JPRAS Open 39 (2024) 257–261

Contents lists available at ScienceDirect

JPRAS Open

journal homepage: www.elsevier.com/locate/jpra

Case Report

Digital papillary adenocarcinoma: An important differential diagnosis for digital soft tissue masses

RJ Goodall*, MA Pickford

Department of Plastic Surgery, Queen Victoria Hospital NHS Foundation Trust, East Grinstead, Sussex, UK

ARTICLE INFO

Article history:
Received 26 November 2023
Accepted 7 January 2024
Available online 10 January 2024

Keywords:
Digital papillary adenocarcinoma
Case report
Digital soft tissue swelling
Homodigital island flap

ABSTRACT

Digital papillary adenocarcinoma (DPAC) is a rare, aggressive cancer with significant metastatic potential which arises from digital sweat glands. We present a case of a DPAC managed with surgical excision and reconstruction with a reversed homodigital island flap.

Level of evidence: V

Introduction

Most swellings arising from the skin or subcutaneous tissues in the digits are benign,[1] with ganglia (50–80 %), tendon sheath giant cell tumours (7–12 %) and epidermal inclusion cysts (5–9 %) representing the most common pathologies.[1-3] Nonetheless, clinicians should always be suspicious of the possibility of malignancy, with cutaneous squamous and basal cell carcinomas being the commonest malignancies affecting the digits.[4] Digital papillary adenocarcinoma (DPAC) is a rare malignant tumour arising from sweat glands, usually those of digits, with an annual incidence of 0.8 cases per 1,000,000.[5] DPAC histopathological diagnosis can be challenging, but accurate and timely diagnosis is

* Corresponding author.
E-mail address: r.goodall@imperial.ac.uk (R. Goodall).

https://doi.org/10.1016/j.jpra.2024.01.004

important given the high associated metastatic rates, reported to vary from 14 to 41 %.[5-9] Here, we present a case of a DPAC affecting the left index finger, managed with surgical excision and reconstruction with a reversed homodigital island flap.

Case

A 68-year-old, right-hand-dominant, Caucasian male presented to our service with a 6-month history of a 2 × 2 cm painless, smooth, firm, fixed nodule over the dorsum of his left index finger distal interphalangeal joint (Figure 1). The lesion was referred by the local dermatology service as an incision biopsy-proven benign adnexal tumour, favouring a spiradenoma. However, following this initial biopsy, a second incision biopsy was performed by the referring team, which was this time reported as a DPAC. Due to diagnostic uncertainty, an urgent narrow-margin excision was performed to allow accurate and reliable histological analysis. The lesion was therefore excised with 3 mm peripheral margins to the paratenon of the underlying extensor tendon and was closed with a full-thickness skin graft under local anaesthetic. The histological analysis of this excision specimen again initially diagnosed a benign spiradenoma, however a second opinion was sought, which revised the diagnosis to DPAC with involved peripheral and deep margins.

Re-excision was therefore undertaken with 5 mm peripheral margins, down to the periosteum of the distal phalanx, inclusive of the entire nail complex, but preserving the extensor tendon. This wider excision obtained histological clearance; the defect was reconstructed using an ulnar-based, reversed homodigital island flap. The donor site was closed using a full-thickness skin graft harvested from the ipsilateral volar forearm. Both digital nerves were preserved.

Post-operatively, both the flap and graft healed promptly without complications. At 14-months follow-up the patient has maintained full, pain-free range of motion (Figure 2). There has been no evidence of local or regional recurrence identified during each of his 4-monthly surveillance clinic appointments, with no palpable axillary lymphadenopathy and no radiological signs of pulmonary metastasis on annual surveillance chest radiographs. He will be monitored for 5 years before being discharged from the service.

