CASE REPORT
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A rare case of porokeratosis mibelli in 3-year-old boy

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ABSTRACT

Introduction: Porokeratosis mibelli is a rare, chronic, and slowly progressive genodermatotic characterized by abnormality of keratinization. The clinical manifestations are hyperkeratotic papules or plaques surrounded by a thread-like elevated border that widens centrifugally. Rarity of this case means the science behind it is underdeveloped. Thus, we present this case as an educational vehicle and to further develop the body of knowledge surrounding this disease.
Case report: We reported a 3 years old boy, suspect of porokeratosis mibelli. His mother complained of a few reddish spots and patches that sometimes itchy, healed into white and brownish patches with atrophic centers in the last 5 months before admission. Physical examination: few lesions on the back, waist, left side of buttock, back of the neck, and lateral upper left arm. Hypopigmented and hyperpigmented patches with atrophic centers, and erythematous plaques with central erosion were found. No abnormality of internal organs or laboratory findings. Histopathology examination showed epidermis contains of invagination rete malpighia filled keratin. Slightly thinned stratum germinativum with corneum contains basket weave pattern. Topical therapy with 1% 5-fluorourasil cream twice daily showed mild improvement after 10 days.
Conclusion: Although clinical examinations of the lesions on this patient did not fit the criteria of porokeratosis mibelli, however, the results of histopathology examination supported the diagnosis. Treatment with 1% 5-fluorourasil cream showed slow but progressive healing on the patient

Keywords: Children, porokeratosis mibelli, topical 1% 5-Flourourasil.
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INTRODUCTION

Porokeratosis mibelli is a keratinization disorder with diverse morphology. It manifests as hyperkeratotic papules or plaques circled by elevated border which widens centrifugally. Porokeratosis mibelli is a genetic disorder with several loci. The pathogenic mechanisms are still difficult to understand, however, prior studies suggested that males are more prone to this disorder compare to females and usually occurred during childhood.[1]

Classic porokeratosis started in the course of infancy or childhood as asymptomatic annular papules, with brown to skin color and annular border trait. Hyperkeratotic border is well-confined with a specific longitudinal furrow which generally more than 1 mm in height. Hyperpigmentation or hypopigmentation, with atrophic or anhidrotic center can be found. The diameter of lesion varies from millimeters to several centimeters but a giant lesion may develop to 20 cm. A giant porokeratosis mibelli rarely happened and mostly found on the lower leg and foot. Huge lesions are linked to higher probability of malignancy. Multiple lesions can appear locally (regionally) and unilaterally (generally). The circumstances could be familial and descended as an autosomal dominant mark. Skin lesions might appear endlessly.[1,2]

All forms of porokeratosis mibelli have similar histopathologic features, which is hyperkeratotic of stratum corneum and thin stake of parakeratotic cells with defective staining. At the lesions area, the cornoid lamella and granular layer may not present or greatly reduced or have normal thickness. Cornoid lamella could be preferential in clinical apparent even though it could also be found in other conditions like ichthyoses, viral wart and nevoid hyperkeratosis.[1,3]

Porokeratosis mibelli lesions are chronic, evolve slowly and although intense pruritus has been reported, it comparatively asymptomatic. Treatment for porokeratosis mibelli is not standardized yet and the result of therapy are variable. Various medication and therapy can be considered, such as potent topical preparation of steroids and keratolytic, including retinoids, 5-fluorouracil, imiquimod 5%, calcipotriol, and anthralin. Other modalities included advanced therapy such as cryotherapy and lasers, including carbon dioxide laser, pulsed dye laser and neodymium: yttrium-aluminum-garnet laser.[1] Although rarely happen, some cases reported spontaneous resolve of the lesions.[1-4] The disease is generally considered a benign process; however, malignant degeneration may occur such as squamous cell carcinoma, Bowen disease and basal cell carcinoma. Malignant degeneration has been described in all forms of porokeratosis, with a reported incidence of 7.5–11%.[2]

The rarity of this diagnosis, coupled

with the unavailability of standardized therapy, make the report and discussion of every case important for the development of its evidence-based care. Thus, in this case report, we present a rare case of porokeratosis mibelli with the aim to further develop the body of knowledge surrounding this disease entity.

CASE REPORT

A 3-year-old boy came to Out-patient Clinic of Dermato-Venereology Department, dr. Zainoel Abidin Hospital, Banda Aceh, with complaints of white and brownish patches with central atrophic, reddish patches that sometimes itchy on the back, waist, left of buttock, back of the neck, and upper right arm for 5 months before admission. The reddish spots and patches were coalescent and the margin of the lesion was mildly elevated. About a month later, the lesions healed spontaneously, left white and brownish patches with central atrophic healing. No history of application of chemical agent, medicine, or insect bite. No history of fever, excessive sun exposure, weight loss, hepatitis or complaints associated with his joint either. Consumption of medicine before the appearance of the patch such as prednisolone was denied. No decreased or loss of sensation on the reddish or white patch on his skin, neither history of hair loss or dry skin. The patient was born full term through spontaneous delivery, weighed 2800 gram, and was assisted by a doctor. The boy was the youngest child of five brothers.

