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Case Report

Squamous cell carcinoma in rare case of Huriez Syndrome: The role of distant flaps

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ABSTRACT

Context: Huriez syndrome is a rare de rmatological condition characterized by severe sclerotic and atrophic changes in the extremities (hands and feet) and an increased tendency to develop squamous cell carcinomas, with no established gold standard for the surgical treatment of these patients, who are difficult to manage due to the inability to perform reconstructions using local flaps.
Clinical Case: We report the case of a patient with severe Huriez syndrome who had developed SCC in both the hands and foot over time. After multiple surgeries at other centers, all resulting in recurrences, we planned wide excisions followed by reconstruction using distant flaps. The postoperative course for both reconstructive procedures was complication-free. Follow-up revealed not only an excellent reconstructive outcome but also highlighted the untapped potential of the flap in managing this condition: thanks to its independent vascularization, the flap remained unaffected by the surrounding diseased skin and significantly improved the scaly, atrophic appearance of the affected areas.
Discussion: Huriez syndrome is a dermatological condition characterized by a predisposition to developing squamous cell carcinomas. In these patients, it is essential to plan wide excisions that ensure oncological radicality. The reconstruction must be carefully planned, and microvascular flaps can be prioritized.

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Conclusion: The radial forearm flap has proven effective for reconstruction in individuals with SCC associated with Huriez syndrome. The skin of the flap not only remained unaffected by the surrounding pathology but also induced a previously undescribed benefit on the diseased skin. This oncological-reconstructive approach could become the gold standard in treating these patients.

Introduction

Huriez syndrome (HRZ), firstly described by Huriez et al.,[1] is a rare genetic skin disorder featuring sclerotic and atrophic alterations in the hands and feet and a significantly elevated risk - up to 100 times higher - of developing cutaneous squamous cell carcinoma (cSCC) with a poorly differentiated histotype.[2] The treatment of squamous cell carcinomas in patients with Huriez syndrome poses a major challenge for plastic and orthopedic surgeons. The rapid growth of these soft tissue tumors requires reconstruction using flaps or grafts. However, using local flaps from nearby areas is often impractical, as these regions are also prone to developing additional precancerous and cancerous lesions. We present here a patient with severe Huriez syndrome who experienced malignant degeneration of the skin on hands and feet and underwent multiple reconstructions and amputations. The role of pedicled and free flaps was crucial after oncological resection in preserving function, preventing recurrences and improving the quality of the surrounding skin.

Case report

In February 2020, a 55-year-old man was admitted to the Hand Surgery Unit at the Jewish Hospital of Rome with a diagnosis of cutaneous squamous cell carcinoma (cSCC) in the hands, linked to a severe case of Huriez syndrome. Physical examination showed up a notable thickening of the skin on palms and soles, underdeveloped fingernails and a pronounced form of palmoplantar keratoderma. His medical history revealed multiple surgeries to remove squamous cell carcinomas, resulting in increasingly extensive amputations that significantly impacted his quality of life.

Right hand

In 2014 the patient underwent amputation of the second finger of the right hand for SCC. The histological diagnosis was poorly-differentiated and ulcerated SCC with resection edges free of disease. Four years later, multiples recurrences were treated with additional and sequential resections, with a residual non-functional stump. In February 2020 when he came to our attention, he had an ulcerated recurrence of SCC, involving the first ray and the wrist **(**Figure 1**).** An oncological staging was performed confirming a locally aggressive disease, without nodal or organ metastasis. A new biopsy confirmed the histological pattern of invasive, ulcerated, SCC. In agreement with the patient we decided to perform total amputation of the residual stump, proximal to the wrist joint.

