A case report of discrete papular subtype of lichen myxedematosus.

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ABSTRACT:

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Lichen myxedematosus is a chronic, idiopathic disorder characterized by an abnormal accumulation of mucin in the skin. The localized variants of lichen myxedematosus are subdivided into four subtypes: (1) a discrete papular form; (2) acral persistent papular mucinosis; (3) cutaneous mucinosis of infancy; (4) a pure nodular form.

We reported a case of 48-year-old female patient who presented with multiple discrete flat-topped whitish papules on the chest wall, upper back, both forearms, and dorsum of both hands and feet. The histopathology and special staining were compatible with lichen myxedematosus. Laboratory investigations for further systemic involvements were performed. Lichen myxedematosus is not associated with sclerosis, paraproteinemia, systemic involvement or thyroid disease. The disease has a good prognosis and does not require any treatment. Wait-and-see approach is recommended.

Key words: Discrete papular subtype of lichen myxedematosus, primary cutaneous mucinosis

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บทคัดย่อ:

ณิชา เจนมานะชัยกุล นิอร บุญเผื่อน รายงานผู้ป่วยโรคมิวซินสะสมที่ผิวหนังที่มีรอยโรคแบบตุ่มนูน วารสารโรคผิวหนัง 2560; 33: 186-193.

สถาบันโรคผิวหนัง กรมการแพทย์

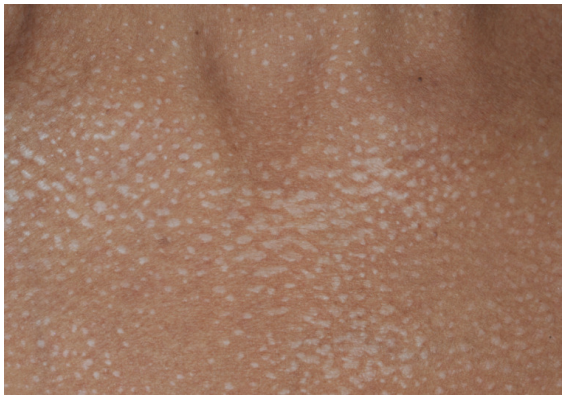
โรคมิวซินสะสมที่ผิวหนัง สาเหตุไม่ทราบแน่ชัด โดยมีลักษณะทางคลินิกเป็นตุ่มนูนสีขาวมันวาว จำนวนมาก ขนาดเล็ก กระจายตามแขนขาและลำตัว กรณีรอยโรคเป็นเฉพาะที่ สามารถจำแนกตามรอยโรคเป็น 4 แบบ คือ discrete papular form, acral persistent papular mucinosis, cutaneous mucinosis of infancy และ pure nodular form

รายงานฉบับนี้เป็นการนำเสนอผู้ป่วยหญิงอายุ 48 ปี ที่มาด้วยตุ่มนูนราบสีขาวหลายตุ่ม กระจายตามหน้าอก หลัง ปลายแขน 2 ข้าง หลังมือและหลังเท้า 2 ข้าง จากผลการตรวจทางพยาธิวิทยาและการย้อมพิเศษ ให้การวินิจฉัยว่าเป็นโรคมิวซินสะสมที่ผิวหนังชนิดตุ่มนูน การตรวจร่างกายและผลเลือดพบว่าอยู่ในเกณฑ์ปกติ โรคนี้ไม่สัมพันธ์กับโรคหนังแข็ง, paraproteinemia, อวัยวะอื่นๆ และโรคไทรอยด์ การพยากรณ์โรคดี ไม่จำเป็นต้องรักษา ยังมีการนัดเพื่อติดตามอาการในระยะยาวต่อไป

คำสำคัญ: โรคมิวซินสะสมที่ผิวหนังแบบตุ่มนูน, โรคมิวซินสะสมที่ผิวหนัง

Case report

A 48-year-old Thai female patient presented five months history of multiple whitish papules on the chest wall, upper back, both forearms, and dorsum of both hands and feet. The lesions initially developed on the chest wall. After a month, the lesions developed on the upper back, both forearms, and dorsum of both hands and feet. She denied history of any systemic symptoms, prior trauma, exposure to chemical compound, or keloidal scar. No family members experienced similar condition. Physical examination revealed multiple discrete flat-topped whitish papules on the chest wall, upper back, both forearms, and dorsum of both hands and feet. Neither hepatosplenomegaly nor lymphadenopathy was observed. Complete blood count (CBC), liver function tests (LFTs), and thyroid function tests (TFTs) were unremarkable. Serology tests for human immunodeficiency virus (HIV) and hepatitis C virus (HCV) were all negative. The serum protein electrophoresis was within normal limits. A 4-mm punch biopsy from right shoulder was performed. The dermis showed a mild superficial perivascular infiltration of lymphocytes. Alcian blue staining demonstrated some fibroblastic proliferation in the dermal interstitium with increased deposition of mucin. Thus, the final diagnosis was consistent with discrete papular subtype of lichen myxedematosus. The treatment was advising the patient that the disease rarely resolves spontaneously.



