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CASE REPORT

Familial Reactive Perforating Collagenosis with Adolescence-Onset: A Rare Case Report and Literature Review

Laila Tsaqilah[1], Hedwika Advina Nastiti[1], Hermin Aminah Usman[2], Erda Avriyanti[1], Hartati Purbo Dharmadji[1], Risa Miliawati Nurul Hidayah[1]

[1]Department of Dermatology and Venereology, Faculty of Medicine, Universitas Padjadjaran – Dr. Hasan Sadikin General Hospital, Bandung, West Java, Indonesia; [2]Department of Anatomical Pathology, Faculty of Medicine, Universitas Padjadjaran – Dr. Hasan Sadikin General Hospital, Bandung, West Java, Indonesia

Correspondence: Laila Tsaqilah, Department of Dermatology and Venereology, Faculty of Medicine, Universitas Padjadjaran – Dr. Hasan Sadikin General Hospital, Jl. Pasteur No. 38, Bandung, West Java, 40161, Indonesia, Tel/Fax +62 822 8447 4849, Email laila.tsaqilah@gmail.com

Abstract: Familial Reactive Perforating Collagenosis (FRPC) is a very rare form of benign dermatosis frequently presented during early childhood and not associated with systemic diseases. Less than 50 FRPC patients have been reported in the literature. Due to the limited number of cases, the pathophysiology of this unique entity remains elusive; moreover, no standard treatment has been agreed upon. Here, we report a case of FRPC in a 20-year-old male who was presented with generalized multiple discrete papules covered with central keratotic plugs in all regions of his body, particularly in the facial area, neck, abdominal, and extensor region of the extremities for more than 7 years. Similar symptoms were acknowledged in the patient's family members. Histopathological analyses identified the crateriform shape invagination in the epidermis filled with inflammatory lymphocytes and basophilic debris and perforated by basophilic collagen bundles from the underlying dermis. Based on the clinical and histopathological findings, the patient was diagnosed with FRPC. He was treated with topical desoximetasone 0.25% cream applied 2–3 times daily. A follow-up evaluation after 4 weeks revealed a near-complete resolution of skin papules. To our knowledge, this is the first report of FRPC case from Indonesia. Unlike the majority of FRPC patients who had their disease onsets during infancy or early childhood, FRPC skin manifestations in our patient started during the adolescence period. The resolution of skin manifestations after daily application of topical desoximetasone suggests that topical corticosteroids are a potential treatment option for FRPC patients.

Keywords: familial reactive perforating collagenosis, benign dermatosis, central keratotic plug, crateriform shape invagination, topical desoximetasone

Introduction

Reactive perforating collagenosis (RPC) is a rare benign dermatosis characterized by transepidermal elimination with extruding abnormal dermal collagen through the epidermis.[1] The patient's common clinical manifestation of RPC includes erythematous maculopapular lesions in the facial area and extremities, often covered by a central keratotic plug. Removal of the keratotic plug will reveal the crateriform shape of the erythematous papules.[2] The skin lesion distribution, size and depth of the keratotic plug, along with histopathological results that showed transepidermal elimination of keratin and deposition of abnormal collagen fibers are the basis for diagnosing RPC in patients.[3]

Based on its origin, RPC can be classified into two types: acquired (ARPC) and familial (FRPC).[1] As the terms implied, skin manifestations in ARPC are predisposed by systemic conditions, such as diabetes mellitus, neoplasms, autoimmune disorders, and liver and renal diseases; meanwhile, any of these underlying conditions are usually absent in FRPC patients.[1,4] The prevalence of FRPC in siblings and consanguineous families indicates the genetic origin of the disease.[2,4,5] Based on the number of reported cases, FRPC is the rarer form of RPC. To our knowledge, only 43 FRPC patients have been reported in the literature (see Table 1). In contrast to the adult onset of ARPC, the typical onset of

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Table 1 Summary of Other Reported Cases of Familial Reactive Perforating Collagenosis (FRPC)

