Aquagenic syringeal acrokeratoderma is a newly described disease entity of unclear etiology, affecting predominantly adolescents and young adults. It manifested as multiple whitish papules on the palms or rarely soles, which became accentuated after immersion in water and improved after drying the involved area. Burning, pain, pruritus or tightening sensation may be present and usually there is an association with palmar hyperhidrosis. The histopathology revealed dilated sweat duct ostia with focal mild hyperkeratosis. Most patients responded well to topical aluminum chloride, but recurrence could occur. We presented a 19-year-old healthy male with multiple whitish, translucent, asymptomatic papules mainly on his left palm for 7 years. Such a condition became more pronounced after exposure to warm water and resolved to residual smaller flesh-colored to white papules after a drying period. He reported occasional mild palmar hyperhidrosis of his left hand and had a family history of atopy. The dermatoscopy revealed dilated puncta on the papules. The histopathologic examination revealed dilatation of intracorneal eccrine ducts with mild focal hyperkeratosis and acanthosis. After receiving topical 20% anhydrous aluminum chloride nightly for 5 weeks, he reported almost disappearance of the papules when the palms were dry. (Dermatol Sinica 26: 145-150, 2008)

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first became aware of this condition on the volar aspect of his left little finger after taking showers. Several weeks later, the whole left palm was involved. The condition was more pronounced but remained asymptomatic after exposure to warm water for around 5 minutes and resolved to residual smaller flesh-colored to white papules after a drying period of 1 hour. He was otherwise in excellent health. He reported mild left palmar hyperhidrosis occasionally. The sweating did not accentuate the papules. His hairs are normal in thickness, and he denied a personal history of atopy. There was no family history of similar skin lesions. However, his father had asthma and his elder sister had asthma and allergic rhinitis. He was right-handed. He did not have trauma or hand dermatitis previously. On the initial dermatological examination before exposure to water, the left palm, the volar aspects of fingers and, to a less extent, the lateral aspect of the right thumb were affected by multiple, 1-2 mm, non-scaling, smooth-surfaced, white papules after a drying period of 1 hour. (Fig. 1) A punch biopsy was taken from the medial aspect of the left palm after water immersion for 10 minutes. Histopathologic examination revealed compact orthohyperkeratosis with acanthosis, elongation of the rete ridges and dilatation of intracorneal eccrine ducts. (Fig. 2) Iodine starch test for bilateral palms revealed no evidence of hyperhidrosis. Trans-epidermal water loss was decreased on the left palm. Dermoscopy revealed dilated puncta. He received desoximetasone ointment (Esperson ointment) once in the morning and aluminum hydroxychloride & Cetrimide (Joyla® cream) once nightly for 1 week. However, the effect was limited. Thereafter, he received 20% anhydrous aluminum chloride (Odaban® antiperspirant pump spray) instead once nightly for 5 weeks. There was significant improvement with almost disappearance of whitish papules grossly when the skin was dry. However, multiple whitish papules still appeared on the left palm after immersion to water for 5 minutes after the 5-week treatment.
DISCUSSION

Aquagenic syringeal acrokeratoderma is a rare and probably underdiagnosed disease, which predominantly affects adolescents and young adults.\(^4\) It is mostly observed in females and occasionally in males.\(^5\) The age of onset ranges from 9-42 years.\(^6,16\) In affected patients, the skin either looks normal, or there is hyperlinearity, or appear as multiple unremarkable translucent white papules on the palms and rarely on the soles prior to immersion in water.\(^7,8\) The papules become more pronounced with pebbly appearance after brief immersion in water, especially hot water for 2 to 10 minutes.\(^3,7\) The condition is called “hand-in-the-bucket” sign, which is a useful clinical clue for diagnosis.\(^9\) There are markedly dilated puncta on the papules.\(^9\) A burning sensation, pain, pruritus or tightening sensation may be present, although the condition may be asymptomatic.\(^3,5\) Within minutes to an hour after drying the involved area, the papules resolve or become unremarkable.\(^7\)

Aquagenic syringeal acrokeratoderma and hereditary papulotranslucent acrokeratoderma are two forms of papulotranslucent acrokeratoderma reported in the literature.\(^10\) The latter was first described in 1973 by Onwukwe et al.\(^11\) It begins at puberty and is characterized by persistent, asymptomatic, bilateral, symmetric, translucent, yellowish-white papules and plaques on the margins of the hands and feet, increased in areas of pressure and trauma.\(^4,11,12\) The existing lesions did not disappear after.\(^10\) Worsening of the condition may occur after soaking in water.\(^7\) Males and females are equally affected.\(^7\) It is usually inherited in an autosomal dominant mode with an association with fine-textured scalp hair and an atopic diathesis.\(^12,13\) The pathogenesis of hereditary papulotranslucent acrokeratoderma is unknown. Physical trauma may play a role in initiating the condition.\(^10\) The histological findings of hereditary papulotranslucent acrokeratoderma were focal hyperkeratosis, acanthosis and normal eccrine ducts.\(^13\) Despite the differences between hereditary papulotranslucent acrokeratoderma and aquagenic syringeal acrokeratoderma, these two diseases may represent variants within a spectrum of disease.\(^4\)

