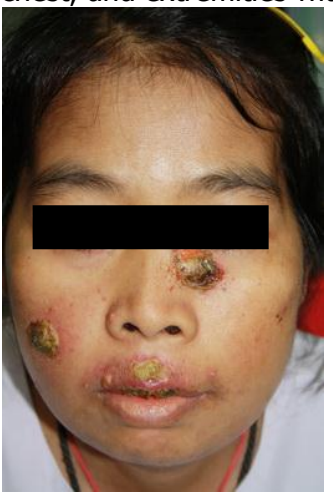
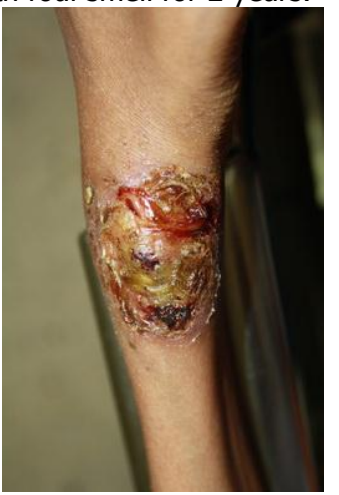
Case 4

A 34-year-old woman from Surin.

Chief complaint: Multiple ulcerated plaques with yellowish crusts on face, chest, and extremities with foul smell for 2 years.





Present illness: Two years previously, the patient noticed small lump on her chest. Then more lesions came up on the right lower leg, face and intranasal. The lesions later ulcerated with yellowish discharge and foul smell, some healed with atrophic scars. She also had low grade fever, low back pain, fatique, decrease appetite with weight loss 20kg in two years.

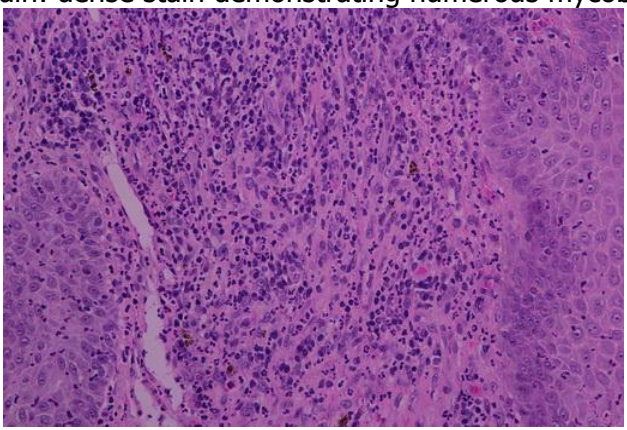
Past history: Four years previously, she was diagnosed SLE with AIHA from the other hospital and was on prednisolone 20mg/day. However, further evaluations at Ramathibodi hospital revealed no evidence of systemic lupus erythematosus, so prednisolone was tapered off.

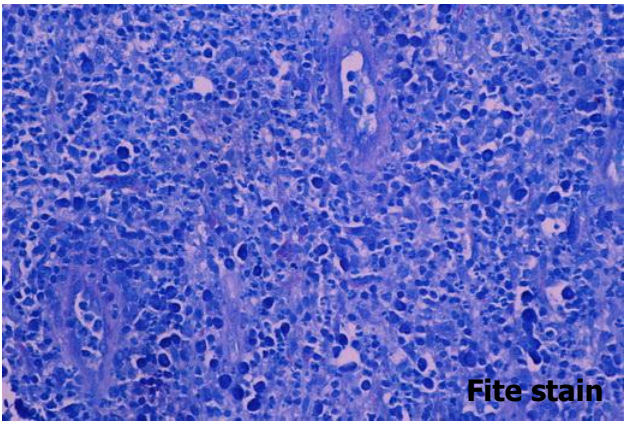
Family history: Her elder sister has systemic lupus erythematosus

Physical examination: Multiple ulcerated plaques with yellowish crusts and purulent discharge on the right cheek, the left cheek, upper chest wall, the right forearm, and the right lower leg which had foul smell