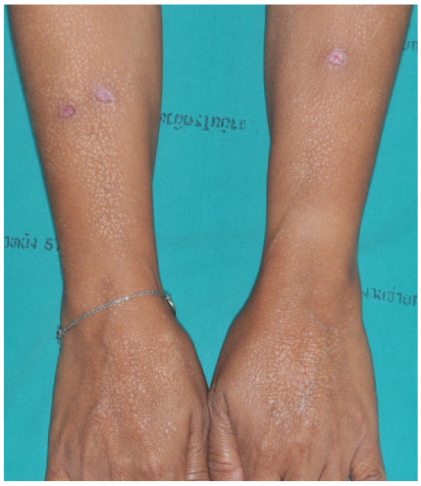


Figure 1A, 1B Multiple discrete flat-topped whitish papules on the chest wall, both forearms, and dorsum of both hands.

Discussion:

Lichen myxedematosus or papular mucinosis is an idiopathic cutaneous mucinosis.[1] It is characterized by asymptomatic, multiple, small, firm, waxy papules (or nodules and plaques produced by the confluence of papules) usually on the upper and lower limbs and trunk.[2] The skin is the only site of involvement. Lichen myxedematosus is not associated with sclerosis, paraproteinemia, systemic involvement or thyroid disease, in contrast to scleromyxedema (Table 1).[3]

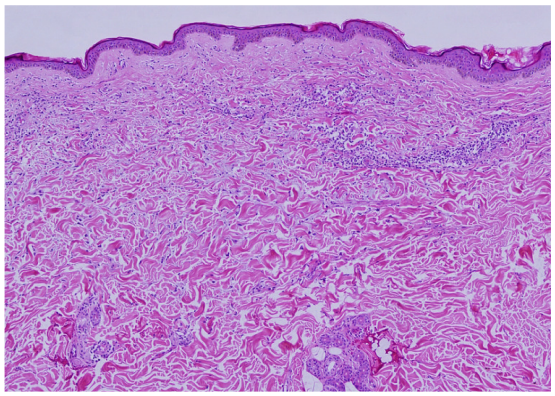


Figure 2 Histopathological image displays unremarkable epidermis with hyperkeratosis. Neither spongiosis nor interface change is noted. No necrotic keratinocyte is present. The dermis shows a mild superficial perivascular infiltration of lymphocytes. No pigmentary incontinence is seen.

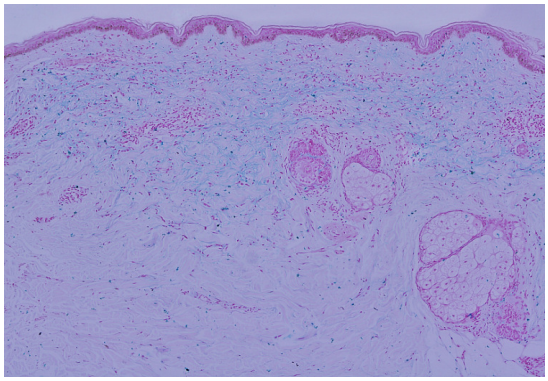


Figure 3 Alcian blue staining demonstrates some fibroblastic proliferation in the dermal interstitium with increased deposition of mucin.

Table 1 Diagnostic criteria of scleromyxedema versus lichen myxedematosus

Scleromyxedema	Lichen myxedematosus
Generalized papular eruption and sclerodermoid features	Papular eruption (or nodules and/or plaques due to confluence of papules)
Microscopic triad (mucin deposition, fibroblast proliferation, fibrosis)	Mucin deposition with variable fibroblast proliferation
Monoclonal gammopathy	Absence of monoclonal gammopathy
Absence of thyroid disorder	Absence of thyroid disorder

The pathogenesis of lichen myxedematosus remains unknown.[4] The incidence and prevalence rates are not known.[2] The localized variants of lichen myxedematosus are subdivided into four subtypes: (1) a discrete papular form; (2) acral persistent papular mucinosis; (3) cutaneous mucinosis of infancy; (4) a pure nodular form (Table 2). Localized variants of lichen myxedematosus have been reported, in patients with HIV infection, hepatitis C viral infection, generalized morphea, morbid obesity, subclinical hypothyroidism, and in the setting of toxic oil syndrome and L-tryptophan-associated eosinophilia-myalgia syndrome.[5-11]

Table 2 Localized variants of lichen myxedematosus

	Lichen myxedematosus (localized variants)			
	Discrete papular type	**Acral persistent type**	**Cutaneous mucinosis of infancy**	**Nodular type**
Characteristics of lesion	• 2-5 mm papules • Few to hundreds • Symmetrical pattern • No induration	• Multiple ivory to skin-colored papules	• Firm opalescent papules	• Multiple nodules
Location	• Limbs, trunk • Spare face	• Dorsal aspect of the hands, extensor surface of the distal forearms	• Neck, upper arms (especially the elbows), trunk	• Limbs, trunk
Prognosis	• Rarely resolve spontaneously	• Lesions persist without systemic manifestations	• No spontaneous resolution • No systemic symptoms	