The pathogenesis of aquagenic syringeal acrokeratoderma remains unclear. It has been considered a variant of hereditary papulotranslucent acrokeratoderma, yet other proposed aquagenic syringeal acrokeratoderma a separated entity from hereditary papulotranslucent acrokeratoderma.\(^1,2\) There may be pathological barrier defect in the stratum corneum with tendency to mild hyperkeratosis, which allows an increased water absorbance and further causes the stratum corneum swelling during water immersion.\(^7,12\) The dilated eccrine duct is probably a secondary phenomenon.\(^11,12\) Keratin defect, or altered cornified envelope such as a mutation in transglutaminase or involucrin might be responsible for the pathological barrier defect in the stratum corneum.\(^7\) Other hypotheses suggest that an aberration of the sweat glands and external factors such as occlusion or friction may predispose the dilatation of the eccrine ostia.\(^13\) It was also proposed that the patients may have defective chloride channels as in cystic fibrosis or elevation of skin sodium as is seen in the effect of celecoxib or rofecoxib.\(^15,17\) The hypertonic sweat creates osmotic gradient that makes the isotonic fluids dilate the eccrine ostia and flow into the eccrine ducts.\(^15,17\)

There are several findings that have previously been reported in association with aquagenic syringeal acrokeratoderma, including palmar hyperhidrosis, increased foot odor, asthma, allergic rhinitis, cystic fibrosis, palmar erythema, malignant melanoma, and treatment with celecoxib or rofecoxib.\(^3,10,12,14\) As siblings with this condition have been reported, a genetic susceptibility may be pos-
The pathological examination of aquagenic syringeal acrokeratoderma after immersion in water revealed hyperkeratosis.
around dilated sweat duct ostia. Most cases had normal dermis except for one case reporting hyperplastic eccrine glands in the dermis with unremarkable epidermis.

Multiple treatments for aquagenic syringeal acrokeratoderma were tried mainly on the basis of the associated hyperhidrosis, including topical aluminum chloride, iontophoresis, and botulinum toxin. Some patients had spontaneous amelioration. Reviewing the English literature, a total of 21 cases are found, including our case. 11 patients received aluminum chloride treatment and 9 patients responded well. Five patients who responded well to aluminum chloride had hyperhidrosis. Among the 8 patients who responded well (other than the present case), 2 patients had recurrences, 5 patients had no record of recurrence, and 1 patient had no recurrence after a short period of follow-up for 1 year. Three patients had spontaneous amelioration (Table 1).

In conclusion, we reported a case of aquagenic syringeal acrokeratoderma. It manifests as multiple whitish, flat-topped papules on the palms and rarely on the soles, which are accentuated after immersion to water and resolve or become unremarkable after a short drying period. There is an association with palmar hyperhidrosis. The histopathological examination reveals intracorneal dilated eccrine ostia with hyperkeratosis. Our patient responded well to topical aluminum chloride.

REFERENCES
水因性汗管肢端角皮症

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水因性汗管肢端角皮症是一個原因不明，近來才被報告的疾病，主要影響年輕人，病灶為手掌出現很多白色小丘疹，少數例子是出现在腳掌，這樣的白色丘疹在泡水後會更加明顯；病灶乾燥後，白色小丘疹會恢復原來的樣子。病人偶爾會抱怨有灼熱、疼痛、搔癢或是緊繃感，常伴隨多汗症。病理學檢查發現有擴張的汗管開口，開口處常局部過度角化。大多數的病人對於局部使用氯化鋁反應良好，但仍有可能會再復發。我們報告一例19歲健康的男性，有許多白色透明、無症狀的丘疹主要分布於左手掌，在遇到溫水後丘疹會變的更加明顯，在風乾一段時間之後，恢復原來較小的樣子。他宣稱左手掌偶有多汗的情形，家人有異位性體質。以皮膚視鏡檢查可發現丘疹上有擴張的小孔。病理檢查為表皮層有擴張的汗管伴隨局部過度角化和棘層肥厚。在每晚接受了百分之二十無水氯化鋁局部治療共五週之後，在乾燥的情形下，小丘疹幾乎都消失了。（中華皮誌：26: 145-150, 2008）