Rhinoscopy: irregular surface of right nasal septum with yellowish crusts
Lungs: occasional wheezing
Abdomen: no hepatosplenomegaly
Investigations: CBC : Hb 11g/dL, Hct 37.8%, WC 25,700 (N92%, L5%, M2%, E1%), Plt 448,000
CXR : non-specific reticular infiltrate both lungs predominant on left side
Spine X-ray : compression fracture at L3 vertebrae
Wound pus : AFB positive 2+
Tissue C/S for mycobacteria: Scotochromogenic mycobacterium
Pus PCR for mycobacteria: *Mycobacterium szulgai*
Anti-HIV antibody: negative
Anti-interferon γ IgG autoantibody: positive high titer
Immunoglobulin level: normal
Lymphocyte panel: normal
Histopathology (S12-08354B: face)

- Dense dermal mixed inflammatory-cell infiltrate of histiocytes, multinucleated giant cells, lymphocytes and numerous plasma cells extending to subcutis.
- Mild epidermal hyperplasia with focal spongiosis.
- Fite stain: dense stain demonstrating numerous mycobacteria.





Diagnosis: Disseminated *Mycobacterial szulgai* infection (skin, lung, and bone)
Positive anti-interferon γ IgG autoantibody

Management: Clarithromycin 500mg oral twice a day
Ethambutol 800mg oral once a day
Rifampicin 450mg oral once a day

All the lesions healed with atrophic scars. Her lower back pain improved. Plan to continue treatment for 18 months.

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Discussion

Our patient presented with multiple ulcerated plaques with purulent discharge on face, chest, extremities and intranasal for two years with low grade fever, fatique, decreased appetite with weight loss 20 kg. The differential diagnosis in this patient should include; first, infections that run chronic course for example nontuberculous mycobacteria, *Mycobacterium tuberculosis*, deep fungal infection, nocardiasis and leishmaniasis; second, Wegener's granulomatosis; and third, lymphoma cutis. Tissue culture revealed

scotochromogenic mycobacterium and PCR identified species *Mycobacterium szulgai*. Moreover, the spine X-ray showed compression fracture of L2 vertebrae most likely from infection. From the clinical and all of the investigations the most likely diagnosis in our patient is disseminated *M. szulgai* infection with at least two organs involved.

M. szulgai is a slow-growing nontuberculous Mycobacterium that was first described in 1972 by Mark et al.[1] It produces a yellow-orange pigment: the organism is scotochromogenic when grown at 37°C, but photochromogenic at 25°C.[2] It has been recovered from environmental sources, including a snail, aquarium water, swimming pool water, and tropical fish. The environment is the suspected source of infection.[3] Pulmonary *M. szulgai* disease is the most common presentation with extrapulmonary disease restricted to patients with an impaired systemic immunity (e.g., receipt of immunosuppressive medication, HIV infection, interferon γ receptor deficiency or hematologic malignancy).[4, 5] So it is important to recognize the presence of some immunologic defects predisposing immunocompetent host to develop disseminated *M. szulgai* infection. Large case series from Thailand and Taiwan have proposed a syndrome of adult-onset immunodeficiency in HIV-uninfected adults who have disseminated mycobacterial infection (especially with rapid growing mycobacteria)and other opportunistic infections (e.g., *Cryptococcus neoformans*, *Histoplasma capsulatum*, *Penicillium marneffei*, disseminated salmonellosis, or severe varicella-zoster virus infection) with concomitant reactive dermatosis especially neutrophilic dermatosis.[6] The study showed that this syndrome is strongly associated with high titer neutralizing antibodies to interferon γ which in turn supporting the main role of interferon γ in controlling numerous pathogens.

Our patient had high IgG antibody titer to interferon γ, which explained her clinically immunocompromised status and features. To my knowledge this is the first case of disseminated

M. szulgai infection associated with high titer of anti-interferon γ antibody. After treatment with combination anti tuberculous drugs of rifampin, ethambutol and clarithromycin, all the lesions regressed and healed with atrophic scars along with the improvement of systemic symptoms and low back pain.

References

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