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CASE REPORT

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The first reported case of porokeratosis of mibelli in Syria: A case report and literature review

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ABSTRACT

Porokeratosis of Mibelli is a chronic dermatosis that has rare spontaneous remission and can degenerate into malignant tumors. Here we present, the first reported case of porokeratosis of Mibelli in Syria, which mimics clinically many skin disorders in a 48-year-old Syrian woman. The patient completely recovered after receiving three cycles of cryotherapy.

KEYWORDS

Porokeratosis; Cornoid lamella; Porokeratosis of Mibelli

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Introduction

Porokeratosis is a rare skin disorder, which can be inherited or acquired, with a broad array of clinical variants and unknown etiology [1-3]. It is characterized by the histopathological feature of clonal expansion of keratinocytes [2,4,5]. Porokeratosis is frequently described as erythematous-brownish plaques or well-delimited papules with keratotic ridge borders and variations of sizes and shapes [2,6]. Any part of the body can be affected by porokeratosis, including the oral mucosa [1]. Porokeratosis of Mibelli (PM) is the second most common type of porokeratosis [2]. It was first named and described in 1893 by Mibelli [7]. Here, we present the first reported case of this type in Syria.

Case Report

A 48-year-old Syrian woman, whose skin color is fair to beige, presented with a 5-year history of solitary lesion on her lower left leg. It started as an itchy papule, then slowly increased in size and became an asymptomatic plaque.

On physical examination, there was a well-defined, erythematous brownish plaque (measuring 1.5x2.5 cm) with central atrophy, the presence of a groove in a ridge-like border and overlying slight scales (Figure 1).

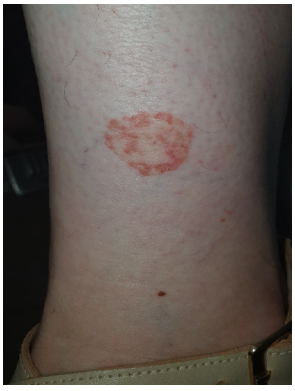

Figure 1. Solitary erythematous brownish plaque with central atrophy on the patient's lower leg.

The diagnosis of psoriasis was made by a physician two years ago, and the patient was treated with topical steroids for two weeks. However, the treatment with topical steroids was not effective. Nail changes, including brittleness and pitting, were observed on all fingernails. According to the patient, these changes developed years ago, and there was no history of nail biting or trauma. Besides, she has a history of arthralgia. Her mucous membranes and other systemic physical examinations were completely normal. The patient was employed as a nurse and denied any activity related to sun exposure or radiation. Her dressing routine mostly consists of pants that cover her legs. She has no history of allergies or any immunosuppressive treatments. Her family history of skin disorders was unremarkable. The dermoscopic examination of the lesion showed yellow circles with brown dots. A skin biopsy of the edge of the lesion was performed and the histopathological results revealed mild epidermal acanthosis with focal parakeratosis, hyperkeratosis, and keratin-filled epidermal invagination with angulated parakeratotic column. The epithelium deep into the column is vacuolated and devoid of the granular layer (cornoid lamella). Dyskeratotic cells are present with focal epithelial dysplasia. In the dermis, telangiectasia was seen with non-specific chronic inflammatory cells (Figure 2). Based on the patient's histopathological and clinical examination, the PM diagnosis was confirmed. The lesion was effectively treated after three cycles of cryotherapy.

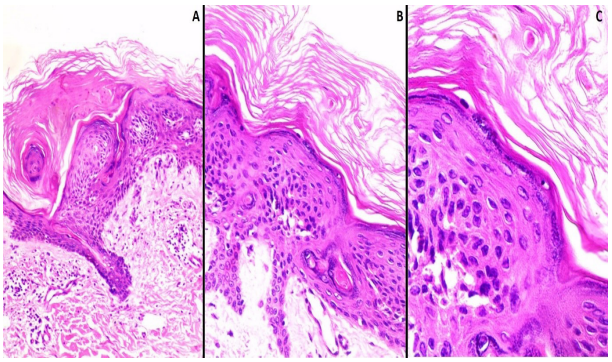

Figure 2. Histopathology of the lesion reveals: (A) Focal parakeratosis, hyperkeratosis, and keratin-filled epidermal invagination with angulated parakeratotic column (H&E x 40), (B) Mild epidermal acanthosis and cornoid lamella (the epithelium deep to the column is vacuolated and devoid of the granular layer) (H&E x 100), (C) Dyskeratotic cells are present with focal epithelial dysplasia, in addition, liquefactive degeneration of the basal cell layer with cleavage formation is present (H&E x 200).

