ISSN 2307-8960 (online)

World Journal of *Clinical Cases*

World J Clin Cases 2023 May 6; 11(13): 2855-3113

Published by Baishideng Publishing Group Inc

World Journal of Clinical Cases

Contents

Thrice Monthly Volume 11 Number 13 May 6, 2023

OPINION REVIEW

REVIEW

MINIREVIEWS

ORIGINAL ARTICLE

Case Control Study

Retrospective Study

Observational Study

World Journal of Clinical Cases

Thrice Monthly Volume 11 Number 13 May 6, 2023

Contents

World Journal of Clinical Cases

Thrice Monthly Volume 11 Number 13 May 6, 2023

Contents

World Journal of Clinical Cases

Thrice Monthly Volume 11 Number 13 May 6, 2023

Contents

NAME OF JOURNAL
World Journal of Clinical Cases

ISSN
ISSN 2307-8960 (online)

LAUNCH DATE
April 16, 2013

FREQUENCY
Thrice Monthly

EDITORS-IN-CHIEF
Bao-Gan Peng, Jerzy Tadeusz Chudek, George Kontogeorgos, Maurizio Serati, Ja Hyeon Ku

EDITORIAL BOARD MEMBERS
https://www.wjgnet.com/2307-8960/editorialboard.htm

PUBLICATION DATE
May 6, 2023

COPYRIGHT
© 2023 Baishideng Publishing Group Inc

INSTRUCTIONS TO AUTHORS
https://www.wjgnet.com/bpg/gerinfo/204

GUIDELINES FOR ETHICS DOCUMENTS
https://www.wjgnet.com/bpg/GerInfo/287

GUIDELINES FOR NON-NATIVE SPEAKERS OF ENGLISH
https://www.wjgnet.com/bpg/gerinfo/240

PUBLICATION ETHICS
https://www.wjgnet.com/bpg/GerInfo/288

PUBLICATION MISCONDUCT
https://www.wjgnet.com/bpg/gerinfo/208

ARTICLE PROCESSING CHARGE
https://www.wjgnet.com/bpg/gerinfo/242

STEPS FOR SUBMITTING MANUSCRIPTS
https://www.wjgnet.com/bpg/GerInfo/239

ONLINE SUBMISSION
https://www.f6publishing.com

World Journal of Clinical Cases

Submit a Manuscript: https://www.f6publishing.com

World J Clin Cases 2023 May 6; 11(13): 3086-3091

DOI: 10.12998/wjcc.v11.i13.3086

ISSN 2307-8960 (online)

CASE REPORT

Solitary acral persistent papular mucinosis nodule: A case report and summary of eight Korean cases

Yu Jeong Park, Hui Young Shin, Woo Kyoung Choi, Ai-Young Lee, Seung Ho Lee, Jong Soo Hong

Specialty type: Medicine, research and experimental

Provenance and peer review: Unsolicited article; Externally peer reviewed.

Peer-review model: Single blind

Peer-review report's scientific quality classification

Grade A (Excellent): 0
Grade B (Very good): 0
Grade C (Good): C
Grade D (Fair): 0
Grade E (Poor): 0

P-Reviewer: Fan L, China

Received: December 29, 2022
Peer-review started: December 29, 2022
First decision: March 10, 2023
Revised: March 22, 2023
Accepted: March 31, 2023
Article in press: March 31, 2023
Published online: May 6, 2023

Yu Jeong Park, Hui Young Shin, Woo Kyoung Choi, Ai-Young Lee, Seung Ho Lee, Jong Soo Hong, Department of Dermatology, Dongguk University Ilsan Hospital, College of Medicine, Dongguk University, Goyang-si, Gyeonggi-do, Korea 10326, South Korea

Corresponding author: Jong Soo Hong, MD, PhD, Professor, Department of Dermatology, Dongguk University Ilsan Hospital, College of Medicine, Dongguk University, Dongguk University Ilsan Medical Center, 27, Dongguk-ro, Ilsandong-gu, Goyang-si, Gyeonggi-do, Korea 10326, South Korea. jsttjstt@hanmail.net

Abstract

BACKGROUND

Acral persistent papular mucinosis (APPM) is a rare idiopathic subtype of localized lichen myxedematosus. To date, there have been 40 APPM cases reported worldwide; however, only 7 cases have been reported in the Korean literature.