Author, Country of Study, Year of Publication	Number of cases	Gender, Ethnicity	Age of Onset; Age at Presentation	Consanguinity	Affected Family Member(s)	Skin Manifestation; Suspected Precipitating Factor	Histopathological Results	Treatment	Outcome
Kanan MW, Kuwait, 1974[4]	7	Male, Kuwaiti (Sib 1a)	Early childhood (Age not specified); 26 years old	Consanguineous parents	Brother (Sib 1b) and sister (Sib 1c)	Multiple acne-like, maculopapular lesions on the forehead, sides of the face, chin, and fingers; Trauma and seasonal (Winter season).	Cup-shaped invagination filled with bundles of collagen and a solid mass of necrotic crust made up by numerous degenerated inflammatory cells, necrotic connective tissue, and keratinous flakes.	Emollient	NA
		Male, Kuwaiti (Sib 1b)	Early childhood (Age not specified); 20 years old	Consanguineous parents	Brother (Sib 1a) and sister (Sib 1c)	Multiple acne-like, maculopapular lesions on the forehead, sides of the hands and feet; Trauma.	Cup-shaped invagination filled with bundles of collagen and a solid mass of necrotic crust made up by numerous degenerated inflammatory cells, necrotic connective tissue, and keratinous flakes.	NA	NA
		Female, Kuwaiti (Sib 1c)	Early childhood (Age not specified); 28 years old	Consanguineous parents	Brothers (Sibs 1a and 1b)	Multiple acne-like, maculopapular lesions on the face, hands, and feet; Trauma and seasonal (Winter season).	NA	NA	NA
		Male, white Palestinian Arab (Sib 2a)	Early childhood (Age not specified); 40 years old	Consanguineous parents	Maternal grandfather, brothers (Sibs 2b and 2c), and sister (Sib 2d)	Two dome-shaped papular lesions on the dorsum of both thumbs; Trauma.	Only stated as comparable to incidental RPC	NA	NA
		Male, white Palestinian Arab (Sib 2b)	Early childhood (Age not specified); 32 years old	Consanguineous parents	Maternal grandfather, brothers (Sibs 2a and 2c), and sister (Sib 2d)	Lesions on the hands and feet; Trauma.	NA	NA	NA
		Male, white Palestinian Arab (Sib 2c)	Early childhood (Age not specified); 32 years old	Consanguineous parents	Maternal grandfather, brothers (Sib 2a and 2b), and sister (Sib 2d)	Lesions on the hands and feet; Trauma.	NA	NA	NA
		Female, white Palestinian Arab (Sib 2d)	Early childhood (Age not specified); 24 years old	Consanguineous parents	Maternal grandfather and brothers (Sibs 2a, 2b, and 2c)	Lesions on the hands and legs; Trauma.	NA	NA	NA
Kachhawa D, India, 1993[7]	1	Male, NA	15 years old; 21 years old	NA	Brother	Multiple, slow growing, discrete, hyperkeratotic, hyperpigmented papules on the facial area and limbs; Trauma.	Atrophic epidermal layer at the base of the plug, perforated by basophilic bundles of collagen from the underlying dermis.	NA	NA
Kumar V et al, India, 1998[8]	1	Male, NA	7 years old; 11 years old	No consanguinity	Brother and sister	Asymptomatic, multiple, discrete, hyperkeratotic papules on the facial area, both palms, and fingers with history of recurrencies; Trauma and seasonal (Winter season).	Hyperkeratinization and cup-shaped invagination of the dermis by degenerated collagen bundles.	NA	NA