Left hand

In 2015 the patient underwent amputation of the second finger of the left hand for excision of a SCC. The histological diagnosis was poorly-differentiated and ulcerated SCC with resection edges free of disease. While preparing for surgery on the right hand, the patient mentioned the appearance of a rapidly growing lesion on the third finger of his left hand (Figure 1). It was decided to remove this lesion after completing the controlateral amputation. Our focus was on achieving oncological clearance and preserving function in the remaining hand. The resection involved removing the skin with

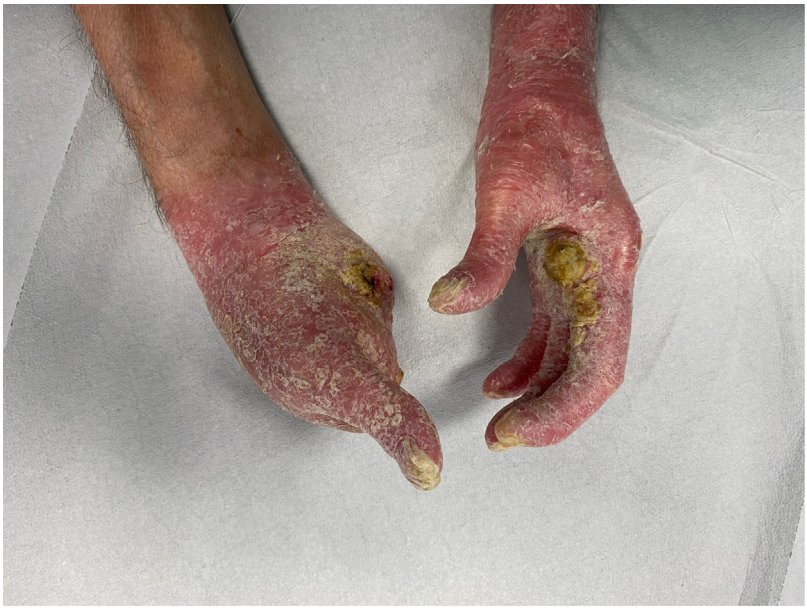

Figure 1. Right hand presenting an ulcerated recurrence of SCC of the residual stump, involving the first ray and wrist. Simultaneously, left hand presenting a rapidly growing lesion on the third finger of his left hand.

wide margins, the radial neurovascular bundle, and the flexor digitorum superficialis (FDS) and flexor digitorum profundus (FDP) tendons. A graft was used to reconstruct the profundus tendon, and a pedicled radial forearm flap was prepared for tissue coverage. Since the forearm skin was unaffected by Huriez's disease, the flap's skin paddle was pliable, well-nourished, and free from sclerotic changes. The postoperative recovery was smooth, and the patient was discharged five days later.

Right foot

Another rapidly growing lesion appeared on the medial surface of right foot in October 2022, strongly suggestive of squamous cell carcinoma (Figure 2). After biopsy, a wide excision was performed and a free radial forearm flap harvested from the right arm (with a total hand amputation) was used for reconstruction (Figure 3). Similarly, the skin of the right forearm was not affected by Huriez's disease. The histological diagnosis was poorly differentiated SCC. The patient was discharged after 5 days without complications.

Follow up

Follow-ups were scheduled at 3, 6, and 9 months, followed by oncological check-ups every year. Four years later, there is no sign of recurrence. The patient has achieved excellent functional recovery. The skin areas covered by the flap remained unaffected by the surrounding diseased skin, likely due to the fact that the reconstructed tissues had their own vascular supply. Interestingly, the areas around the radial forearm flaps on both the hand and foot exhibited unexpected improvements, showing enhanced trophism and local conditions (Figure 4).