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Discussion

PM is most common in adults and children and is more prevalent in males [2]. The most common variant of PM is characterized by one or more annular plaques with a hyperkeratotic or atrophic center that usually includes the limbs and trunk with a unilateral distribution [2,8]. Other parts, like the neck, shoulders, and genitals may also be affected [8]. Facial and mucosal lesions are rare to be involved with PM [8]. PM can present as a mild pruritic or asymptomatic lesion [9]. In our case, the lesion started as an itchy papule then it extended to an asymptomatic plaque. The incidence of PM is commonly reported in Caucasians and white-skinned individuals with a poor prognosis, particularly in Italian ethnicity, and it is rarely seen in people with dark skin [5,7].

Dermoscopic findings of porokeratosis generally reveal a homologous central white area that may contain a scar-like appearance or includes red-brown dots or globules, in addition to vascular structures with different patterns (linear, globular, or dotted vessels) [2,4].

The diagnosis of PM can be challenging for dermatologists due to its nonspecific clinical characteristics [9]. The differential diagnosis of PM can include psoriasis, Bowen's disease, and squamous cell carcinoma [8]. PM that we presented can mimic clinically many skin disorders including psoriasis, Bowen's disease, and pagetoid reticulosis (Woringer-Kolopp disease). Psoriasis was highly suspicious due to the above-mentioned presentation of the lesion that corresponds with psoriasis.

The histopathological examination along with the clinical examination and clinical history of our patient confirmed the diagnosis of PM. Actinic porokeratosis was also considered as a differential diagnosis, but due to the dressing routine of the patient and her clinical history that did not include any chronic sun exposure, the diagnosis of actinic porokeratosis was excluded.

The histopathological examination of PM reveals cornoid lamella, which contains a column of tightly piled parakeratotic cells stabilized in a keratin-filled epidermal invagination without a granular layer [9,10].

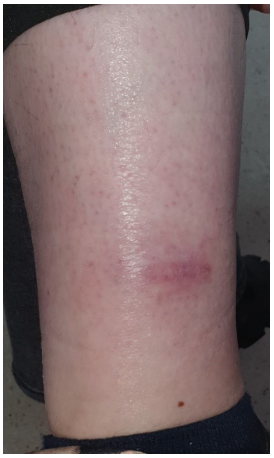
In the epidermis underlying the parakeratotic column, keratinocytes are arranged irregularly and have pyknotic nuclei with perinuclear edema [11]. Dyskeratotic keratinocytes can be found at the base of the cornoid lamella in the epithelium [11]. Ultraviolet radiation, immunosuppression, neoplastic disorders, infection, genetic factors, mechanical trauma, and drugs have all been considered to be causes of porokeratosis [2,7]. Also, it has been reported cases of PM that are associated with HIV infection, hematologic malignancies, organ transplantation, use of immunosuppressant drugs, and chemotherapy [4].

Many skin diseases can associate with porokeratosis such as psoriasis, lichen planus, lichen sclerosis, pemphigus, discoid lupus, alopecia areata, hidradenitis suppurativa, and pyoderma gangrenosum [2].

The cases of ungual porokeratosis reported to date are hallmarked by chronicity and a tendency to scarring of the nail apparatus with a tendency to destroy the nail plate [12]. Nail scarring occurs as the result of clinical conditions in which there is irreversible damage to the nail matrix or nail bed [12]. Clinically, nail scarring varies from mild (nail plate ridging) to moderate (splitting) to severe (pterygium and anonychia) [12]. PM has a high rate of malignant transformation (about 6.8-11% of cases) [1]. Malignant transformation has been noted in porokeratosis types with degeneration into basal cell carcinomas, squamous cell carcinomas, and Bowen's disease [9,13]. Large-sized lesions, long duration of appearance, and limbs involvements are considered risk factors [5,10]. Therefore, it is important to diagnose and treat PM as it can develop into malignant epithelial tumors [9].

Porokeratosis is usually refractory to treatment, with only 16% of patients obtaining a full response [2]. The response to various treatments is usually temporary with recurrent relapses [8]. Sun protection, moisturizers use, and regular follow-up to check for malignancies is preferable in many patients [1,2,5].

Topical application of retinoids, corticosteroids, imiquimod (5%), 5-fluorouracil (5%), and diclofenac gel (3%) as well as oral administration of acitretin, cryotherapy, electrocautery, surgery, and CO2 laser therapy agents have been used in the treatment of porokeratosis [5,6,9,10]. Surgery is the most effective treatment of PM; However, it may not always be the appropriate procedure [1]. An isolated porokeratosis lesion can be completely removed surgically, but depending on the location, number, and size of the lesion, this procedure may present technical challenges [13].