Ramesh V et al, India, 2007[2]	10	Male, NA (Sib 3a)	9 months old; 21 years old	NA	Brother (Sib 3b)	Asymptomatic keratin-plugged nodules on dorsum of hands and feet, and over lower legs; Multiple superficial scars on face; Seasonal (More in winter season).	Cup-shaped crater filled with keratinous material and basophilic cellular debris and vertically oriented collagen fibers.	Topical Tretinoin (0.1%)	NA
		Female, NA (Sib 3b)	11 months old; 13 years old	NA	Sister (Sib 3a)	Asymptomatic keratin-plugged nodules on dorsum of hands, forearms, elbows, legs, forehead, and scalp; multiple superficial scars on both cheeks; Seasonal (More in winter season).	Cup-shaped crater filled with keratinous material and basophilic cellular debris and vertically oriented collagen fibers.	Topical Tretinoin (0.1%)	NA
		Female, NA	6 months; 4.5 years old	NA	NA	Small umbilicated lesions on the head, face, forearms, hands, buttocks, and lower legs. No significant scarring was seen; Seasonal (more in summer season).	Cup-shaped crater filled with keratinous material and basophilic cellular debris and vertically oriented collagen fibers; additionally, follicular involvement was identified, with hair shafts were noted among the debris.	Topical Tretinoin (0.1%) and zinc oxide	NA
		Female, NA	3 years old; 14 years old	NA	NA	Keratin-plugged papules on the periorbital area, dorsum of hands, forearms, and lower back. No significant scarring was seen; Seasonal (more in summer season).	Cup-shaped crater filled with keratinous material and basophilic cellular debris and vertically oriented collagen fibers. Follicular involvement was identified, with hair shafts were noted among the debris. Additionally, follicular and extrafollicular pathology were identified in the lesion on the back.	Topical Tretinoin (0.1%) and zinc oxide	NA
		Female, NA	1 year old; 4 years old	Consanguineous parents	Brother	Small umbilicated lesions on the facial area, dorsum of hands, trunk, legs, and feet. No significant scarring was seen; Seasonal (more in summer season).	Cup-shaped crater filled with keratinous material and basophilic cellular debris and vertically oriented collagen fibers.	Topical Tretinoin (0.1%) and zinc oxide	NA
		Female, NA	12 years old; 22 years old	NA	NA	Crateriform papules on the scalp, facial area, proximal limbs up to elbows and knees. No significant scarring was seen; Seasonal (more in summer season).	Cup-shaped crater filled with keratinous material and basophilic cellular debris and vertically oriented collagen fibers; additionally, follicular involvement was identified, with hair shafts were noted among the debris.	Topical Tretinoin (0.1%) and zinc oxide	Size and number of lesion reduced.
		Male, NA (Sib 4a)	1 year old; 8 years old	NA	Brother (Sib 4b)	Small papules on the facial area, forearms, and dorsum of hands. No significant scarring was seen; Seasonal (More in winter season).	Cup-shaped crater filled with keratinous material and basophilic cellular debris and vertically oriented collagen fibers.	Topical Tretinoin (0.1%)	NA
		Male, NA (Sib 4b)	9 months old; 7 years old	NA	Brother (Sib 4a)	Lesions on the dorsum of hands; Seasonal (More in winter season).	Cup-shaped crater filled with keratinous material and basophilic cellular debris and vertically oriented collagen fibers.	Topical Tretinoin (0.1%)	NA
		Female, NA (Sib 5a)	4 years old; 14 years old	NA	Brother (Sib 5b)	Keratin-plugged lesions on the scalp, facial area, pinnae, and limbs. Multiple scarring on the facial area; NA.	Cup-shaped crater filled with keratinous material and basophilic cellular debris and vertically oriented collagen fibers; additionally, follicular involvement was identified, with hair shafts were noted among the debris.	Topical Tretinoin (0.1%)	NA
		Male, NA (Sib 5b)	NA; 12 years old	NA	Sister (Sib 5a)	Lesions on the dorsum of hands. Scarring on the facial area and back; NA.	Cup-shaped crater filled with keratinous material and basophilic cellular debris and vertically oriented collagen fibers.	Topical Tretinoin (0.1%)	NA

(*Continued*)

Table 1 (Continued).

