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Case Report

Local subungual excision and graft use in patients with incontinentia pigmenti: A case report and review of the literature☆

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ABSTRACT

Incontinentia pigmenti (IP) is a rare genetic skin disorder with an X-linked dominant inheritance that is seen almost exclusively in females. Subungual keratoacanthoma (SUKA) is a rare benign neoplasm of the nail bed associated with IP. The recommended initial treatment of SUKA is conservative thereby sparing the digit in the form of excision and curettage. However, definitive treatment involves terminalisation of the affected digit. We report a case in a 54-year-old woman with IP who developed SUKA who was treated with local subungual excision and subsequent skin grafting who had previous terminalisation of other digits for the same condition.

Subungual keratoacanthoma is a rare benign neoplasm that is typically treated with curettage and excision at the initial stages with terminalisation as the definitive treatment. Given the highly positive outcome in this case with minimal sacrifice to the dexterity of the patient and preservation of digit length, we feel that local subungual excision and skin grafting should be strongly considered as an alternative to amputation in moderate to severe cases of the condition.

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Introduction

Incontinentia pigmenti (IP) is a rare genetic skin disorder with an X-linked dominant inheritance that is seen almost exclusively in females.[1] IP is caused by a mutation of the IKBKG/NEMO, which leads to a loss of function of NF-kB, thereby leaving cells susceptible to apoptosis.[2] IP is a multi-system disorder with associated extra-cutaneous manifestations affecting the skin, eyes, hair, teeth and central nervous system.[3–5]

Subungual keratoacanthoma (SUKA) is a rare benign neoplasm of the nail bed associated with IP. SUKA typically presents as a painful, rapidly growing lesion of the terminal phalanx with early underlying bone destruction.[6] The recommended initial treatment of SUKA is conservative thereby sparing the digit in the form of excision and curettage. However, definitive treatment involves terminalisation of the affected digit.[7] We report a case of local subungual excision and subsequent skin grafting in the treatment of SUKA in patient with IP who had previous terminalisation of other digits for the same condition.

Case report

A 54-year-old right hand dominant female with IP, initially presented to a tertiary centre with discoloured patches of skin of the nail bed. She later developed painful lesions on the fingers involving the distal phalanx, identified by MRI and histopathology as subungual keratoacanthomas.

In **2011**, the patient had both her left little and ring fingers terminalised, at the PIPJ and DIPJ respectively and her right middle finger terminalised at the DIPJ. Despite this surgical intervention, the pain and swelling continued in the other fingers.

In **2013**, ablation or excision of the underlying nail bed with a subsequent skin graft was suggested as a more conservative alternative.

Between **2013** and **2016**, the patient has undergone this procedure several times for the remaining fingers – the thumb, index and middle finger of the left hand and thumb, index, ring and little finger of the right hand with replacement skin grafts taken from the right groin region (Figures 1 and 2).

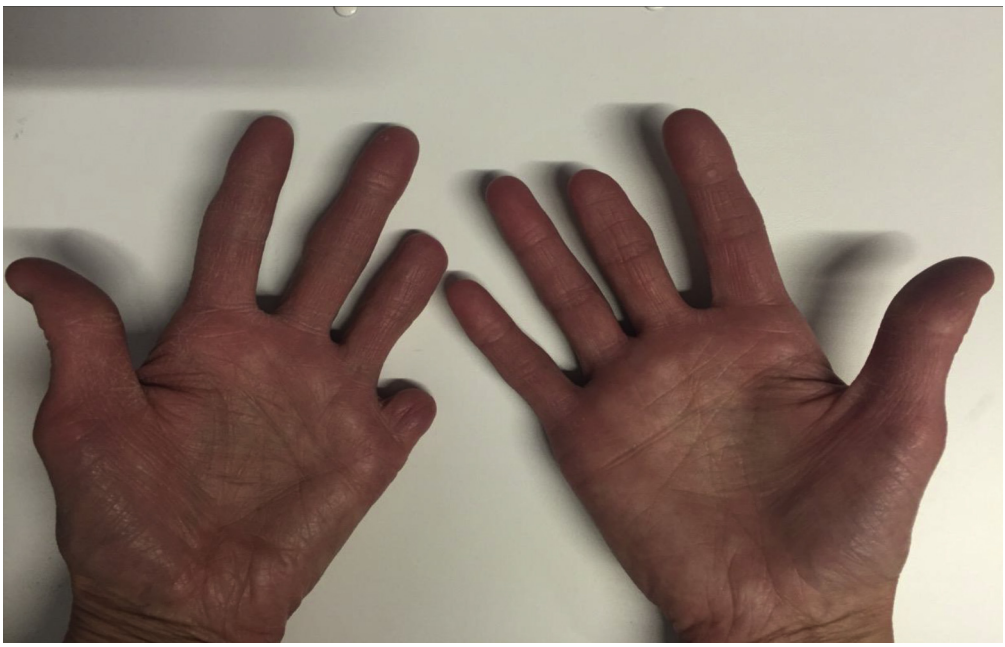

Figure 1. Palmar aspect of the hand.

