Chromoblastomycosis and disseminated superficial porokeratosis with secondary cutaneous amyloidosis: a case report

Sakchai Chaiyamahapurk MD, PhD,

Chaiyaporn Virochsangaroon **MD,**

Prateep Warnnisorn **MD, PhD.**

ABSTRACT:

CHAIYAMAHAPURK S, VIROCHSANGAROON **C,** WARNNISORN **P. CHROMOBLASTOMYCOSIS AND DISSEMINATED SUPERFICIAL POROKERATOSIS WITH SECONDARY CUTANEOUS AMYLOIDOSIS: A CASE REPORT. THAI J DERMATOL 2018; 34:**169-175.

FACULTY OF MEDICINE, NARESUAN UNIVERSITY, PHITSANULOK, THAILAND.

Chromoblastomycosis is an uncommon skin disease which could lead to delay diagnosis and disfiguring lesion. The patient has verrucous lesion with black dots, KOH finding of sclerotic bodies and mixed cell granulomatous inflammation from tissue section help confirm diagnosis.

Disseminated superficial porokeratosis is a rare skin disease with the presence multiple scattered macules and papules on body and extremities. The characteristics are annular ridge at the rim of lesion and cornoid lamella and the presence of parakeratotic column in histopathological study.

We report a co-existence of chromoblastomycosis and disseminated superficial porokeratosis with secondary cutaneous amyloidosis in a 70 year old male patient.

Key words: Chromoblastomycosis, Porokeratosis, Amyloidosis, Itraconazole

From: Faculty of Medicine, Naresuan University, Phitsanulok, Thailand

Corresponding author: Sakchai Chaiyamahapurk MD PhD, email:sakchaipurk@gmail.com

บทคัดย่อ:

ศักดิ์ชัย ไชยมหาพฤกษ์ ชัยพร วิโรจน์แสงอรุณ ประทีป วรรณิสสร

รายงานผู้ป่วยโรค CHROMOBLASTOMYCOSIS ที่พบร่วมกับโรค DISSEMINATED SUPERFICIAL POROKERATOSIS ที่มี AMYLOID ในชั้นหนังแท้ วารสารโรคผิวหนัง 2561; 34: 169-175.

คณะแพทยศาสตร์ มหาวิทยาลัยนเรศวร จังหวัดพิษณุโลก

Chromoblastomycosis เป็นโรคผิวหนังซึ่งพบได้ไม่บ่อยและอาจทำให้ได้รับการวินิจฉัยช้าและมีรอยโรคลุกลามได้ ผู้ป่วยมีลักษณะทางคลินิกของผื่นบริเวณแขนซ้ายที่หนาตัวและมีจุดดำ การตรวจเชื้อราด้วยสารละลายโปตัสเซียมไฮดรอกไซด์โดยขูดจากบริเวณจุดดำพบเชื้อราลักษณะกลมสีน้ำตาล ตรวจพยาธิสภาพผิวหนังพบการอักเสบของผิวหนังแท้แบบแกรนูโลม่าชนิดเซลล์ผสม และพบเชื้อราลักษณะกลมติดสีน้ำตาล

Disseminated Superficial Porokeratosis เป็นโรคที่พบได้ไม่บ่อยและมีลักษณะรอยโรคเป็นตุ่ม ผื่นเล็กๆจำนวนมาก บริเวณลำตัว แขนขา เมื่อคลำรอยโรคจะมีลักษณะนูนบริเวณขอบ การตรวจพยาธิสภาพผิวหนังพบ Cornoid lamella

รายงานฉบับนี้เป็นการนำเสนอผู้ป่วยชายไทยอายุ 70 ปี ซึ่งพบการเกิดโรค Chromoblastomycosis ในผู้ป่วย Disseminated superficial porokeratosis และ มี amyloid ฝังในชั้นหนังแท้

คำสำคัญ: Chromoblastomycosis, Porokeratosis, Itraconazole

A 70 year old Thai man came to outpatient clinic with single verrucous plaque on left elbow for two years. It gradually increased in size. Dermatological examination showed single hyperkeratotic erythematous plaque on left elbow, size 6 x 10 centimeters (Figure 1). Several small minute black dots were seen in edematous verrucous plaque. Also many scattered individual hyperpigmented macules and papules with a subtle threadlike annulus on the rim of the lesions were observed on arm, leg and body. He did not concern about these scattered hyperpigmented lesions which had been since 30 years before. His grandfather, father and brother also developed scattered lesions on body and extremities like him. The patient was usually healthy with no history of chronic or recurrent illness.