Discussion

Huriez syndrome is a dermatological condition that often predisposes individuals to the development of skin carcinomas, especially squamous cell carcinoma. The surgical management of these carcinomas in these delicate patients presents a significant challenge, and a gold standard that has yet to be described in literature.[3]

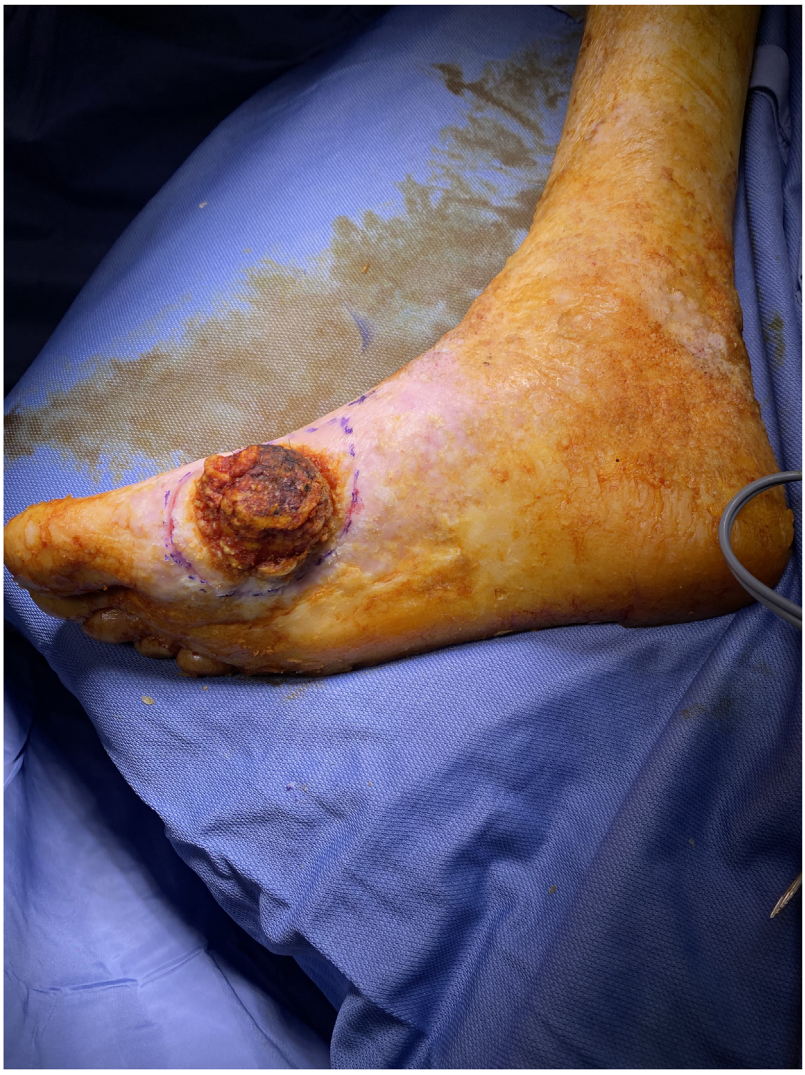

Figure 2. Right foot presenting another rapidly growing lesion, strongly suggestive of squamous cell carcinoma.

A first crucial consideration in these individuals is the imperative of achieving oncological radicality. Due to their predisposition to high rates of local recurrences, surgeries must be conducted with wide margins (>6 mm as reported in AIOM Guidelines, 2024).[4] The reconstructive approach must also be carefully evaluated. In normal skin conditions, when primary closure is not feasible after the removal of skin carcinomas, reconstructive techniques involving local flaps are commonly employed[5,9] providing a dependable method for replacing "like with like".[6] A primary challenge for patients with Huriez syndrome lies in the inability to utilize skin from surrounding regions for reconstruction, because the adjacent skin is also affected by the syndrome and prone to developing additional neoplastic lesions, making it necessary to increase the surgical complexity.

Reconstruction of the hand - To reconstruct wide tissue loss of the hand, pedicled flaps like the radial forearm flap or the posterior interosseous flap are commonly employed. The pedicled radial forearm flap is preferred for addressing extensive palmo-digital soft tissue defects and typically yields

Figure 3. Intraoperative photo of the free radial forearm flap, harvested from the right harm, corresponding to the amputation stump, then placed to reconstruct the tissue loss from the demolitive procedure.