CASE SUMMARY

A 70-year-old man was referred to our hospital with a solitary pinkish nodule on the dorsum of his right hand. Despite the absence of symptoms, the patient wanted to know the exact diagnosis; thus, a biopsy was performed. Histopathological examination of a biopsy specimen obtained from the nodule on the dorsum of his hand revealed orthokeratotic hyperkeratosis with patchy parakeratosis, prominent hypergranulosis, and diffuse dissecting mucinous deposition between collagen bundles, along with some bland-looking spindle cells throughout the dermis. The nodule was histologically diagnosed as an APPM, and an intralesional triamcinolone injection (2.5 mg/mL) was started every 2 wk. After three sessions of treatment, the patient showed marked improvements.

CONCLUSION

To the best of our knowledge, this is the first case of a Korean APPM presenting as a solitary nodule that showed a marked response to triamcinolone intralesional injection. Since it is a rare disease, we report this case to contribute to future research on the pathogenesis and treatment of APPM.

Key Words: Acral persistent papular mucinosis; Localized lichen myxedematosus; Cutaneous mucinosis; Mucin; Case report

Core Tip: Acral persistent papular mucinosis (APPM) is a rare idiopathic subtype of localized lichen myxedematosus. To date, 40 cases have been reported worldwide; however, only seven cases have been reported in the Korean literature. This article reports on a rare case of solitary APPM, which was histologically diagnosed in a 70-year-old Korean man with a pinkish nodule on the dorsum of his hand. The patient showed marked improvement after three sessions of intralesional triamcinolone injection. This is the first reported case of a Korean APPM presenting as a solitary nodule and emphasizes the importance of continued research into the pathogenesis and treatment of this rare disease.

Citation: Park YJ, Shin HY, Choi WK, Lee AY, Lee SH, Hong JS. Solitary acral persistent papular mucinosis nodule: A case report and summary of eight Korean cases. *World J Clin Cases* 2023; 11(13): 3086-3091
URL: https://www.wjgnet.com/2307-8960/full/v11/i13/3086.htm
DOI: https://dx.doi.org/10.12998/wjcc.v11.i13.3086

INTRODUCTION

Acral persistent papular mucinosis (APPM) is a rare idiopathic subtype of localized lichen myxedematosus (LM)[1]. It is characterized by white or skin-colored, 2–5 mm papules on the hands, wrists, and extensor aspects of the forearms[1]. The number of papules varies from a few to hundreds and is usually asymptomatic; however, some patients complain of pruritus[1].

Most cases have a favorable prognosis and require no treatment; however, in some cases, tacrolimus ointments or triamcinolone intralesional injections have been administered.

To date, there have been 40 APPM cases reported worldwide[1,2], however, only 7 cases have been reported in the Korean literature. Herein, we present a new case of an older Korean adult with solitary APPM as an atypical manifestation. In addition, we summarized 8 cases of APPM, including ours.

CASE PRESENTATION

Chief complaints

A 70-year-old man was referred to our hospital with a solitary pinkish nodule on the dorsum of his right hand (Figure 1A).

History of present illness

The patient was asymptomatic and did not complain of pruritus or pain.

History of past illness

His medical history showed that he had psoriasis for decades, which was managed using topical agents.

Personal and family history

He denied any other familial history of dermatologic or endocrinologic diseases.

Physical examination

Despite the absence of symptoms, the patient wanted to know the exact diagnosis; thus, biopsy and blood tests were performed.

Laboratory examinations

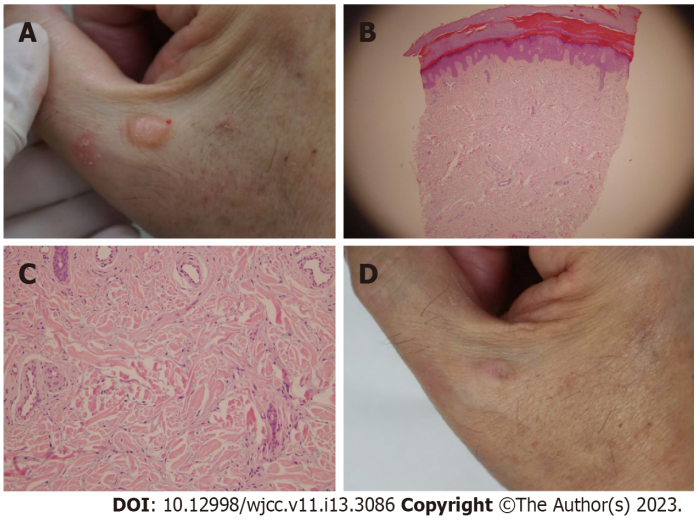
Blood tests showed no endocrine abnormalities including thyroid function.

