Case Report

Recalcitrant Digital Porokeratosis of Mibelli: A Successful Surgical Treatment

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Abstract

Context: Porokeratosis of Mibelli (PM) is a rare, benign, asymptomatic, epidermal hyperkeratinization dermatitis that is characterized by annular plaque that expands through the edges and leaves an atrophic center. Many therapies have been attempted for the treatment of PM, but none of these have given satisfactory results. The efficacies of treatment options are limited, and currently there is no gold standard. **Case Report:** This paper reports the case of a 22-year-old female with 3-years history of PM, who had not responded to routine therapies like topical corticosteroids, topical tretinoin, topical salicylic acid, and various emollients and keratolytic agent. Cryosurgery and laser ablation did not have acceptable response. We used surgical treatment with successful cosmetic outcome. **Conclusion:** The treatment of PM should be individualized considering the aesthetic and functionality, and the patient's preferences. Complete surgical excision for isolated digital PM had good results.

Keywords: Cornoid lamella, Porokeratosis of Mibelli (PM), Treatment

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Introduction

Porokeratosis of Mibelli (PM) is a rare, benign, asymptomatic, epidermal hyperkeratinization lesion that is characterized by annular plaque that expands through the edges and leaves an atrophic center.[1] It can affect any part of the body but is mostly seen on the trunk and extremities, and is more frequent in males.[1,2] Involvement of an individual digit of the hand, in isolation, is not common in the literature.[3] There are different approaches to treatment of PM including oral and topical retinoids, topical and intralesional corticosteroids, vitamin D3 analogs, keratolytic agents, CO_2 laser, dermabrasion, cryotherapy, topical 5-fluorouracil, and surgical option. None of these modalities are appropriate for all cases.[4]

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This report describes the successful surgical treatment of a recalcitrant case of histopathologically diagnosed PM in a 22-year-old female with a split-thickness skin graft (STSG).

Case Presentation

A 22-year-old female was presented to Al Zahra hospital's dermatology clinic, Isfahan, Iran, with an approximately 3-years history of a solitary, slowly progressive, scaly plaque, with a slightly raised hyperkeratotic border on the second digit of her hand [Figures 1 and 2]. She did not have a history of any underlying disease. Skin examination of the second digit indicated a 3-4 cm hyperkeratotic plaque, with a distinct raised annular border and an atrophic center that was compatible with PM. Histopathological analysis showed "cornoid lamella" and confirmed the diagnosis of PM. The lesion was refractory to common treatments. She had not responded to routine therapies like topical corticosteroids, topical tretinoin, topical salicylic acid and various emollients, and keratolytic agent. Cryosurgery and laser ablation did not have acceptable response. Recently the lesion became increasingly large, pruritic, and cosmetically displeasing that induced psychological stress, shame, and anxiety

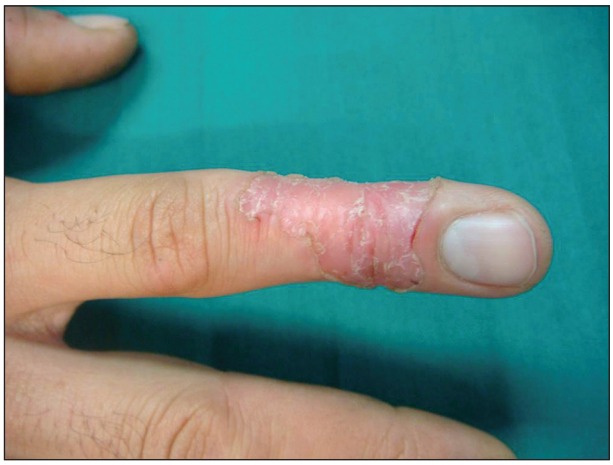

Figure 1: PM (before treatment); raised annular hyperkeratotic border on the digit

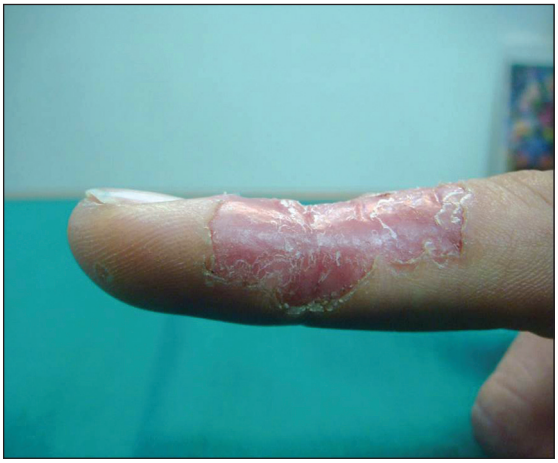

Figure 2: PM (before treatment); solitary, slowly progressive, scaly plaque with a hyperkeratotic border

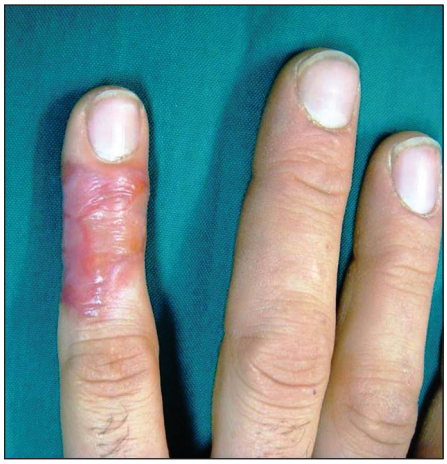

Figure 3: PM (after treatment); complete surgical excision and grafting

about the general appearance, along with cosmetic concern. Due to the recalcitrance of lesions to common topical treatments after 2 years of appropriate treatment, it was decided that the patient would get treated with excision of the plaque and application of a STSG from her thigh. After a 4-month follow-up, hyperkeratotic lesion was resolved; there was no recurrent lesion and an acceptable result was obtained [Figure 3].

Discussion

Many therapies have been attempted for the treatment of PM, but none of these have given satisfactory results.[4] The efficacy of these methods is limited, and there is currently no gold standard.[4] Topical 5-fluorouracil cannot be used for disseminated lesions since an inflammatory response is likely to occur.[5] Oral etretinate has been associated with undesirable side effects, the need for a prolonged period of therapy, and rapid recurrence.[4] Cryotherapy and laser therapies, including CO_2 laser, have mostly been effective, but these treatments may have recurrences and typically require anesthesia, wound care, and a lengthy recovery period.[4] None of the currently available therapies is effective in all cases, and this situation exposes the patient to multiple treatment cycles.[6] The case at hand was recalcitrant to routine medical treatments, and due to the patient's symptoms and cosmetic problems we decided to treat her with excision of the plaque and STSG. Complete surgical excision, as in our case, should offer a cure, although in widespread cases, nonoperative treatment may be more appropriate.[3]

Conclusion

The treatment of PM should be individualized considering the aesthetic and functionality and the patient's preferences. In our case, surgical treatment of isolated digital PM had good results.

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