miliaris disseminatus faciei. *J Dermatol*. 2011 Sep;38(9):864-873.

2. Rocas D, Kanitakis J. Lupus miliaris disseminatus faciei: report of a new case and brief literature review. *Dermatol Online J*. 2013;19(3):4.

3. Schaarschmidt ML, Schlich M, Schmieder A, Goerdt S, Peitsch WK. Lupus miliaris disseminatus faciei: not only a facial dermatosis. *Acta Derm Venereol*. 2017.

4. Shimizu A, Funasaka Y, Ueno T, Kanzaki A, Saeki H. Case of lupus miliaris disseminatus faciei associated with marked formation of cysts, successfully treated with intralesional injections of triamcinolone acetonide. *J Dermatol*. 2017;44(3):e164-e165.

5. Jih MH, Friedman PM, Kimyai-Asadi A, et al. Lupus miliaris disseminatus faciei: treatment with the 1450-nm diode laser. *Arch Dermatol*. 2005;141(2):143-145.