Digital papillary adenocarcinoma: A case report

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Abstract
We present the case of a 64-year-old male with a 3-month history of a mass involving the distal portion of the fifth finger on the left hand which was histologically confirmed as digital papillary adenocarcinoma. Although the tumor is low grade, the chances of recurrence and metastasis are high.

KEYWORDS
digital, digital papillary adenocarcinoma

INTRODUCTION

Digital papillary adenocarcinoma (DPA) is a rare and aggressive neoplasm of the sweat glands. It was first described by Helwig in 1979 as a tumor with high rate of recurrence and metastasis. The term aggressive digital papillary adenocarcinoma (ADPA) was later introduced by Duke et al. It is often misdiagnosed and characterized by digital location particularly in the fingers/toes and adjacent palmar/plantar skin. Most tumors present as a slow-growing solitary, nodular, and usually painful mass that is grossly cystic. We report the case of a 64-year-old male with a DPA involving the fifth digit of the left hand.

CASE PRESENTATION

A 64-year-old man presented with an 18-month history of a scar-like lesion on the pulp of the fifth digit of the left hand. The lesion was rapidly increasing in size in the last three months, and it became associated with occasional pain along with darkening. Moreover, he reported no weight loss, anorexia, or fatigability on exertion. On physical examination, he was healthy looking with a mass at the volar surface of the distal phalanx of the fifth digit on the left hand. The mass was mobile, nontender, darkish, roundish, and firm in consistency. The mass measured 1 cm in its greatest diameter. The preliminary investigations included body temperature 36.8°C, blood
pressure 162/98 mmHg (mildly hypertensive), pulse rate 78 beats/minute, random blood sugar 9 gm/dl, hemoglobin level 10 gm/dl (mild anemia), white blood cell count, and its differentials (all were normal), body mass index 28.6 kg/m², and creatinine 24.1 mmol/L. Incisional biopsy was done which revealed infiltrating and proliferating hyperchromatic glandular structures with micropapillary structures delineated within the dermis. Additionally, the overlying skin was not ulcerated. The histomorphological features were highly suggestive of a metastatic papillary adenocarcinoma probably originating from the breast, colon, or thyroid gland.

Therefore, this necessitated for meticulous search of the primary site of the tumor. Laboratory workups include thyroid function test, carcinoembryonic antigen (CEA), carbohydrate antigen (CA)-19, and carbohydrate antigen (CA)-125 levels which were normal. Esophagogastrroduodenoscopy (OGD), colonoscopy, and ultrasound scan around the neck area were also normal. Additionally, thoracoabdominal computed tomography (CT) scan was normal.

Wide local excision was done, and the mass was taken for histological evaluation. The microscopic findings revealed a tumor which was composed of well-formed atypical glands with papillary infoldings lined by malignant cells with moderated atypia and some mitoses (about 2 mitoses per HPF), and the tumor cells were confined within the dermis, Figure 1(A), (B), and (C). Neither lymphovascular nor perineural invasion was evident. Immunohistochemistry testing was done to confirm the diagnosis which included pan cytokeratin (positive), EMA (positive), CEA (patchy positively), C-Kit (negative), and S100 (negative).

The patient was discharged home on 50 mg tabs of diclofenac and 500 mg tabs of metronidazole both for five days. He was being clinically evaluated after two months postoperatively. After a follow-up period of almost two years, his medical condition remained uneventful.

3 | DISCUSSION

DPA has a male predominance and is commonly diagnosed between 50 and 70 years of age similar to the patient reported in the present case albeit of a small number of cases that have been reported in the pediatric population. Initial studies by Kao et al suggested that it is more in Caucasians however several studies have reported cases in the non-Caucasian population.

The tumor mainly grows while confined in the dermis, poorly delineated and it consists of well-formed atypical glands with papillary projections which are lined by malignant cells with moderated atypia and few mitoses. Other studies have reported that DPA is a well-circumscribed, infiltrative growth pattern, tubuloalveolar and ductal structures with areas of papillary projection protruding into the cystic lumina. The ductal structures are usually larger and more dilated compared to those of papillary eccrine adenoma. Morphologically, the tumor has solid and cystic components with closely aggregated back-to-back glandular architectures lined by cuboidal to columnar epithelia that are surrounded by a basal myoepithelial layer. Cytologic atypia, mitotic counts, and necrosis have also been observed, however,
not so frequent. The parenchymal component shows variation in appearance and commonly has either thin fibrous septa or areas of dense hyalinized collagen with cysts that contain either necrotic debris or eosinophilic secretory material.²

Misdiagnosis and late diagnosis of DPA are a common phenomenon because of its rarity. It is often mistaken clinically for benign pathologies such as calluses, ganglion cysts, gout, and soft tissue infections due to its slow growth pattern.⁸ Additionally, microscopically the tumor may be misdiagnosed for adenocarcinomas arising from the breast, lung, and thyroid as was the case in our patient.⁹

Considering the confirmation of the diagnosis of DPA, immunohistochemical studies have shown that DPA is positive for S100 protein, and some express estrogen and/or progesterone receptor.³–⁵ In a study which was done by Suchak et al, it was found that Ki67 proliferation index was ranging from 2% to 30%, whereas smooth muscle actin (SMA), calponin, and P63 stained the myoepithelial layer surrounding the glandular structure.¹⁰ However, the presence of myoepithelial cells does not reflect the benign biological behavior of the tumor.

The recommended treatment for patients with DPA is surgical excision of the primary tumor with the possibility of proximal amputation; however, neither objective margin nor evidence exists to support the use of chemotherapy or radiotherapy.³⁷ Patients treated with either wide local excision (WLE) or amputation have 5% rate of recurrence compared to 50% for those who are not treated all.⁶ The local recurrence rate varies according to the management plan. Recurrence and metastasis rates of 47% and 41.7%, respectively, have been reported with reduction rates of 16% and 19% for recurrence and metastasis, respectively, following surgical treatment with either WLE or digital amputation.²¹⁰ Metastasis of the tumor to the lungs and lymph nodes has been reported to be approximately 14%.³ Even after developing metastasis, the 5-year overall survival was once reported to be 20 years.⁸ Therefore, this indicates that this is a low-grade tumor, and if it is diagnosed at its early stage and treated appropriately, patients would have very good prognosis.

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None.

CONFLICT OF INTEREST
The authors have no conflicts of interest to declare.

AUTHOR CONTRIBUTIONS
BAK: curation of the information regarding the patient and wrote the first draft, VA: curation of the information regarding the patient and wrote the first draft, JJY: organized the manuscript, performed in-depth literature review, and wrote the first draft. All authors revised the final version, and they agreed for its intellectual content.

ETHICAL APPROVAL
We confirm that publication of the case details required institutional approval and we also confirm that we obtained ethical clearance from the institution review board (IRB) of the School of Biomedical science, Makerere College of Health Sciences (MakCHS).

CONSENT
We confirm that a written informed consent was provided by the parent to have the case details and any accompanying images get published.

DATA AVAILABILITY STATEMENT
Data regarding the patient reported in this paper are available upon request from the authors.

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