Two skin biopsies were performed. Histopathological study of lesion from the solitary lesion on the left elbow (Figure 1) show pseudoepitheliomatous hyperplasia overlying mixed cell suppurative granulomatous inflammation in dermis with lymphocyte, neutrophil, epithelioid histiocytes, and multinucleated giant cells infiltration. Few round bronze colored bodies were found in the dermis. KOH preparation from left elbow plaque also reveals pigmented sclerotic bodies. Fungal

culture from biopsy specimen was positive for dark septate hyphae.

Histopathological study of lesion from hyperpigmented macule on the left arm (Figure2) show cornoid lamella containing parakeratotic corneocyte. Superficial dermis show aggregate of eosinophilic aggregation material, positive with Congo Red stain with apple-green birefringence under polarized light.

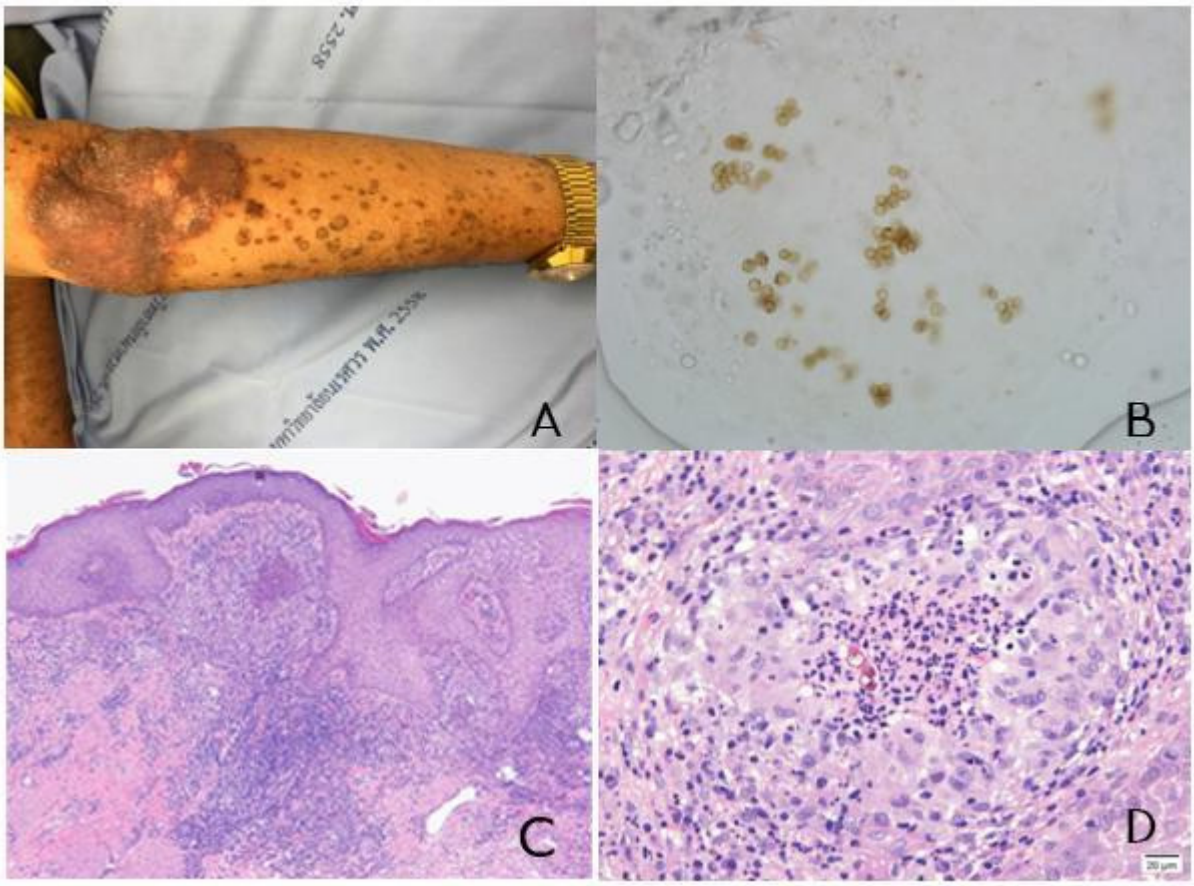

Figure 1 (A) Skin lesion on left elbow and forearm.

