中文題目： 全身性紅斑狼瘡與全身性皮膚石灰沉著症－ 一病例報告

英文題目： Calcinosis cutis universalis in systemic lupus erythematous: a case report

作者：黃虹綾[1]、歐燦騰[2]、吳正欽[2]、蔡文展[2]、顏正賢[2]

服務單位：高雄醫學大學附設醫院內科部[1]、過敏免疫風濕內科[2]

Introduction

Calcinosis cutis is a rare disorder characterized by calcium deposition in skin and subcutaneous tissues. It had been well known as association with connective tissue diseases , such as systemic sclerosis, dermatomyositis, and overlap syndromes. It is a very rare entities in systemic lupus erythematosus (SLE) populatin. Only 38 cases reports were been proposed in English literature up to 2012. The etiology of SLE is dystrophic in nature and almost presented as calcinosis circumscripta , only few cases presented as calcinosis cutis univeralis.

Case report

A 37-year-old woman presented to our emergency department with progression of local ulcerations with purulent pus and chalky milk-like material from the wounds of left posterior thigh.

She had been diagnosed as systemic lupus erythematosus(SLE) 13 years ago, with characters as alopecia, malar rash, Raynaud's phenomenon, pulmonary hypertension, positive antinuclear antibodies, high titer of anti-dsDNA and anti-Ro antibodies. She kept regular medication control by oral prednisolone(the dose varied from 5~30mg per day, depending on her clinical condition), hydroxychloroquine and irbesartan. Four years later, Calcinosis cutis of her lower legs had developed and gradually spread into her buttocks, back, and four extremities. The laboratory revealed normal calcium, phosphorus, PTH and muscle enzyme level.

Later she received several times of surgical removal of the newly, isolated calcified mass of her buttock and four extremities. She also took diltiazem, bisphosphate, intravenous ceftriaxone, and intralesional corticosteroid injection for calcinosis control. But these therapies all failed to control the spread of calcinosis. From then on, she had experienced several times of cellulites of her left thigh, buttock, bilateral hands, and right foot with the isolated pathogens included salmonella, Streptoccocus epidemidis, Staphylococcus aureus, Staphylococcus agalactiae, and Escherichia coli respectively.

Thirteen years later, her skin lesions progressed more rapidly, especially in recent six months. Multiple various sized hard nodules and hyperpigmented skin lesions extended quickly ,then fused into a whole plate, which deposit on her anterior abdomen, bilateral forearms, both thighs, and legs and caused the limitation of joint

motion.

Figure 1: multiple various sized hard masses on her buttocks and some masses became erythematous and tender.