Author, Country of Study, Year of Publication	Number of cases	Gender, Ethnicity	Age of Onset; Age at Presentation	Consanguinity	Affected Family Member(s)	Skin Manifestation; Suspected Precipitating Factor	Histopathological Results	Treatment	Outcome
Bhat YJ et al, India, 2009[5]	10	Female, NA (Sib 6a)	8 years old; 19 years old	No consanguinity	Sister (Sib 6b)	Multiple, discrete, pruritic skin colored papules on the dorsum of hands; then, lesions became keratin-plugged, hyperkeratotic, and hyperpigmented; Trauma and seasonal (Winter season).	Central invagination with basophilic collagen and inflammatory cells, and parakeratosis.	Oral isotretinoin and emollient cream	Complete regression
		Female, NA (Sib 6b)	8 years old; 12 years old	No consanguinity	Sister (Sib 6a)	Multiple, discrete, asymptomatic skin colored papules on the dorsum of hands; then, lesions became keratin-plugged, hyperkeratotic, and hyperpigmented; Trauma and seasonal (Winter season).	Central invagination with basophilic collagen and inflammatory cells, and parakeratosis.	Oral isotretinoin and emollient cream	Complete regression
		Male, NA (Sib 7a)	1 year old; 9 years old	No consanguinity	Sister (Sib 7b)	Multiple, discrete, asymptomatic skin colored papules on the dorsum of hands; then, lesions became keratin-plugged, hyperkeratotic, and hyperpigmented; Trauma and seasonal (Winter season).	Central invagination with basophilic collagen and inflammatory cells, and parakeratosis.	Oral isotretinoin and emollient cream	Complete regression
		Female, NA (Sib 7b)	3 years old; 14 years old	No consanguinity	Brother (Sib 7a)	Multiple, discrete, asymptomatic skin colored papules on the dorsum of hands; then, lesions became keratin-plugged, hyperkeratotic, and hyperpigmented; NA.	Central invagination with basophilic collagen and inflammatory cells, and parakeratosis.	Topical tretinoin and steroid antibiotic cream	Temporary improvement
		Male, NA (Sib 8a)	6 years old; 11 years old	Consanguineous parents	Sister (Sib 8b)	Multiple, discrete, pruritic skin colored papules on the dorsum of hands; then, lesions became keratin-plugged, hyperkeratotic, and hyperpigmented; Trauma.	Central invagination with basophilic collagen and inflammatory cells, and parakeratosis.	Oral isotretinoin	Complete regression
		Female, NA (Sib 8b)	7 years old; 8 years old	Consanguineous parents	Brother (Sib 8a)	Multiple, discrete, asymptomatic skin colored papules on the dorsum of hands; then, lesions became keratin-plugged, hyperkeratotic, and hyperpigmented; Trauma and seasonal (Winter season).	Central invagination with basophilic collagen and inflammatory cells, and parakeratosis.	Topical tretinoin, emollient cream, and oral vitamin A supplement	NA
		Male, NA (Sib 9a)	8 years old; 38 years old	No consanguinity	Brother (Sib 9b) and sister (Sib 9c)	Large intensely pruritic lesions on the face and extremities that leaves scars when it heals; Trauma and seasonal (Winter season).	Central invagination with basophilic collagen and inflammatory cells, and parakeratosis.	Oral isotretinoin and emollient cream	Complete regression
		Male, NA (Sib 9b)	1 year old; 9 years old	No consanguinity	Brother (Sib 9a) and sister (Sib 9c)	Multiple, discrete, asymptomatic skin colored papules on the dorsum of hands; then, lesions became keratin-plugged, hyperkeratotic, and hyperpigmented; Trauma and seasonal (Winter season).	Central invagination with basophilic collagen and inflammatory cells, and parakeratosis.	Topical tretinoin and oral vitamin A supplement	NA
		Female, NA (Sib 9c)	2 years old; 7 years old	No consanguinity	Brothers (Sibs 9a and 9b)	Multiple, discrete, asymptomatic skin colored papules on the dorsum of hands; then, lesions became keratin-plugged, hyperkeratotic, and hyperpigmented; Seasonal (Winter season).	Central invagination with basophilic collagen and inflammatory cells, and parakeratosis.	Topical tretinoin and emollient cream	NA
		Male, NA	7 years old; 10 years old	No consanguinity	NA	Multiple, discrete, asymptomatic skin colored papules on the dorsum of hands; then, lesions became keratin-plugged, hyperkeratotic, and hyperpigmented; NA.	Central invagination with basophilic collagen and inflammatory cells, and parakeratosis.	Topical tretinoin and steroid antibiotic cream	Temporary improvement