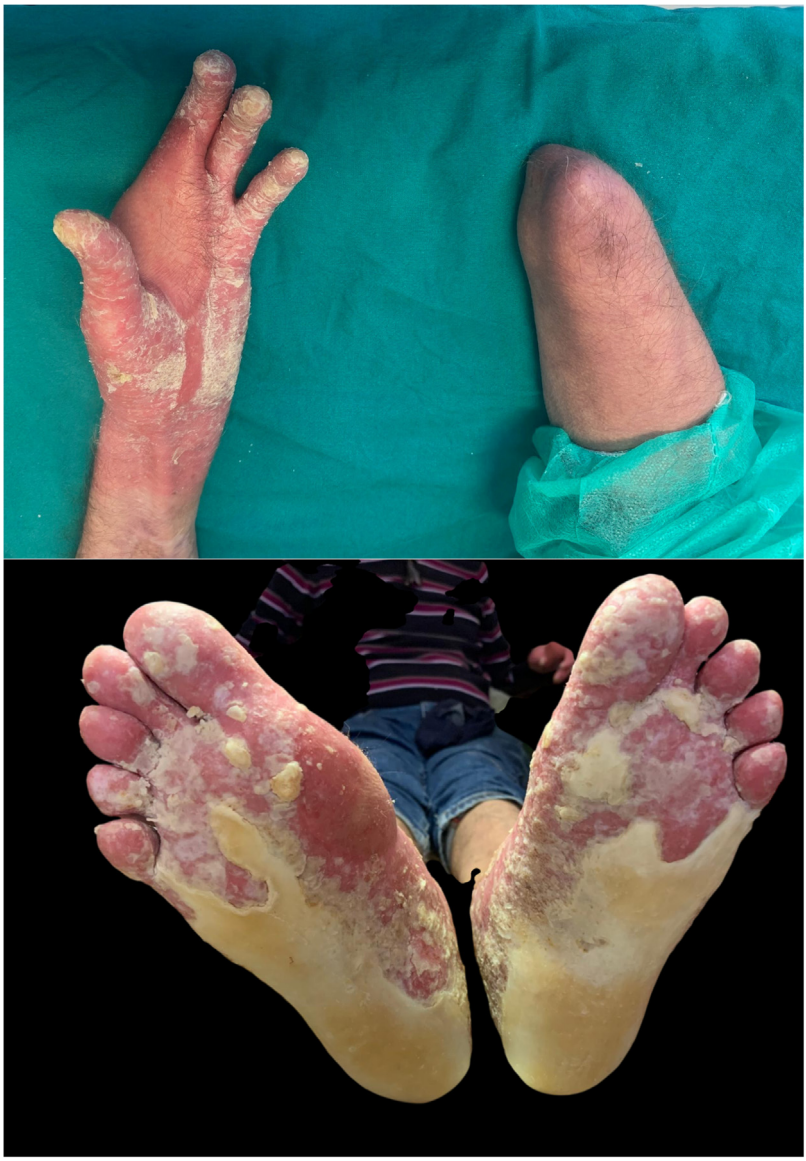

Figure 4. Postoperative photo four years after the surgery: hand and foot areas around radial forearm flaps showed up improvements in local conditions with enhanced trophism.

excellent outcomes. However, it is known for compromising the radial vascular supply and poor cosmetic appearance at the donor site.[7] Regarding the posterior interosseous artery flap, while it offers the advantage of sparing major perfusing vessels, there have been reports of distal necrosis when the flap extends beyond the metacarpophalangeal joints to cover areas as far as the proximal interphalangeal (PIP) joint.[8] For this reason we did not select it for our reconstruction.

Reconstruction of the foot- Free flaps such as the superficial circumflex iliac artery perforator (SCIP) flap or anterolateral thigh (ALT) flap are frequently used facing a wide forefoot tissue loss.[8] In our case, we used a radial artery flap harvested from the amputated hand site because the flap was thin, pliable and hairless, and sacrifice of the vascular axis would not pose significant issues.