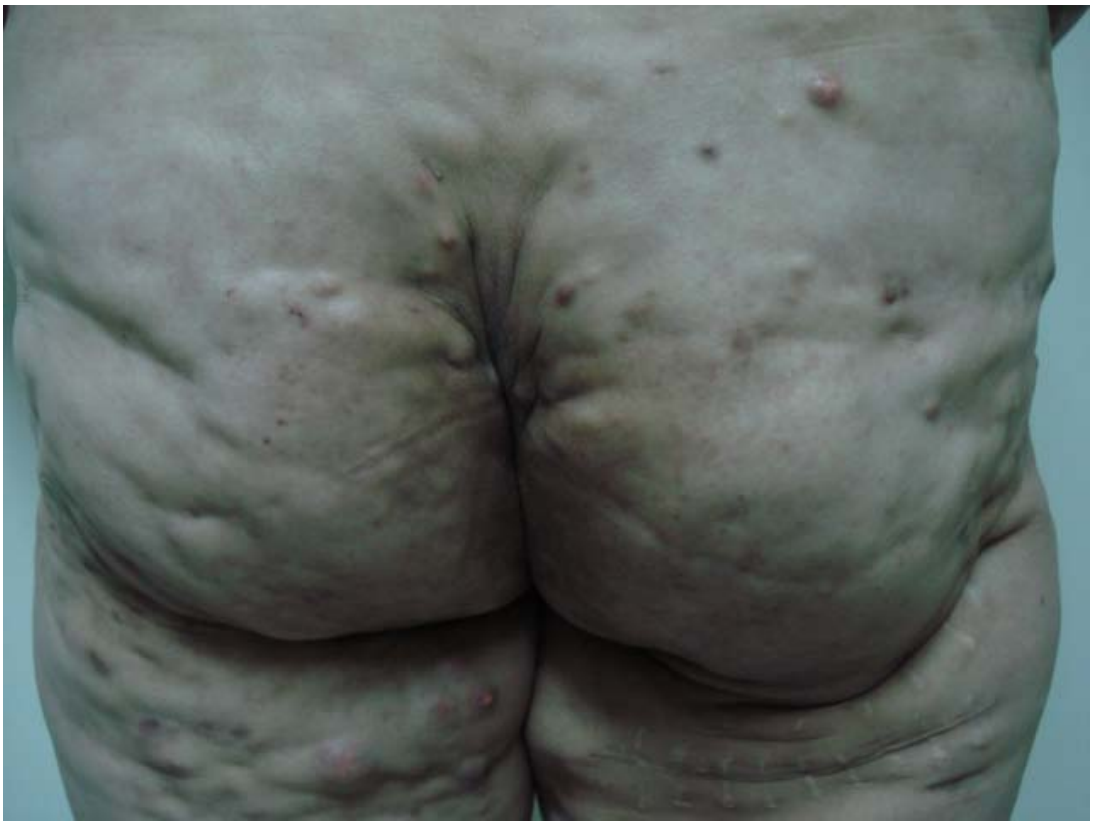


Figure 2: Multiple soft tissue calcifications around her buttock area and thighs.

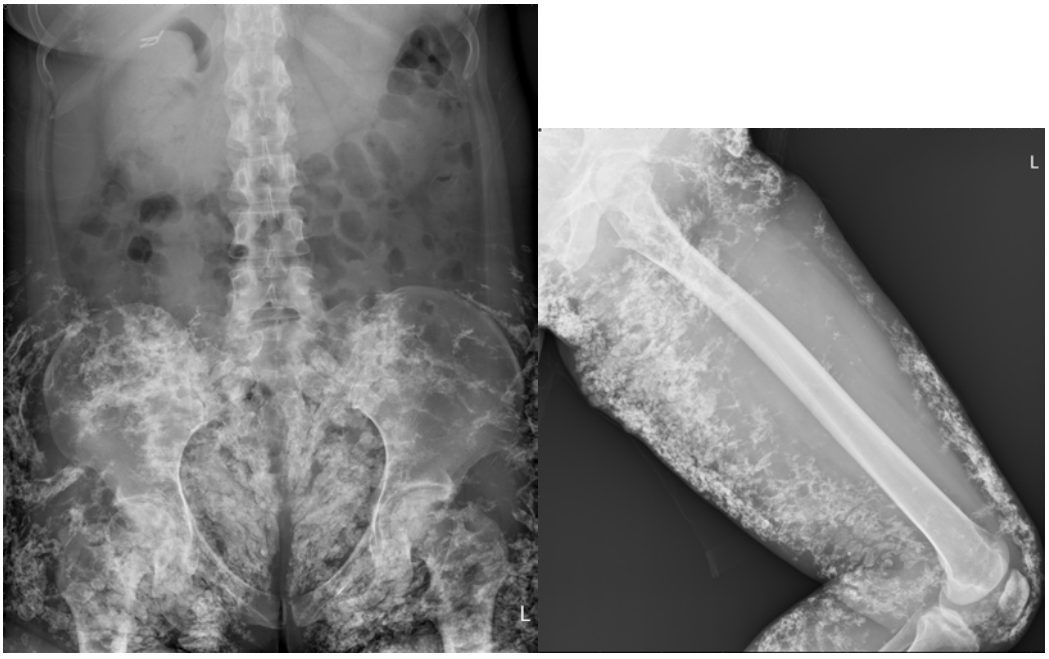


Figure 3: CT showed only subcutaneous tissue calcification without visceral

involvement.