Swagata TA et al, India, 2011[9]	1	Female, NA	2 years old; 19 years old	No consanguinity	Mother	Multiple hyperpigmented papules with central keratosis on the back and upper extremities; NA.	Cup-shaped invagination in the epidermal layer of the lesion, surrounded by acanthosis in the surrounding epidermis. At the base of invagination, altered collagen fibers were identified.	Topical Retinoids	NA
Verma R, India, 2013[10]	1	Male, NA	2 years old; 4 years old	No consanguinity	Brother	Multiple, dark-colored, intensely pruritic raised lesions on the face, elbows, and outer area of legs; Trauma.	Vertically oriented collagen bundles from the dermis that perforated epidermis, hyperkeratinized crater that were also filled with polymorphonuclear leukocytes and basophilic fibres.	Topical tretinoin 0.05% and intralesional injection of corticosteroids	Flattening of lesions, but new lesions appear
Sehgal VN et al, India, 2013[11]	1	Male, NA	8 years old; 8 years old	NA	Brother	Facial progressive asymptomatic skin eruptions as numerous skin-colored umbilicated keratin-plugged nodules; Non-traumatic.	Cup-shaped depression filled with basophilic debris and collagen fibers.	Narrow-Band Ultraviolet B	After 25 doses, complete skin regressions was identified, leaving atrophic scars and pigmented skin.
Kandhari R et al, India, 2014[12]	3	Female, NA (Sib 10a)	3 years old; 8 years old	No consanguinity	Sisters (Sibs 10b and 10c)	Multiple, well defined facial scars that preceded with multiple, erythematous, mildly pruritic, raised lesions, and a single crateriform lesion; Seasonal (Summer season).	Cup-shaped invagination of the epidermis filled with keratotic debris and vertically oriented collagen.	Topical tretinoin 0.05%	Lesions healed with scarring after 1 month
		Female, NA (Sib 10b)	3 years old; 11 years old	No consanguinity	Sisters (Sibs 10a and 10c)	Multiple, well defined facial scars that preceded with multiple, erythematous, mildly pruritic, raised lesions; Seasonal (Summer season).	Cup-shaped invagination of the epidermis filled with keratotic debris and vertically oriented collagen.	Topical tretinoin 0.05%	Lesions healed with scarring after 1 month
		Female, NA (Sib 10c)	3 years old; 13 years old	No consanguinity	Sisters (Sibs 10a and 10b)	Multiple, well defined facial scars that preceded with multiple, erythematous, mildly pruritic, raised lesions. Additionally, few discrete, skin-colored papules on the facial area were identified as well; Seasonal (Summer season).	Cup-shaped invagination of the epidermis filled with keratotic debris and vertically oriented collagen.	Topical tretinoin 0.05%	Lesions healed with scarring after 1 month
Pai VV et al, India, 2014[13]	2	Female, NA (Mother 1a)	Childhood (Age not specified); 30 years old	NA	Daughter (Daughter 1b)	Multiple mildly pruritic papules with scarring occurred when lesions resolved; NA.	Cup-shaped invagination with hair shaft in its crater, and filled with parakeratotic cells, basophilic materials, and collagen bundles.	Topical fluticasone, retinoic acid, and moisturizer	Minimal improvement after 2 months
		Female, NA (Daughter 1b)	1 year old; 10 years old	NA	Mother (Mother 1a)	Multiple mildly pruritic papules with scarring occurred when lesions resolved; Trauma.	Cup-shaped invagination with hair shaft in its crater, and filled with parakeratotic cells, basophilic materials, and collagen bundles.	Topical fluticasone, retinoic acid, and moisturizer	Minimal improvement after 2 months
Ghosh SK et al, India, 2016[3]	1	Male, NA	5 years old; 7 years old	Consanguineous parents	Grandfather	Multiple facial scars and pruritic skin eruptions over different parts of body; NA.	Extrusion of collagen fibers, inflammatory cell debris, and epidermal cup-shaped depression.	NA	NA

(*Continued*)

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Table 1 (Continued).

Author, Country of Study, Year of Publication	Number of cases	Gender, Ethnicity	Age of Onset; Age at Presentation	Consanguinity	Affected Family Member(s)	Skin Manifestation; Suspected Precipitating Factor	Histopathological Results	Treatment	Outcome
Tiwary AK et al, India, 2016[14]	1	Male, NA	Infancy (Age not specified); 13 years old	Consanguineous parents	Brother	Multiple, mildly pruritic, hyperpigmented papules and nodules face, the upper and lower extremities, particularly on the extensors; NA.	Epidermal invagination filled with basophilic collagen bundles, neutrophils, and orthokeratotic corneocytes; meanwhile, the dermal layer was infiltrated with lymphocytes and neutrophils.	Topical tretinoin, emollient, and oral antihistamines	NA
Rai AK et al, India, 2018[15]	2	Male, NA (Sib 11a)	NA; 7 years old	No consanguinity	Brother (Sib 11b)	Multiple, dark-colored, intensely pruritic keratin-plugged lesions on the face and extremities; Trauma.	Hyperkeratotic crater in the epidermis, and vertically oriented collagen bundles perforating both dermal and epidermal layers.	Topical tretinoin, emollient, oral antihistamines, and intralesional steroid injection.	Resolved existing lesions, but new lesions appear
		Male, NA (Sib 11b)	NA; 4 years old	No consanguinity	Brother (Sib 11a)	Multiple, dark-colored, intensely pruritic keratin-plugged lesions on the face and extremities; Trauma.	Hyperkeratotic crater in the epidermis, and vertically oriented collagen bundles perforating both dermal and epidermal layers.	Topical tretinoin, emollient, oral antihistamines, and intralesional steroid injection.	Resolved existing lesions, but new lesions appear
Albadri W et al, India, 2023[16]	2	Female, NA (Sib 12a)	3 years old; 11 years old	Fourth-degree consanguineous marriage	Sister (Sib12b)	Multiple, hyperpigmented keratin-plugged lesions on the upper and lower extremities, particularly on the extensors; NA.	Orthokeratotic plug with basophilic degenerated collagen fibers invaginated the epidermis, and vertically oriented collagen fibers infiltrated the dermis.	Oral isotretinoin and topical ammonium lactate	NA
		Female, NA (Sib 12b)	6 months old; 10 years old	Fourth-degree consanguineous marriage	Sister (Sib 12a)	Multiple, hyperpigmented keratin-plugged lesions on the upper and lower extremities, particularly on the extensors; NA.	Orthokeratotic plug with basophilic degenerated collagen fibers invaginated the epidermis, and vertically oriented collagen fibers infiltrated the dermis.	Oral isotretinoin and topical ammonium lactate	NA

Abbreviation: Sib, sibling.