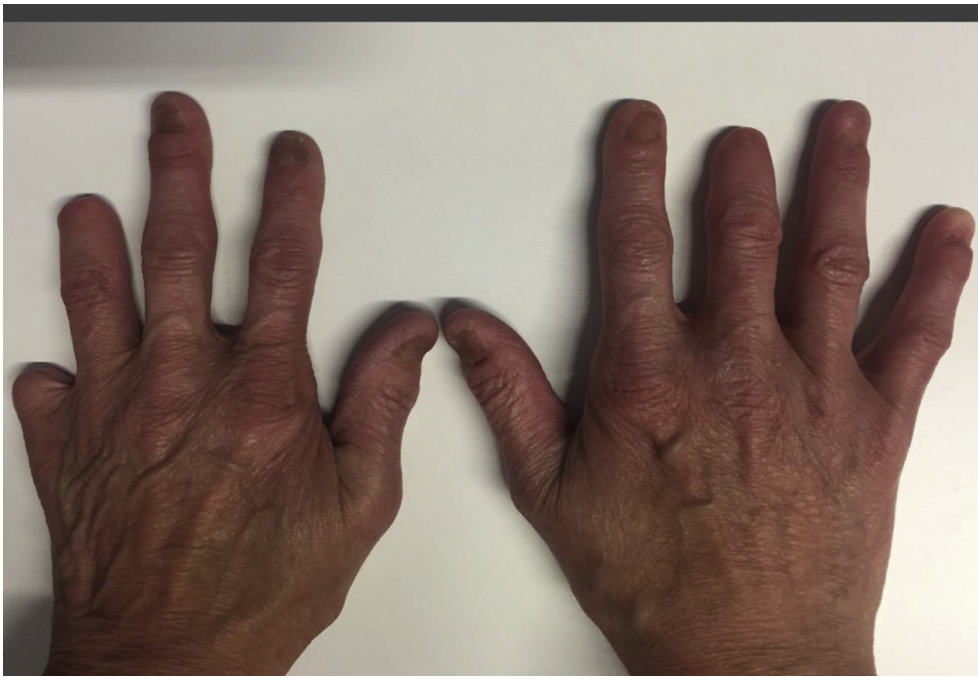

Figure 2. Dorsal aspect of the hand.

Recent clinic reviews have been largely positive, with all fingers healing well, minimal sacrifice to dexterity and no remaining areas of IP affecting the patients' quality of life.

Discussion

First-line surgical treatment for SUKA is widely considered to be curettage and local excision, as this has proven to be highly successful in current literature.[8] However, some surgeons prefer to use Mohs micrographic surgery as the first-line procedure, as the rates of recurrence are lower, due to its highly accurate margins of excision. Failing this, amputation is currently the only recognised definitive surgical treatment in patients with multiple recurrences, underlying bony disruption or suspected squamous cell carcinoma.[8] SUKA may show locally aggressive behaviour, hence are at times mistaken for squamous cell carcinoma, but does not metastasize.[9] A study by Pellegrini and Tompkins[8] reviewed 18 cases in the literature, which revealed terminalisation as the definitive form of treatment in only 8 cases, with the remaining cases treated via curettage or excision. It was suggested that aggressive ablative surgery as initial treatment was discouraged. A further study by Baran and Goettmann[10] revealed 12 cases of distal digital keratoacanthoma (DKA) and their treatment demonstrating 9 cases treated via surgical removal, 1 case by excision and curettage, 1 by terminalisation and 1 case by systemic methotrexate. It has been debated that subungual tumours found in IP should be considered as true keratoacanthoma whereas DKA is a different entity. Until now, the combination of local subungual excision and subsequent skin grafting has not been reported.

Conclusion

Subungual keratoacanthoma is a rare benign neoplasm that is typically treated with curettage and excision at the initial stages with terminalisation as the definitive treatment. To the best of our knowledge we report the first case in which SUKA in an IP patient was treated with local excision and skin graft. Given the highly positive outcome in this case with minimal sacrifice to the dexterity of the

patient and preservation of digit length, we feel that this procedure should be strongly considered as an alternative to amputation in moderate to severe cases of the condition.

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