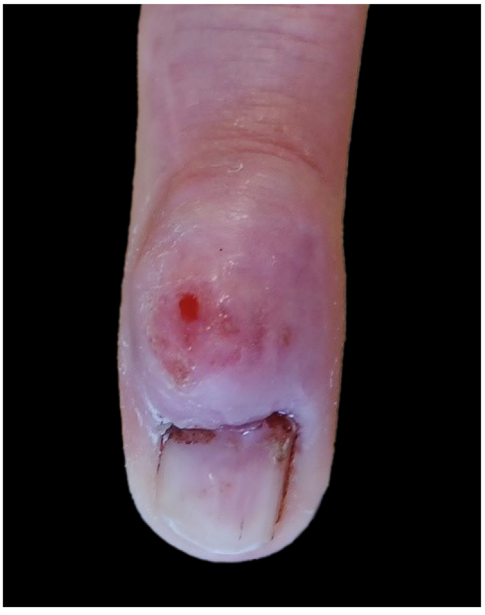

Figure 1. Pre-operative clinical photograph of the left index finger digital papillary adenocarcinoma.

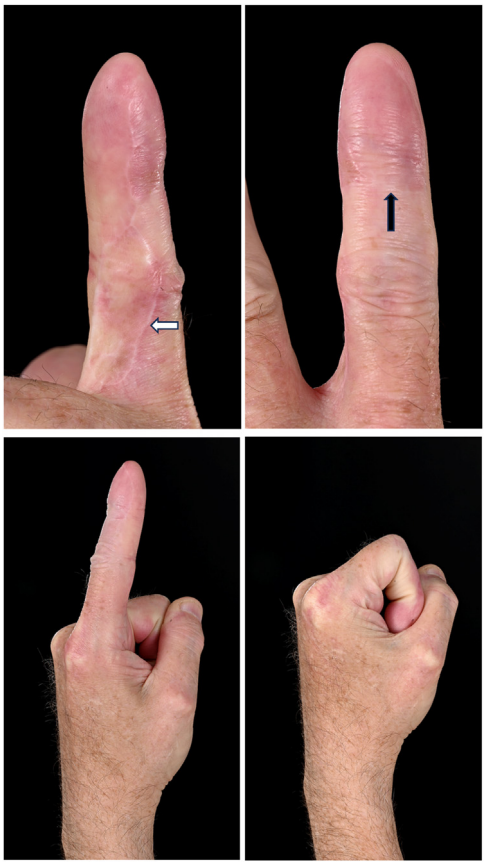

Figure 2. Post-operative result following wide excision of digital papillary adenocarcinoma and reversed homodigital island flap reconstruction. *Black arrow = healed flap, white arrow = skin grafted donor site.*

Discussion

Digital papillary adenocarcinoma is a very rare cancer arising from sweat glands, which typically affects the distal ends of both upper and lower limb digits.[5] DPACs have been reported in individuals as young as 19,[7] however the majority affect those between 50 and 70 years with a predominance for males (male to female ratio between from 4 to 9:1).[5]

Helwig first described DPAC in 1979 but did not formally publish data on these tumours until 1987, when he and Kao described DPACs as "aggressive digital papillary tumours" because of their locally aggressive behaviour and tendency to occur on digits.[6] Kao and Helwig's histological analysis of 57 cases in 1987 subdivided DPACs into lower grade "digital papillary adenomas" and higher grade "aggressive digital papillary adenocarcinomas" based on histological parameters such as glandular differentiation and mitotic rate.[6] However, in 2000, Duke's subsequent analysis of 67 cases (of whom 14 % developed metastases with 3 individuals dying from metastatic dissemination of the disease) none of these histologic features had any prognostic significance.[7] These observations prompted changes in nomenclature to encourage all digital papillary sweat gland tumours to be considered and managed as aggressive digital papillary adenocarcinomas (DPACs), and the term digital papillary adenoma was abandoned.