Localized forms of lichen myxedematosus histopathologically contains fairly large amounts of mucin which have shown to accumulate in the upper and mid reticular dermis, and is associated with variably increased fibroblasts. Fibrosis is not marked and may even be absent. There is no significant increase in dermal collagen.[12] Histology helps to distinguish localized variants of lichen myxedematosus from several papular eruptions that can have a similar appearance, such as granuloma annulare, lichen amyloidosis, lichen planus and other lichenoid eruptions.[2]

We reported one case of lichen myxedematosus in which the clinical features and histology fit into the diagnostic criteria. Our case is a discrete papular subtype. Discrete papular lichen myxedematosus (DPLM) is a very rare entity, affecting both genders, but is more common in males than females.[13] A PubMed search of the literature for published cases of DPLM found reports of 18 cases (Appendix 1).[14-28]

Appendix 1 Cases of discrete papular subtype of lichen myxedematosus in normal patients[14-28]

Reference	Sex	Age	Lesion	Location	Lab	Treatment	Prognosis
Montgomery, 1953	Female	38	Discrete, rounded, pale-yellow papules about 2 mm in diameter	Dorsal surface of forearms, wrists, hands, and medial aspects of knees	Normal	Not reported	Worsened after 10 years
Woerdeman, 1960	Male	19	Small, millet seed sized, yellowish, non-itchy papules	Wrists, elbows, knees	Not reported	Not reported	Not reported
Tay, 1970	Male	41	Discrete papular lesions, waxy, shining, indurated, and cobblestone-shaped with size varying from a small pea to that of a coin	Trunk, shoulders, extensor aspects of arms and forearms	Normal	Failed therapy with oral thyroxine	Unchanged 2 years
Coskey, 1977	Male	22	Discrete flesh-colored, pea-sized papules	Left deltoid, right arm	Normal	Not reported	Unchanged 8 years
Abd El-Aal, 1981	Female	32	Pale red to yellowish papules glistening and generalized	Back, extensors of the upper and lower limbs	Normal	Not reported	Not reported
Kaymen, 1989	Male	63	Minute fleshy papules	Face	Normal	CO_2 laser and intralesional corticosteroids	Improved
Reynolds, 1992	Male	32	Numerous skin-colored papules	Chest, back, upper extremities > lower extremities	Normal	Intralesional and topical corticosteroids	Improved
Rongioletti, 1998	Female	59	Multiple 2 to 4 mm flesh-colored papules	Arms, forearms	HCV Ab: positive	None	Worsened with interferon therapy

Reference	Sex	Age	Lesion	Location	Lab	Treatment	Prognosis
Poswig, 2000	Male	62	Flat, flesh-colored papules with a diameter of 2-3 mm	Lower back	Normal	Not reported	Unchanged 18 months
Montesu, 2001	Male	70	Multiple, linear, flesh-colored papules with a diameter of 2-4 mm	Face, neck, clavicle	HCV Ab: positive HCV RNA: positive	Topical corticosteroids and emollient creams	Improved
Sulit, 2005	Female	80	Multiple 2 to 4 mm discrete, flesh-colored shiny papules without scale	Bilateral and posterior neck	Normal	Failed therapy with topical corticosteroids; successful symptomatic relief with pimecrolimus	Unchanged 14 months
Seike, 2005	Male	78	Discrete papules with 3-5 mm in diameter, reddish-brown, smooth surface	Upper back	Normal	Failed therapy with topical corticosteroids and cryotherapy	Unchanged
Bragg, 2008	Female	56	Numerous 2 to 4 mm skin-colored, firm, waxy papules	Central chest, abdomen, upper anterior thighs	Normal	Intralesional and topical corticosteroids	Unchanged
Rongioletti, 2008	Female	67	Numerous, small, firm, waxy, asymptomatic papules	Neck	Normal	Topical 0.1% tacrolimus ointment	Improved
Rongioletti, 2008	Female	50	Small, firm, flesh-colored papules	Shins	Normal	Topical 0.1% tacrolimus ointment	Improved
Concheiro, 2009	Female	21	Numerous well-defined reddish-brown papules, 2-4 mm in diameter	Outside of the arms and thighs	Normal	None	Unchanged
Hadj, 2014	Male	42	Multiple 2 to 4 mm discrete, flesh-colored shiny papules without scale	Bilateral posterior neck, left pectoral region, hypogastric region	Normal	Topical corticosteroids	Improved
Tam, 2014	Male	23	Segmentally 1-5 mm indurated, dark brown, slightly erythematous papules	Right lower abdomen	Normal	Intralesional and topical corticosteroids	Improved
Our case	Female	48	Multiple discrete whitish papules	Chest wall, upper back, both forearms, and dorsum of both hands and feet	Subclinical hypothyroidism	Failed therapy with topical corticosteroids	Unchanged

Since localized lichen myxedematosus is usually limited to the skin, it has a good prognosis and does not require any treatment. Wait-and-see approach is recommended. Topical application of corticosteroids, pimecrolimus or tacrolimus may be of some benefit. However, spontaneous resolution may occur, even in the setting of HIV infection.[25]

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