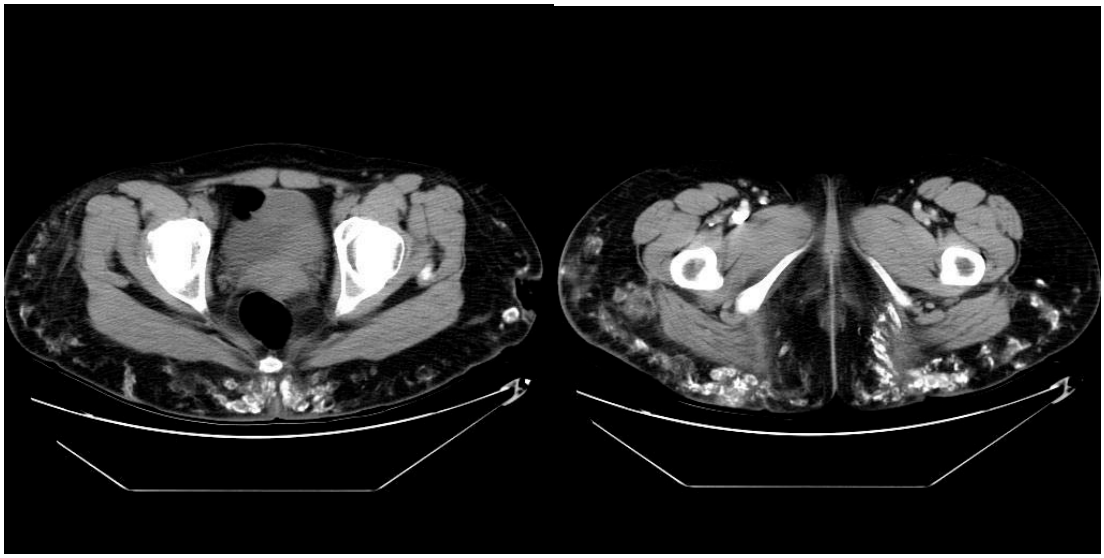
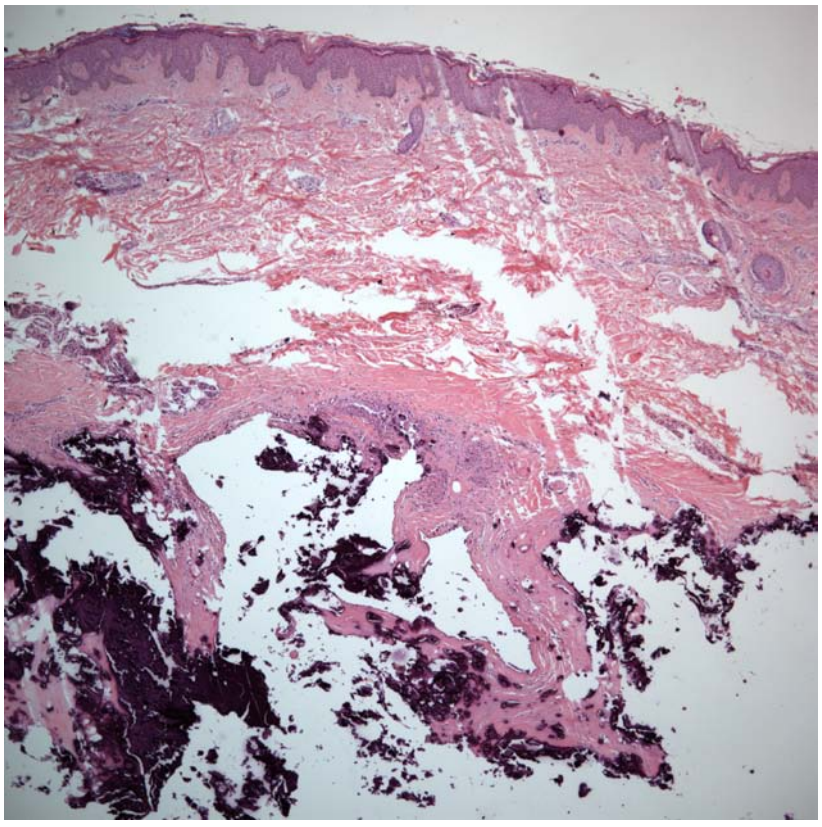


Figure 4: Skin biopsy revealed subcutaneous tissue calcification (HE stain, 40×)



Discussion

Calcinosis cutis is very rare in systemic lupus erythematous . We report this case with calcinosis cutis univeralis, who presented the first calcinosis cutis following 4 years of diagnosis. She received several different treatment, surgical excision for the newly formed localized lesion, it seems under temporarily control ,without newly calcification over the scar area. However, it still gets progression and became the

whole plaque with papule formation over other sites, especially over the buttock , four extremity extensor site and lower abdomen area even under systemic medication treatment ,she ever received bisphosphate 10mg/kg/day for 6 months, cefriaxone 2g/day for 30 days, Diltiazem 30mg/day for more than 1 year and intralesional corticosteroids .

Until now, there is no effective treatment for calcinosis cutis.