FRPC is during infancy or early childhood. Concerning their symptoms, pruritus in FRPC is usually milder than ARPC.[6] Detailed pathogenesis of FRPC is not fully understood yet; thus, therapeutic approaches are symptomatic. One of the challenges that prevented the extension of our understanding of this unique entity is its rarity; hence, the report of new FRPC cases is essential. Here, we describe a case of FRPC in an Indonesian patient with adolescence-onset. The patient's paternal grandfather, uncle, and cousins acknowledged a history of similar symptoms. A literature review on previously published FRPC case reports was performed and summarized to discuss this unique and challenging condition comprehensively.

Case Presentation

A 20-year-old male was presented with generalized multiple discrete erythematous papules. Although these lesions could be identified in all regions of his body, many erythematous papules were acknowledged in specific areas, including the facial area, neck, abdominal, and extensor parts of his extremities. When carefully assessed, many of these papules were covered with central keratotic plugs. In papules where the keratotic plug was already removed, the actual crateriform shape of the lesion was identified. Each of these papules is sized 3–5 mm. The polymorphic appearance was acknowledged, suggesting a repetitive cycle of exacerbation and resolution in the patient. The latest exacerbation occurred in the last month before his visit, with the number of papules increasing. Resolved lesions from previous exacerbation could be identified by post-inflammatory hyperpigmented skin (Figure 1A–D).

The patient acknowledged the initial occurrence of the lesion seven years prior to his admission, suggesting that the age of onset is 13 years. Consanguinity was not acknowledged in the patient's family. Nevertheless, similar clinical manifestations were acknowledged in the patient's paternal grandfather, uncle, and cousins, suggesting an autosomal dominant inheritance pattern.

Physical examination of other body parts did not reveal anything remarkable. A routine and specific blood examination was performed to evaluate the presence of systemic diseases that could trigger exacerbation, such as diabetes mellitus, autoimmune disorders, and liver and renal diseases. Based on the blood examination results, no systemic diseases could be identified in the patient.

A punch biopsy specimen was obtained from one of the erythematous papules on the left back of his hand. Histopathological analysis based on H&E staining of the biopsy specimen revealed fibrocollagenous tissue, the keratinized epidermal layer that is covered by stratified squamous epithelium, crateriform appearance on the epidermis, and increased inflammatory lymphocytes (Figure 2A and B). Further histopathological examination by Masson trichrome staining confirmed collagen infiltration through the epidermis (Figure 2C and D). Based on the clinical and histopathological analyses, the patient was diagnosed with FRPC.

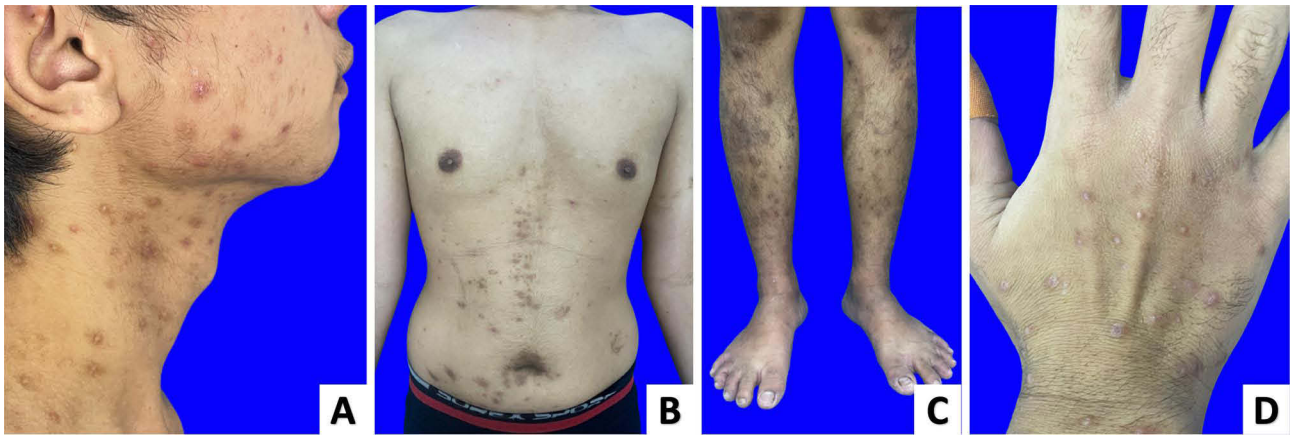

Figure 1 Dermatological symptoms in our familial reactive perforating collagenosis (RPC) patient. The patient presented with generalized multiple hyperpigmented papules, particularly in (**A**) the facial, neck, (**B**) abdominal, and (**C**) extensor parts of the feet and (**D**) hands. As a repetitive cycle of resolution and exacerbation had happened multiple times, resolved lesions could be identified by the occurrence of post-inflammatory hyperpigmented macules.