In early reports of DPAC, the histology was described as "distinctive, with a typically solid and cystic growth pattern with papillae formation of various types and a pattern of fused back-to-back

gland-like structures in solid areas".[7] Despite this, the rarity of DPAC can lead to diagnostic uncertainty, as was observed in the present case wherein the initial incision biopsy, and then the narrow margin excision biopsy, were both reported as a benign spiradenoma by two separate histology departments, leading to confusion and a delay in treatment. Misdiagnosis (or delays to accurate diagnosis) can prove disastrous for patients with DPAC, given its locally aggressive characteristics and high metastatic potential, with metastatic rates reported to be as high as 41 %.[6]

Metastases may occur through lymphatic and/or haematogenous spread, with regional lymph node and pulmonary metastases being the most common sites for spread.[5-10] DPACs capacity for lymphatic spread has prompted investigation into the role of sentinel lymph node biopsy (SLNB). A single-centre retrospective case series of 18 patients treated for DPAC with wide excision and SLNB identified positive nodal metastases in 17 % of patients (none of whom had clinically palpable lymphadenopathy), who subsequently underwent completion lymph node dissection.[10] At a median follow-up of 53 months, no individuals with negative SLNB had evidence of recurrence or metastases. Further data demonstrating the diagnostic test accuracy of SLNB in DPAC is lacking due to a sparsity of clinical cases, therefore robust, evidence-based recommendations for practice are similarly absent. Furthermore, recommendations for excision margins (including the possibility of digital amputation), radiological staging investigations and duration/frequency of follow-up surveillance are also lacking. The patient presented in this case report was not offered SLNB, and instead will undergo regular clinical surveillance in which examination for local recurrence and regional lymphadenopathy will be supplemented with annual chest radiographs, in view of the possibility of pulmonary metastases described in the literature.[10]

The distal location of DPACs, on both the volar and dorsal surfaces, makes the excision defects well suited to reconstruction using reversed homodigital island flaps. The latter provide single-stage reconstruction with excellent return of mobility.

Conclusion

DPAC is a rare but aggressive cancer with significant metastatic potential. Failure to diagnose timely and accurately can prove disastrous. Consensus on the role of SLNB, recommended surgical excision margins and follow-up strategies would help clinicians managing this condition in the future.

Funding

None.

Guidelines

This report was written in line with STROBE guidelines.

Consent

Written patient consent was obtained for use of their images in scientific publication.

Ethical approval

Not required.

Declaration of competing interest

None.

References

1. Hsu CS, Hentz VR, Yao J. Tumours of the hand. *Lancet Oncol*. 2007;8(2):157–166.
2. Nahra ME, Bucchieri JS. Ganglion cysts and other tumor related conditions of the hand and wrist. *Hand Clin*. 2004;20:249–260.
3. Glowacki KA, Weiss AP. Giant cell tumors of tendon sheath. *Hand Clin*. 1995;11:245–253.
4. Ilyas EN, Leinberry CF, Ilyas AM. Skin cancers of the hand and upper extremity. *J Hand Surg Am*. 2012;37(1):171–178.
5. Rismiller K, Knackstedt TJ. Aggressive digital papillary adenocarcinoma: Population-based analysis of incidence, demographics, treatment, and outcomes. *Dermatol Surg*. 2018;44(7):911–917.
6. Kao GF, Helwig EB, Graham JH. Aggressive digital papillary adenoma and adenocarcinoma. A clinicopathological study of 57 patients, with histochemical, immunopathological, and ultrastructural observations. *J Cutan Pathol*. 1987;14(3):129–146.
7. Duke WH, Sherrod TT, Lupton GP. Aggressive digital papillary adenocarcinoma (aggressive digital papillary adenoma and adenocarcinoma revisited). *Am J Surg Pathol*. 2000;24(6):775–784.
8. Weingertner N, Gressel A, Battistella M, Cribier B. Aggressive digital papillary adenocarcinoma: A clinicopathological study of 19 cases. *J Am Acad Dermatol*. 2017;77(3) 549-558.e1.
9. Vanderbilt C, Brenn T, Moy AP, et al. Association of HPV42 with digital papillary adenocarcinoma and the use of in situ hybridization for its distinction from acral hidradenoma and diagnosis at non-acral sites. *Mod Pathol*. 2022;35(10):1405–1410.
10. Bartelstein MK, Schwarzkopf E, Busam KJ, Brady MS, Athanasian EA. Sentinel lymph node biopsy predicts systemic recurrence in digital papillary adenocarcinoma. *J Surg Oncol*. 2020;122(7):1323–1327.