(B) Brown sclerotic bodies on KOH preparation.

(C) Hematoxylin-eosin stain revealed pseudoepitheliomatous epidermal hyperplasia.

(D) Dermal mixed cell granulomas containing brown, round-shaped organism.

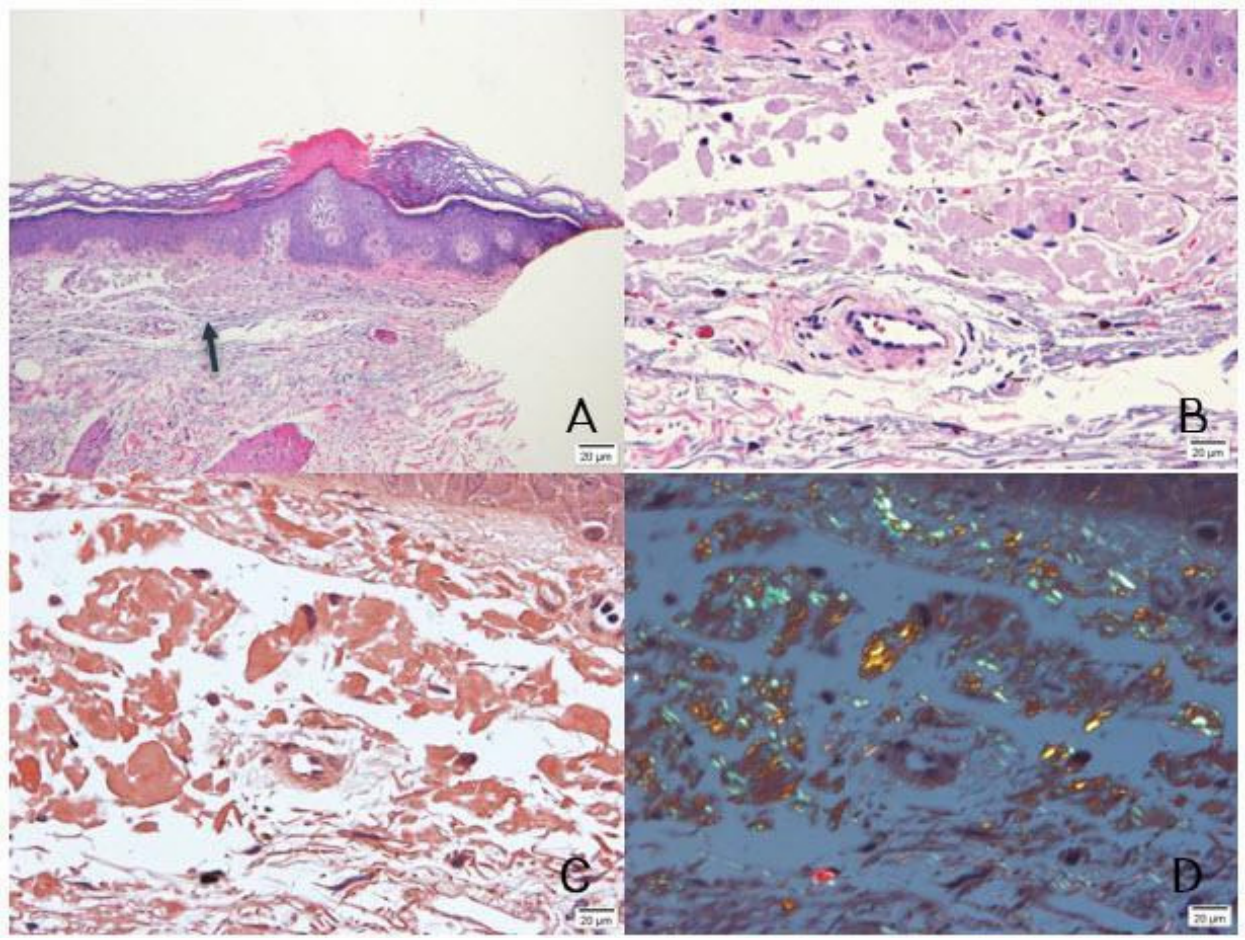

Figure 2

(A) Low magnification of biopsy from scattered hyperpigmented macule. Cornoid lamella and hyperkeratosis.