Imaging examinations

Histopathological examination of a biopsy specimen obtained from the nodule on the dorsum of his hand revealed orthokeratotic hyperkeratosis with patchy parakeratosis, prominent hypergranulosis, and diffuse dissecting mucinous deposition between collagen bundles, along with some bland-looking spindle cells throughout the dermis (Figure 1B and C).

FINAL DIAGNOSIS

The nodule was histologically diagnosed as an APPM.



DOI: 10.12998/wjcc.v11.i13.3086

Figure 1 A pinkish, solitary nodule measuring 1 cm was found on the right dorsum of the hand. A: A clinical image taken before treatment showed the appearance of the nodule; B: Biopsy results showed orthokeratotic hyperkeratosis with patchy parakeratosis, prominent hypergranulosis; C: Diffuse dissecting mucinous deposition between collagen bundles, along with some bland-looking spindle cells throughout the dermis (H&E; B: ×40, C: ×200); and D: Following three sessions of intralesional triamcinolone injection every 2 wk, a marked improvement was observed.

TREATMENT

To reduce the size, intralesional triamcinolone injection (2.5 mg/mL) was administered once every 2 wk.

OUTCOME AND FOLLOW-UP

After three sessions of treatment, the patient showed marked improvements (Figure 1D).

DISCUSSION

Cutaneous mucinosis is a medical term used for a diverse group of skin disorders that involve a localized or widespread accumulation of mucin in the skin or within the hair follicle[3,4]. Mucin is composed of mucopolysaccharides acid or hyaluronic acid and is generally present as part of the connective tissue of the dermis[3,4]. Its main function is to maintain the balance of salt and water in the dermis. The excessive deposition of mucin manifests as a clinically specific lesion or a mucinous rash[3,4]. Cutaneous mucinosis can be divided into two main groups: Generalized form (scleromyxedema), which classically presents with systemic abnormalities, such as monoclonal gammopathy or thyroid disease; and localized form (LM), which is a rare form and classically presents with a lack of systemic disease[3,4].

APPM was first described by Rongioletti *et al*[1] in 1986. It is one of the five subtypes of LM, namely, a discrete form, APPM, self-healing papular mucinosis, papular mucinosis of infancy, and a pure nodular form[3]. APPM is a chronic idiopathic cutaneous mucinosis characterized by lichenoid papules or nodules with mucin deposition and the absence of associated thyroid disease, paraproteinemia, and other systemic abnormalities[3-5]. Patients with this diagnosis are typically women and present with a bilaterally symmetrical distribution on their hands and wrists but not on the trunk and face[6].

The etiology of APPM is still unknown; however, family cases have shown that genetic and environmental factors play an important role[7]. Interleukin-1, interferon gamma, tumor necrosis factor-alpha (TNF-α), and transforming growth factor beta may stimulate glycosaminoglycan synthesis, but the true triggers are unknown, and there is a report of cutaneous mucinosis developing after using a biological agent, such as a TNF-α inhibitor, in patients with psoriasis[8]. Our patient had psoriasis for a long time; however, the direct link between cutaneous mucinosis and psoriasis is not well-known, with no reported literature. Human immunodeficiency virus (HIV) infection can also be linked to primary cutaneous mucinoses, and 18 cases of primary cutaneous mucinoses in HIV-infected patients have been reported, 2 of which are APPM. It has been speculated that direct stimulation of fibroblasts by HIV infection or fibroblast stimulation by activated serum cytokines owing to overactive B-cell function may be associated with mucin deposition, but no definitive mechanism has been elucidated[9].

Table 1 Summary of the Korean cases of acral persistent popular mucinosis

Case	Ref.	Age/Sex	Onset	Past medical history	Clinical manifestation	Treatment	Outcome
1	Kim *et al*[11], 1993	F/56	1 mo ago	s/p Rt. Nephrectomy d/t renal cancer	Multiple, flesh-colored papules on both forearms, wrists and hands without symptoms	None	-
2	Lee *et al*[12], 2000	M/43	6-7 years ago	None	Multiple discrete white to flesh-colored papules are present on the extensor surface of hand without symptoms	None	-
3	Lee *et al*[12], 2000	M/50	7 years ago	None	Multiple whitish papules on dorsum of the hand without symptoms	None	-
4	Song *et al*[6], 2002	F/50	3 years ago	None	Pruritic numerous papular eruption on the dorsa of the hands, and the extensor surface of the forearms	Intralesional triamcinolone injection	No improvement
5	Ryu *et al*[13], 2003	M/64	15 years ago	s/p Subtotal gastrectomy d/t gastric ulcer	Asymptomatic flesh-colored or translucent papules on wrists, back of the hands, and distal forearms	None	-
6	Choi *et al*[9], 2007	M/31	2 mo ago	AIDS	Multiple tiny flesh-colored papules on the extensor surface of the wrist and the distal part of forearm	After 5 wk of starting highly active antiretroviral therapy for AIDS, the skin lesions were also resolved	Resolved
7	Jun *et al*[10], 2016	F/53	7-8 years ago	None	Asymptomatic 1-3 mm flesh-colored papules symmetrically located on both dorsum of hands and wrists, and on anterior chest	Tacrolimus ointment 0.1% once a day	After 15 wk, responded partially