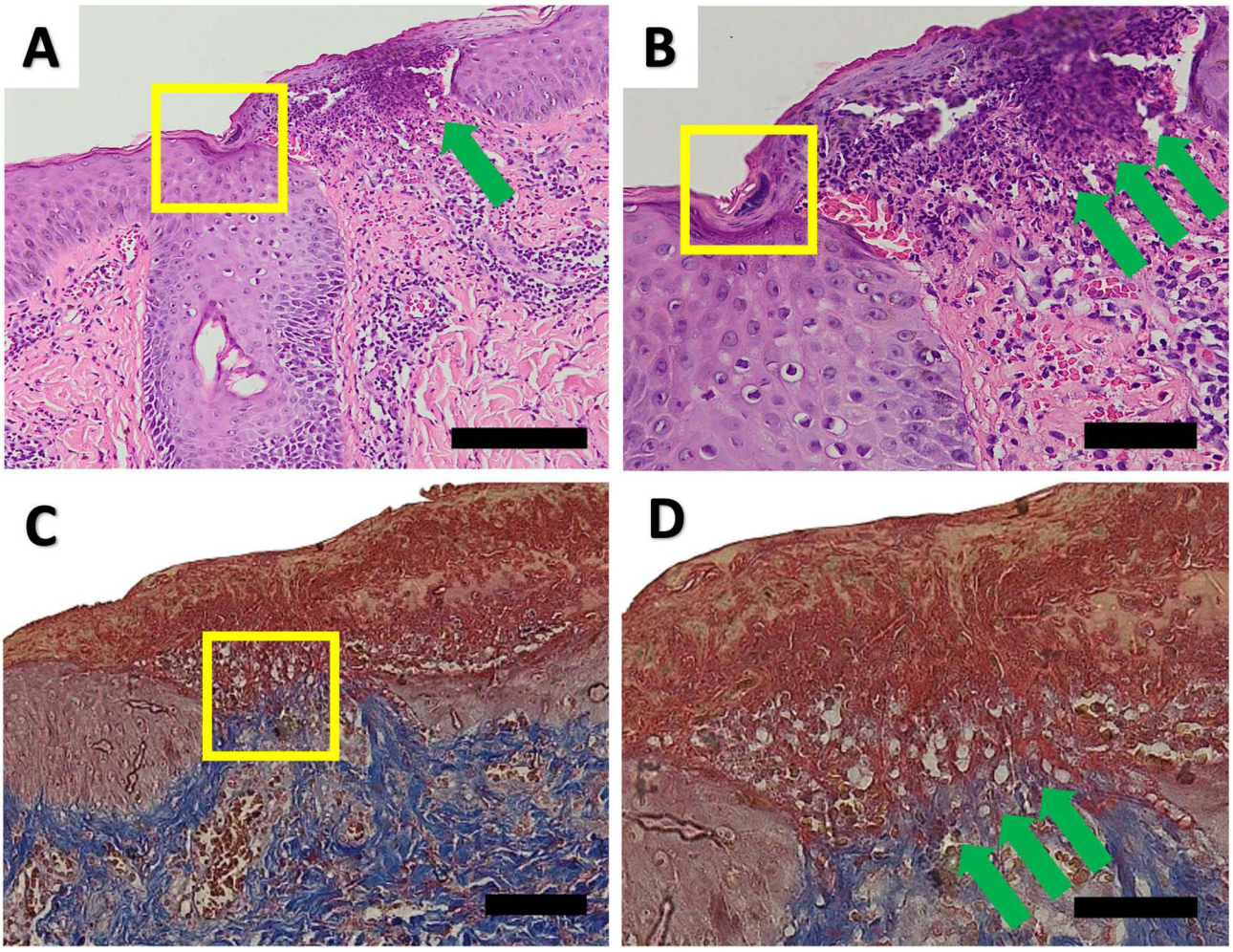

Figure 2 Histopathological results from FRPC patient. (**A** and **B**) H&E staining on the biopsy specimen identified fibrocollagenous tissue, crateriform appearance (yellow box), and enhanced inflammatory lymphocytes (green arrows). (**C** and **D**) Mason's trichrome staining showed an infiltrated area of the epidermal layer (yellow box) with collagen fibers from the underlying dermis (green arrows). (Scale bar (**A** and **C**): 100 μm; Scale bar (**B** and **D**): 50 μm).