Physical examination for general state was within normal range. Cutaneous examination revealed hypopigmented and hyperpigmented patches with central atrophic, erythematous plaque with central erosions, and multiple lesions present on the back, and several on the waist, left side of buttock, at the back of the neck and lateral upper left arm. There was no scaling, discharge or crusts from the lesions. No abnormalities of hematologic malignancy, anal anomalies or craniosynostosis was found.

Skin biopsy from lesions on the back revealed hyperkeratosis, parakeratosis, basket weave pattern. There was invagination rete malpighia filled with keratin. Fibro collagen tissue with mild perivascular lymphocytes was found in the dermis.

Considering the diagnosis of unusual porokeratosis, we initially treated the patient with 0,1% mometasone furoate ointment twice daily for the reddish patches. Mild improvement showed after 2 weeks, however, new lesions still appeared. Then, we changed the treatment to 1% 5-fluorourasil cream twice daily, and mild improvement showed after 10 days. After 1 month of treatment, the reddish lesion decreased, some lesions healed and involuted. One new non-itchy lesion with reddish patches on the back was found. Dermatology examination after 1 month of treatment showed hypopigmented and hyperpigmented patches with central atrophy, erythematous plaques with central atrophy and some lesions with hyperkeratotic, brownish crust margin.

DISCUSSION

We reported a case of rare porokeratosis mibelli in 3 years old boy. Diagnosis was established based on history taking, physical, and histopathology examination. Porokeratosis mibelli is an unusual chronic progressive dermatosis frequently found in India and elsewhere. A 4-years study in University Institute Hospital, Varanasi, reported 10 porokeratosis patients.[5] Another study reported 54 cases of porokeratosis (28 females and 26 males; age range 4 ± 89 years; mean age was 54.5 years) through the period of 1991 to 1998.[6]

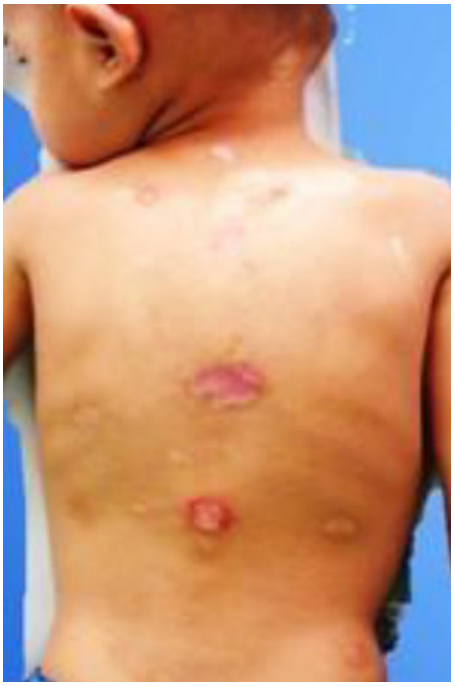

Figure 1. Hypopigmented and hyperpigmented central-atrophic patches with erythematous plaque at the center on the back, and several on the waist, left side of the buttock, back of the neck and lateral upper left arm.

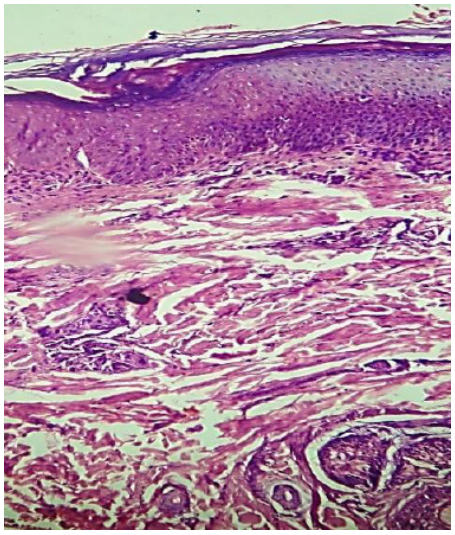

Figure 2. Histopathology examination support diagnosis of porokeratosis

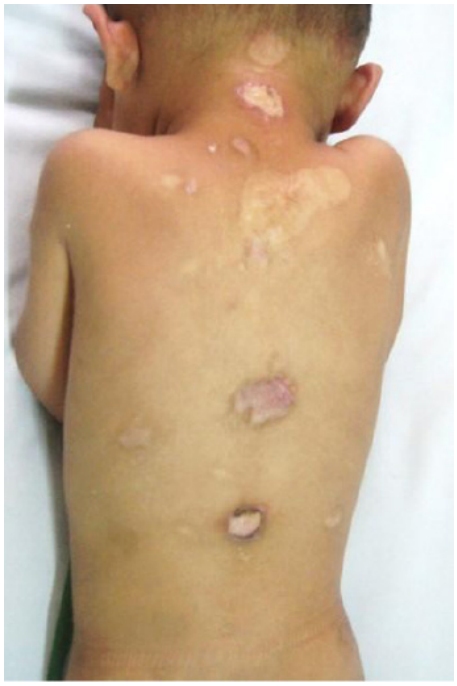

Figure 3. Follow up after 1 month of treatment with 5 FU

When the patient came for the first time, we decided porokeratosis mibelli as the diagnosis and acquired perforating dermatosis as the differential diagnosis because some lesions in this case healed with hyperpigmented and hypopigmented patches with central atrophic and erythematous plaque with erosion. The problem in this case was in establishing the diagnosis because clinically the lesion on this patient did not fit the criteria of porokeratosis, however, the results of histopathology examination support porokeratosis.