AIDS: Acquired immunodeficiency syndrome.

Since APPM has rarely been reported in Korea, we have summarized the characteristics of APPM cases that have occurred in Korea[6,9-13] (Table 1). To the best of our knowledge, there are eight cases, including the present one, of which five were reported in males and three in females. Their ages ranged from 31 to 70 years, and most were middle-aged. Most had no underlying disease and only two patients had reported underlying medical conditions; one, who was a patient in our case, had psoriasis, while the other had HIV. Only one patient complained of itching, while the rest were asymptomatic. Notably, except for our case, all cases presented symmetrical, multiple papules. To the best of our knowledge, this is the first case of a large solitary nodule. As it is a rare form that has not been previously reported, the possibility of APPM should be considered even with a single nodule. Among the 40 cases reported worldwide, it is interesting that 8 were Korean and 24 were Japanese[1,2], suggesting a racial influence; however, further studies are necessary to confirm this because of the small number of samples.

No effective treatment has been established for APPM. Treatment options range from topical steroids or tacrolimus and intralesional corticosteroids to oral tranilast, which inhibits the release of histamine and prostaglandins from mast cells[2]. However, the effects of these treatments are variable. In the Korean cases, four patients were not treated, and the exact outcome could not be identified owing to loss of follow-up. One patient showed some improvement after 15 wk of topical application of tacrolimus. In another case, an intralesional steroid injection was administered but no improvement was noted, and the exact number and interval of treatment were not known. In one case with acquired immunodeficiency syndrome (AIDS) as an underlying disease, APPM improved with antiviral treatment for AIDS without any specific treatment for APPM[6,9-13]. In our case, intralesional triamcinolone injection was effective; therefore, it can be estimated that intralesional corticosteroids are effective in APPM alone, but additional research is needed. Recently, since the main component of mucin is hyaluronic acid, there have been several case reports in which scleromyxedema and popular dermal mucinosis were treated with hyaluronidase[14,15]. Another therapeutic option is electrocoagulation or erbium-YAG laser[16,17].

CONCLUSION

To the best of our knowledge, this is the first case of a Korean APPM presenting as a solitary nodule that showed a marked response to triamcinolone intralesional injection. Since it is a rare disease, we report this case to contribute to future research on the pathogenesis and treatment of APPM.

ACKNOWLEDGEMENTS

The authors declare that they have no proprietary, commercial, or financial interests that could be construed to have inappropriately influenced this study.

FOOTNOTES

Author contributions: Park YJ and Hong JS wrote the first draft of the manuscript, and all authors reviewed and edited the manuscript and approved the final version of the manuscript.

Informed consent statement: The primary version of the consent that has been signed by the patient in the study is attached as a separate file.

Conflict-of-interest statement: All the authors report no relevant conflicts of interest for this article.

CARE Checklist (2016) statement: The authors have read the CARE Checklist (2016), and the manuscript was prepared and revised according to the CARE Checklist (2016).

Country/Territory of origin: South Korea

ORCID number: Yu Jeong Park 0000-0003-2462-5278; Hui Young Shin 0000-0003-2523-9504; Woo Kyoung Choi 0000-0003-1675-3720; Ai-Young Lee 0000-0001-7720-7833; Seung Ho Lee 0000-0001-5057-1379; Jong Soo Hong 0000-0003-3813-3055.