He was treated with topical desoximetasone 0.25% cream applied 2–3 times daily. Evaluation at one week and one month after daily topical corticosteroid application revealed a near-complete resolution of skin papules, leaving post-inflammatory hyperpigmented macules.

Discussion

The term FRPC was first coined in 1974, based on the finding of 7 patients from 2 unrelated consanguineous families.[4] In contrast to ARPC, FRPC skin lesions are not associated with systemic disease; nonetheless, superficial trauma and atmospheric temperature have been suggested as the common predisposing factors in the exacerbation of FRPC.[3,4,8,10,13] As an inherited disease, FRPC is thought to have both autosomal recessive and autosomal dominant patterns of inheritance.[3,4] Consequently, a family history of FRPC suggests the diagnosis of FRPC. Like many other inherited diseases, the clinical manifestation of FRPC usually starts to occur during infancy or early childhood, as early as six months old.[2,16] Rarely, the onset of FRPC occurred during adolescence.[2,7]

Although our patient presented at the age of 20 years old, polymorphic patterns of his skin lesions indicated that he had already undergone a repetitive cycle of resolution and exacerbation of skin lesions. The adolescence-onset of FRPC suggests the possibility of an earlier hypothesis regarding the long quiescence of FRPC.[2,13,17] Like in many other FRPC cases, comorbidity(-ies) with systemic disease was not found in our patient. This suggests that the clinical manifestation

of RPC in familial cases is primarily due to the genetic alteration in the collagen-forming gene(s), leading to abnormal collagen formation through the epidermis.[13] In many FRPC cases, superficial skin trauma and atmospheric temperature have been acknowledged as precipitating factors. Nonetheless, like in the presented case, the precipitating factors in some previously reported FRPC cases are unknown; thus, its risk of recurrence is arguably high.[2,3,5,9,11,13,14,16]

To support the diagnosis of FRPC, a specimen from a punch biopsy on a lesion at the extensor area of the left arm was obtained and underwent histopathological analysis. Mason trichrome staining was performed on the specimen to ensure the collage content that formed the infiltrating fibrous tissue through the epidermis. Results from this staining confirmed the excessive collagen infiltration through the epidermis, supporting the diagnosis of FRPC. The histopathological characteristics identified in the specimen from our patient are similar to those suggested in previous case reports (see Table 1).

Although lesions in FRPC are self-limiting, previous reports have suggested using oral isotretinoin, topical tretinoin, steroid antibiotic cream, and emollient.[5,10,17] Previous studies described that medication with oral isotretinoin resulted in complete regression of symptoms in FRPC patients. Unfortunately, oral isotretinoin is not commercially available in our country; thus, daily application of topical desoximetasone 0.25% cream was advised, leading to a near-complete resolution that left post-inflammatory hyperpigmented macules. This result supports the use of topical corticosteroids as a potential treatment option for FRPC patients. In addition to oral and topical medications, one study suggested applying narrow-band ultraviolet B (UVB) light treatment in FRPC patients.[11] In our case, no further treatment was required since the results of topical corticosteroid application were satisfactory.

In addition to the clinical aspects described above, it is essential to acknowledge that this is Indonesia's first FRPC case report. It is interesting to notice that most of the previously reported FRPC patients lived in India.[2,3,5–17] (See Table 1) Based on our literature review, only 1 study reported the prevalence of FRPC outside of India.[4] Unfortunately, the ethnicity of these patients was not always described; nonetheless, it is tempting to speculate that ethnicity and geography might play essential roles in the pathogenesis of FRPC.

Conclusion

Based on the limited number of FRPC cases that have been reported, it is safe to conclude that FRPC is indeed a rare benign dermatosis in children. Preventing FRPC exacerbation is challenging, as precipitating factors are not always known. Furthermore, although it is self-limiting, various treatments can be applied to accelerate the resolution of FRPC and alleviate its symptoms. Future genetic studies of FRPC are required to unravel its pathophysiology.

Ethics Approval and Consent Statement

The institutional ethical review board of Dr Hasan Sadikin General Hospital has granted permission to publish the case details and images (Institutional approval No. DP.04.03/D.XIV.6.5/63/2024). The authors obtained written consent from the patient for their photographs and medical information to be published in print and online with the understanding that this information may be publicly available.

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Disclosure

The authors declare that this manuscript was written in the absence of any potential conflict of interest.

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