In the literature, there are very few documented cases describing reconstructions following the removal of squamous cell carcinomas in patients with Huriez syndrome. Dumont et al.[3] described their experience with a patient with recurrent squamous cell carcinoma, successfully treated with extensive excision followed by coverage using a robust radial forearm flap. In such patients, it is advisable to opt for more complex reconstructive procedures that can provide lasting benefits, and the authors consider it reasonable to prioritize the use of microvascular flaps in carefully planned reconstructions. This approach helps avoid subjecting patients to multiple reinterventions and amputations, which could otherwise worsen their quality of life. Flap tissue is not only involved in the degenerative processes associated with Huriez syndrome, but also enhances the trophism in the surrounding regions. Distant flaps can have a true 'therapeutic' role in the disease,[10] enhancing function and aesthetics while avoiding or delaying recurrences and the onset of new cancerous lesions.

Conclusion

Huriez syndrome is a rare dermatological condition with evolving surgical treatment strategies. Due to the long survival rates of patients, maintaining control over skin malignancies is essential for their quality of life. Limited literature exists on this, but current experiences may help establish better standards for surgical and reconstructive management. Distant flaps have proven safe and effective for reconstruction, preventing local recurrences and protecting the flap area from characteristic lesions, while also offering unexpected benefits to the surrounding skin—an observation not yet documented in the literature.

Ethical approval

Not required.

Declaration of competing interest

None declared.

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References

1. Loh AYT, Ho CM, Muthiah S, et al. Huriez syndrome caused by a large deletion that abrogates the skin-specific isoform of SMARCAD1. *Br J Dermatol*. 2021;184(6):1205–1207. doi:10.1111/bjd.19.
2. Lee YA, Stevens HP, Delaporte E, Wahn U, Reis A. A gene for an autosomal dominant scleroatrophic syndrome predisposing to skin cancer (Huriez syndrome) maps to chromosome 4q23. *Am J Hum Genet*. 2000;66(1):326–330. doi:10.1086/302718.
3. Dumont L.A., Gahagnon T., Martinot-Duquennoy V., Mortier L., Guerreschi P. Cas clinique: carcinome épidermoïde, lambeau antébrachial radial et syndrome d'Huriez. Mise au point sur une pathologie rare [Case report: squamous cell carcinoma, radial forearm flap.
4. Italian National Institute of Health (ISS). (2024). AIOM Guidelines for squamous cell carcinoma. Retrieved from https://www.iss.it/documents/20126/8403839/LG_313_AIOM_ca_squamocellulare.pdf.
5. Levin LS. The reconstructive ladder. An orthoplastic approach. *Orthop Clin North Am*. 1993;24(3):393–409.
6. Fronek LF, Harrison TA, Gray TE, Blalock TW. Review of cutaneous repairs on the hand and fingers following dermatologic surgery. *J Clin Aesthet Dermatol*. 2023;16(7):45–53.
7. Chaput B, Meresse T, Bekara F, et al. Lower limb perforator flaps: current concept. *Ann Chir Plast Esthet*. 2020;65(5–6):496–516. doi:10.1016/j.anplas.2020.06.009.
8. Rehim SA, Chung KC. Local flaps of the hand. *Hand Clin*. 2014;30(2):137–151. doi:10.1016/j.hcl.2013.12.004.
9. Mohan AT, Sur YJ, Zhu L, et al. The concepts of propeller, perforator, keystone, and other local flaps and their role in the evolution of reconstruction. *Plast Reconstr Surg*. 2016;138(4):710e–729e.
10. Patrignani A, Pagnotta A, Amendola C, Bruno E, Torto FL, Ribuffo D. Hand reconstruction in Huriez syndrome: the "therapeutic" role of distant flaps. *Hand Surg Rehabil*. 2024:101781. doi:10.1016/j.hansur.2024.101781.