Porokeratosis mibelli is characterized by a single plaque or a few numbers of various size plaques with an atrophic center, which might develop for years up to 20 cm in diameter. Although other parts of the body may get affected (e.g., palms, soles, trunk, lips, genitals or mucous membranes), however, lesions are usually located unilaterally on the limbs. Porokeratosis mibelli usually occurs in childhood, affected by hereditary factors, and boys have higher incidence to develop the disease compare to girls (2.17:1).[2] In this case we found dominant hyperpigmented and hypopigmented patches with central atrophic and erythematous plaque with erosion on the back, back of the neck, lateral left upper arm and buttock.

One study reported unusual linear m with erosions at birth. There was no present of vesicles but erosions emerged up to 3 x 1 cm with early scarring at popliteal fossa. Majority of lesions consisted of small focal erosions or ulcerations with erythema and crust. After six weeks of 5-fluorouracil cream 1% applications on the lesions, it showed substantial enhancement and the spread of lesions have been stopped.[6]

The differential diagnosis for this case was acquired perforating dermatosis due to its similar appearances: erythematous papules and plaques with central atrophy, pruritic, majority located in the trunk and limb, except for the systematic symptoms that acquired perforating dermatosis has. It is associated with some systemic diseases such as diabetes with chronic renal failure tumors, chronic hepatopathies and HIV.[7] However, no systemic disease was found in this patient.

The histopathologic features in this case shown the epidermis contains with invagination rete malpighia filled keratin. The stratum germinativum was slightly thinned with corneum contains basket weave pattern. The histopathology examination supported the diagnosis of porokeratosis. From histopathologic pattern, stratum corneum was hyperkeratotic with thin stake of parakeratotic cells which defective staining. At the lesions area, the cornoid lamella and granular layer may not be present or greatly reduced or have normal thickness.[1] Histopathology for acquired perforating dermatosis reveals epidermal invagination with keratotic plug.[7]

There were some modalities of therapy for porokeratosis mibelli. First line therapy for porokeratosis is 5 fluorouracil cream and second line are calcipotriol, imiquimod, topical corticosteroid and topical retinoids.[2,4] We first treated the patient with 0,1% mometasone furoate ointment twice daily for the reddish patches. Mild improvement showed after 2 weeks, however, new lesions still appeared. One study reported the use of 100 mg dexamethasone in 5% dextrose with intravenous administration over 2-3 hours on three consecutive days in a month to treat the patient with porokeratosis. It successfully lead to cessation of new lesions appearance after the first pulse followed by regression of existing lesions after four pulses. There was successful resolution of 60% of the lesion and flattening of all lesions after 12 pulses. After the 18th pulse, all lesions have flattened and 80% of lesions has disappeared completely. There were no side effects reported.[8] A case report of a 5-year-old female patient with porokeratosis of mibelli treatment with topical tretinoin with the good results.[9] Another study reported treatment of porokeratosis with ingenol mebutate 0,015%, once a day for 3 consecutive days, separated by a month which resulted in reduced inflammation.[10]

Then, we changed the treatment to 1% 5-fluorourasil cream twice daily, and mild improvement showed after 10 days. After 1 month of treatment, the reddish lesion decreased, some lesions healed and involuted. Dermatology examination after 1 month of treatment showed hypopigmented and hyperpigmented patches with central atrophy, erythematous plaques with central atrophy and some lesions with hyperkeratotic, brownish crust margin. This is linear with prior study which reported that topical imiquimod 5% at night and 5-fluorouracil 1% every morning could successfully treat porokeratosis.[11]

CONCLUSION

We reported a rare case of porokeratosis mibelli on 3 years old boy. Although clinical examinations of the lesions on this patient did not fit the criteria of porokeratosis, however, the results of histopathology examination support porokeratosis mibelli. Initial treatment of topical steroid did show improvement, however, new lesions still occurred. Thus, we switched the treatment to 1% 5-fluorourasil cream twice daily and slow but progressive healing was shown after 10 days of treatment.

CONFLICT OF INTEREST

The authors declare that there is no conflict of interest regarding the manuscript.

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AUTHOR CONTRIBUTION

All authors are contributed equally to the content of the study.

ETHICAL STATEMENT

The informed consent was declared from patient's parent regarding the publication in this journal.

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