(B) High magnification, dermal deposition of eosinophilic amorphous material (A-arrow).

(C) Positive Congo Red stain.

(D) apple-green birefringence under polarized light.

Further investigation including CBC, renal function and liver function test were normal. Chromoblastomycosis was diagnosed for the solitary plaque, the patient was treated with Itraconazole (200-mg capsule, 4 capsules per day). After 4 months of treatment, regression of skin lesion was marked with almost clear of verrucous surface and black dot.

Disseminated superficial porokeratosis was diagnosed for scattered hyperpigmented macule and papule which show annular ridge on palpation and cornoid lamella in

histopathological study. No treatment was given since he do not concern but advise on sun exposure avoidance and observation for low risk of occurrence of squamous cell carcinoma.

Discussion

Chromoblastomycosis is a disease of tropical and subtropical region. Various saprophytic hyphomycetes fungi of the Dematiaceae (darkly pigmented fungi) family, including Fonsecaea pedrosoi, Phialophora verrucosa, Cladophialophora carrionii cause chromoblastomycosis.[1] The disease were commonly founded and reported in tropical and subtropical region of America, Asia, Africa and Australia including many reports from Madagascar, Brazil and India. The disfiguring from delay diagnosis and long duration of high cost oral antifungal make it to be proposed as a neglected tropical disease.[2] In Thailand, chromoblastomycosis were occasional found.[3] Average three cases were diagnosed per year at Institute of Dermatology, Bangkok, Thailand [4] and six cases were found during four years period (1994-1997) at Siriraj hospital.[5] The disease most likely occurred by occupational exposure in agricultural by inoculation of fungi related to penetrating wound. Treatment with oral antifungal such as itraconazole and terbinafine with long duration of treatment at least 6-7 months duration may have cure rate up to 80-90%. Surgical excision for early lesion is possible. Other treatment option include cryosurgery, local heat and 5-flucytosine.[6,7]

Disseminated superficial porokeratosis is sometimes inherited as an autosomal dominant disorder but most cases appear sporadic. The pathogenesis is thought to be a disorder of keratinization and immunologic response. It can also be triggered by some factors such as UV exposure or immunosuppression. Reported immunosuppression were organ transplantation, HIV, various immunologic diseases and immunosuppression drugs.[8]

There are 5 forms of porokeratosis. (Porokeratosis of Mibelli, Disseminated superficial (actinic) keratosis, Linear, Punctate) Disseminated superficial porokeratosis is the most common form of porokeratosis usually involves the extremities bilaterally and symmetrically while actinic form occurs exclusively in the sun exposed area. Small asymptomatic keratotic papules are characteristic with central atrophy and thin elevated keratotic rim. Cornoid lamella is a distinctive structure present in tissue section but not specific which can be found in wart, solar keratosis, seborrheic keratosis, squamous cell carcinoma and basal cell carcinoma. There were reports of secondary cutaneous amyloidosis in disseminated superficial porokeratosis lesion which premature degeneration of epidermal cell lead to deposit of amyloid protein in upper dermis.[9-11]

Development of squamous cell carcinoma have been reported in porokeratosis lesion (high in linear type, less common in disseminated type) which appear as a papule arising in contiguity with porokeratosis lesion. For the treatment, many modalities have been tried such as cryotherapy, topical 5-FU, topical retinoid, laser, oral acitretin. The curative treatment may result into scar so should be weight against the low risk of developing of squamous cell carcinoma.

In this report, we report occurrence of two rare skin diseases in a patient. Porokeratosis were reported associated with immunosuppression condition but there was no sign of immunosuppression in this patient with 30 years of skin lesion of porokeratosis. There are two polar of infection by melanized fungi. Chromoblastomycosis is usually occurred in healthy individual while infection in immunocompromised host were better referred to as phaeohyphomycosis, another polar of the infection because only hyphae and yeast are observed in the tissue.[12,13] In this patient, Chromoblastomycosis and porokeratosis, two rare diseases might occur by chance and were unlikely to related to each other.

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