Cryosurgery is considered the option of choice compared to other destructive methods because of its simplicity, high rate of cure, short treatment time, low cost, and a few complications [6]. Anyhow, our patient achieved a complete recovery after receiving three circles of cryotherapy (Figure 3).

Figure 3. The lesion regressed completely after three cycles of cryotherapy, however, a scar developed at the site where the biopsy was taken.

Conclusions

This case emphasizes that the histopathology examination is the most important criterion for PM diagnosis. Early diagnosis and treatment of PM are important to decrease the associated malignancy with this disease. Although PM is more common in males and Caucasian children and adults, our patient is a Syrian female who was diagnosed with PM in her fourth decade. Since there are not enough data about the incidence of PM in Arab ethnicity and nations, we hope this case may add to the collective knowledge of this disease in Arabic patients. To our knowledge, this is the first case of porokeratosis of Mibelli to be reported in Syria.

Disclosure Statement

No potential conflict of interest was reported by the author.

References

1. Gu CY, Zhang CF, Chen LJ, Xiang LH, Zheng ZZ. Clinical analysis and etiology of porokeratosis. Exp Ther Med. 2014;8(3):737-741. https://doi.org/10.3892/etm.2014.1803
2. Vargas-Mora P, Morgado-Carrasco D, Fustà-Novell X.

https://doi.org/10.61577/amsd.2024.100007

Porokeratosis: a review of its pathophysiology, clinical manifestations, diagnosis, and treatment. Actas Dermosifiliogr. (English Edition). 2020;111(7):545-560. https://doi.org/10.1016/j.adengl.2020.08.005
3. Alexis AF, Busam K, Myskowski PL. Porokeratosis of Mibelli following bone marrow transplantation. Int J Dermatol. 2006;45(4):361-365. https://doi.org/10.1111/j.1365-4632.2006.02509.x
4. Ottoni LD, Kakizaki P, Pinheiro RR, Sittart JA, Valente NY. Porokeratosis of Mibelli in an HIV-positive patient. An Bras Dermatol. 2016;91:131-133. https://doi.org/10.1590/abd1806-4841.20164253
5. Sertznig P, Von Felbert V, Megahed M. Porokeratosis: present concepts. J Eur Acad Dermatol Venereol. 2012;26(4):404-412. https://doi.org/10.1111/j.1468-3083.2011.04275.x
6. Dereli T, Ozyurt S, Ozturk G. Porokeratosis of Mibelli: successful treatment with cryosurgery. J Dermatol. 2004;31(3):223-227. https://doi.org/10.1111/j.1346-8138.2004.tb00659.x
7. Singh N, Chandrashekar L, Kumar N, Kar R, Thappa DM. Classic porokeratosis of Mibelli. Indian Dermatol Online J. 2014 1;5(Suppl 2):S130-S131. https://doi.org/10.4103/2229-5178.146193
8. Ferreira FR, Santos LD, Tagliarini FA, Lira ML. Poroqueratose de Mibelli-revisão da literatura e relato de um caso. An Bras Dermatol. 2013;88:179-182. https://doi.org/10.1590/abd1806-4841.20132721
9. Giuliodori K, Campanati A, Ganzetti G, Conocchiari L, Cataldi I, Simonetti O, et al. The successful off-label use of photodynamic therapy for classic porokeratosis of Mibelli: case report. Dermatol Ther. 2011;24(5):501-504. https://doi.org/10.1111/j.1529-8019.2012.01473.x
10. Weidner T, Illing T, Miguel D, Elsner P. Treatment of porokeratosis: a systematic review. Am J Clin Dermatol. 2017;18:435-449. https://doi.org/10.1007/s40257-017-0271-3
11. Reed RJ, Leone P. Porokeratosis—A mutant clonal keratosis of the epidermis: I. Histogenesis. Arch Dermatol. 1970;101(3):340-347. https://doi.org/10.1001/archderm.1970.04000030084014
12. Yendo TM, Gabbi TV, Nico MM. Porokeratosis of the Nail Unit: Case Series and Review. Skin Appendage Disord. 2021;7(6):489-492. https://doi.org/10.1159/000516304
13. Harrison S, Sinclair R. Porokeratosis of Mibelli: successful treatment with topical 5% imiquimod cream. Aust J Dermatol. 2003;44(4):281-283. https://doi.org/10.1046/j.1440-0960.2003.00010.x

https://doi.org/10.61577/amsd.2024.100007