Case Report

Digital Papillary Adenocarcinoma, a Rare Malignant Tumor

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Keywords

Digital papillary adenocarcinoma · Hand tumor · Digital mass · Hand · Oncology

Abstract

We present a case report of a digital papillary adenocarcinoma (DPA) on the left thumb with slow growth that was completely excised, without recurrence or metastasis during 24-month follow-up. DPA is a rare malignant tumor often located on the digits of the hand. Due to its slow growth and a non-specific appearance, the diagnosis is often missed or delayed. Treatment should be surgical with clear resection of the margins due to high risk of recurrence and/or metastasis.

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Introduction

Digital papillary adenocarcinoma (DPA) is a rare condition, with a small number of case series reported in the literature. It is an aggressive tumor of the eccrine sweat gland cells, often located on the distal extremities, particularly in the fingers/toes and adjacent palmar/plantar skin. DPA frequently occurs on the volar surface of the digit, near the distal interphalangeal joint or in the periungual region [1].

Clinically, DPA presents as a solitary, usually painful mass with slow growth. Despite being more common in adults with a predominance of male over female patients, it has also been observed in children. The local recurrence rate is of 30% [1], while the prevalence of metastasis has been reported as about 14% [2], particularly to lungs and lymph nodes.

The recommended treatment is surgical excision of the primary tumor, with the possibility of proximal amputation, but no objective margin is suggested by the literature, and no evidence exists to support the use of chemotherapy or radiotherapy [2].

We report a case of DPA, including its clinical course, pathological findings, and therapeutic approach, with a 24-month follow-up period.

Case Presentation

A 53-year-old male had a small, elastic, painless 2 cm nodule, on the volar side of the left thumb distal phalanx, without prior trauma or infection, which had increased in size over the course of the previous year. There was neither redness, discoloration, inflammation of the skin, nor alteration of perfusion. The hand's function was not compromised, presenting the thumb's full range of motion, without any alteration of sensation.

Intraoperatively, we observed a $2.5 \times 2 \times 1$ cm cystic brown-grey tumor with an elastic consistency and well-defined limits, without any adhesions or pedicle. There was no involvement of muscle, tendon, or bone (Fig. 1). Complete surgical resection of the mass was performed, without rupturing the cyst. Postoperative wound healing had no complications and thumb's function was not compromised.

A histopathological study identified a low mitotic grade lesion, mainly cystic, with glandular epithelium protruding as papillae and nuclear pleomorphism with hyperchromatism (Fig. 2). Immunohistochemistry showed focal positivity for EMA (epithelial membrane antigen), CEA (carcinoembryonic antigen), and diffusely for P63 (Fig. 2). These findings were compatible with the DPA diagnosis, without showing any signs of invasion of adjacent structures.

On the thoracoabdominopelvic computed tomography, there was no evidence of organ metastasis or lymphadenopathies. At the 3rd month of follow-up period, magnetic resonance imaging (MRI) of the hand showed neither signs of local recurrence, nor remaining tumor tissue (Fig. 3).

Once the high rates of recurrence and metastasis were explained to the patient, he started a clinical and radiological follow-up with MRI of the hand semi-annually. During the postoperative 24-month follow-up period, the patient presented no signs of local recurrence or distant disease.

Discussion/Conclusion

DPA was first described by Helwig et al. [3] as an "eccrine acrospiroma," and later differentiated between "aggressive digital papillary adenoma" and "aggressive digital papillary adenocarcinoma," taking into account its histological features [4]. Duke et al. [2] suggested a reclassification of both as "aggressive digital papillary adenocarcinoma," and because none of the histological parameters have been predictive of recurrence/metastasis, Suchak et al. [5] removed "aggressive," due to its low-malignancy grade.

The etiology of DPA is largely unknown; a BRAF-V600E mutation has been reported in one case [6], but the significance of this finding is unclear, with new reports of genetic tumor overexpression of FGFR2 [7].

The age at onset is usually between the fifth and seventh decades (the average is 52 years old) [2], affecting males more often than females (7–9:1) [1, 8]. The tumor commonly presents as a solitary mass, may be located between the nail bed and distal interphalangeal joint [8], but it has also been reported on the palmar surface of the hand, plantar surface of the foot, lower leg, and on the web spaces of hands and feet, with rare cases reported on the lips and ears [9]. Often located on the dermis, DPA can invade deeper structures such as muscles, tendons or bones.

Differential diagnosis should include ganglion cyst, foreign body or pyogenic granuloma, paronychia, glomus tumor, squamous cell carcinoma, hemangioma, giant cell tumor, osteomyelitis, and soft tissue infections [1].

Despite being a malignant tumor, it does not always have an aggressive nature, and therefore, a wide surgical excision or an ab initio amputation is not always recommended. Complete excision is the standard treatment for all DPAs, but it has been reported that up to 34% may have the need of subsequent reexcision or amputation [2].

Local recurrence can occur in 30–50% of the cases after surgical excision, which can be reduced to 5% after excision with a clear resection of the margins [2]. The overall rate of reported metastatic disease is between 14 and 26% [2, 5, 8], being more prevalent in the lungs, but other areas have been observed, such as lymph nodes, brain, skin, bones, and kidneys [8, 9].

None of the known clinical or histological features are predictive of local recurrence or metastasis, and there is no recommendation for routine sentinel lymph node biopsy [6]. No effective treatment for widespread metastatic disease has yet been developed, and there is no evidence to support the use of chemotherapy or radiotherapy [2].

The follow-up period should be long-term, given that its growth is insidious with a high recurrence rate, and metastatic disease has been reported as late as 20 years following its excision [2]. The diagnosis of local recurrence through MRI in early stages may be difficult because the appearance is similar to a reactive/inflammatory process following previous surgery. It is still unclear which features can help predict both the outcome and recurrence rate [5].

This case highlights that a rare malignant tumor, presenting with a swelling on the digit, can mimic benign conditions, such as ganglion, cyst, or local infection. Physicians should be aware of the existence of this condition, proper management, and treatment, as well as the long follow-up that is required, whilst it is still unclear which features can help predict DPA behavior. DPA should be treated surgically with a clear resection of the margins, always maintaining a strict follow-up for the detection of recurrence and/or metastatic disease.

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Statement of Ethics

The patient provided informed consent for the publication of this case.

Disclosure Statement

The authors have no conflicts of interest to declare.

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Author Contributions

C. Vale analyzed and interpreted the patient data regarding the disease and was a major contributor in writing the manuscript. A. Carvalho collected the data. T. Roseiro co-wrote the manuscript. J. Antunes reviewed the manuscript and was a supervisor. O. Tellechea performed the histological examination and description. V. Oliveira was the project administrator and supervisor. All authors read and approved the final manuscript.

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Fig. 1. a, b Intraoperative view of cystic tumor. c Macroscopic aspect of the tumor.

Fig. 2. Histopathology (hematoxylin & eosin stain). a Cystic and papillary tumor. b Proeminent cellular atypia and nuclear pleomorphism. c Immunochemistry: diffusely nuclear immunoreactivity with P63.

Fig. 3. MRI of the hand without signs of local recurrence.