S-Editor: Liu XF
L-Editor: A
P-Editor: Zhang XD

REFERENCES

1 **Rongioletti F**, Ferreli C, Atzori L. Acral persistent papular mucinosis. *Clin Dermatol* 2021; **39**: 211-214 [PMID: 34272012 DOI: 10.1016/j.clindermatol.2020.10.001]
2 **Mori A**, Shinkuma S, Mitsui Y, Ogawa K, Miyagawa F, Asada H. Acral persistent papular mucinosis: A case report and summary of 24 Japanese cases. *J Dermatol* 2021; **48**: 1574-1578 [PMID: 34169556 DOI: 10.1111/1346-8138.16043]
3 **Rongioletti F**, Rebora A. Updated classification of papular mucinosis, lichen myxedematosus, and scleromyxedema. *J Am Acad Dermatol* 2001; **44**: 273-281 [PMID: 11174386 DOI: 10.1067/mjd.2001.111630]
4 **Cárdenas-Gonzalez RE**, Ruelas MEH, Candiani JO. Lichen myxedematosus: a rare group of cutaneous mucinosis. *An Bras Dermatol* 2019; **94**: 462-469 [PMID: 31644623 DOI: 10.1590/abd1806-4841.20198478]
5 **Harris JE**, Purcell SM, Griffin TD. Acral persistent papular mucinosis. *J Am Acad Dermatol* 2004; **51**: 982-988 [PMID: 15583597 DOI: 10.1016/j.jaad.2004.07.002]
6 **Song JY**, Lee SW, Kim CW, Kim HO. A case of acral persistent popular mucinosis. *Ann Dermatol***14**: 178-180 [DOI: 10.5021/ad.2002.14.3.178]
7 **Menni S**, Cavicchini S, Brezzi A, Gianotti R, Caputo R. Acral persistent papular mucinosis in two sisters. *Clin Exp Dermatol* 1995; **20**: 431-433 [PMID: 8593726 DOI: 10.1111/j.1365-2230.1995.tb01366.x]
8 **Zhou X**, Mubanga D, Chen Z, Bi X. Secondary Generalized Cutaneous Mucinosis Developed During Etanercept Treatment in a Patient with Psoriasis. *Clin Cosmet Investig Dermatol* 2022; **15**: 987-992 [PMID: 35669090 DOI: 10.2147/CCID.S368390]
9 **Choi JE**, Kim JW, Cho HM, Ahn HH, Kye YC, Kim SN. A case of acral persistent popular mucinosis in patient with HIV infection. *Korean J Dermatol* 2007; **45**: 951-955
10 **Jun JY**, Oh SH, Shim JH, Kim JH, Park JH, Lee DY. Acral Persistent Papular Mucinosis with Partial Response to Tacrolimus Ointment. *Ann Dermatol* 2016; **28**: 517-519 [PMID: 27489445 DOI: 10.5021/ad.2016.28.4.517]
11 **Kim YH**, Yoon JS, Lee JS, Lee JH, Park JK. A case of acral persistent popular mucinosis. *Kor J Dermatol* 1993; **31**: 232-235
12 **Lee MW**, Choi JH, Sung KJ, Moon KC, Koh JK. Acral persistent popular mucinosis. *Kor J Dermatol* 2000; **38**: 236-239
13 **Ryu HJ**, Son SW, Kim Aeree, Kim IH. A case of acral persistent popular mucinosis. *Ann Dermatol* 2003; **15**: 8-11 [DOI: 10.5021/ad.2003.15.1.8]
14 **Akaslan TÇ**, Yıldız P, Onsun N. Clinical and histopathological improvement of scleromyxedema-induced microstomia after hyaluronidase injection. *J Cosmet Dermatol* 2022; **21**: 4319-4322 [PMID: 35441782 DOI: 10.1111/jocd.15007]
15 **Ramamurthi A**, Bicknell L, McCarty M. Hyaluronidase in the treatment of papular dermal mucinosis: first case reported

in North America. *J Surg Dermatol* 2016; **4**: 9-13 [DOI: 10.18282/jsd.v1.i2.43]

16 **André Jorge F**, Mimura Cortez T, Guadalini Mendes F, Esther Alencar Marques M, Amante Miot H. Treatment of acral persistent papular mucinosis with electrocoagulation. *J Cutan Med Surg* 2011; **15**: 227-229 [PMID: 21781629 DOI: 10.2310/7750.2011.10030]

17 **Graves MS**, Lloyd AA, Ross EV. Treatment of acral persistent papular mucinosis using an Erbium-YAG laser. *Lasers Surg Med* 2015; **47**: 467-468 [PMID: 25952726 DOI: 10.1002/Lsm.22368]

Published by **Baishideng Publishing Group Inc**
7041 Koll Center Parkway, Suite 160, Pleasanton, CA 94566, USA
Telephone: +1-925-3991568
E-mail: bpgoffice@wjgnet.com
Help Desk: https://www.f6publishing.com/helpdesk
https://www.wjgnet.com