Case Report

Aggressive Digital Papillary Adenocarcinoma–A rare Entity Posing a Diagnostic Challenge

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Abstract

Aggressive digital papillary adenocarcinoma (ADPCA) is rare tumor of the sweat glands, which is characterised by lesions on the fingers, toes and the digits. The lesion is serious but often overlooked because it is confused clinically with benign and non-tumorous entities. In this paper, we present an interesting case of ADPCA in a 40-year-old lady, suspected clinically as pyogenic granuloma, with the initial excision biopsy indicating a malignant tumor in the sweat gland. Ray amputation of the affected finger was done followed by a thorough work up to rule out metastasis. Histopathological features were studied in detail, along with required markers. We present this case as the documentation of this malignant tumor is limited in literature and also it requires a high index of suspicion when dealing with all masses arising on the digits.

Keywords: *amputation, digital carcinoma, histopathology, recurrence, metastasis*

Introduction

Aggressive digital papillary adenocarcinoma (ADPCA) is distinct from other sweat gland tumors, not only because of its rarity but also its potential for recurrence and distant metastasis. As this neoplasm can be mistaken for benign lesions, both clinically and histologically, substantial delays can occur prior to the start of a definitive therapy (1). Unlike other cancers, the histological and immunohistochemical features of this tumor are not predictive of outcome. The aggressive behavior of this entity is often overlooked.

Case Report

A female, aged 40 years had noticed a skin-colored, solitary, dome-shaped nodule measuring 5 × 5 mm on the tip of the right index finger with mild pain since a year. Suspecting it to be a pyogenic grauloma because of the above-mentioned features, the swelling was excised and sent for histopathological examination. A provisional diagnosis of a malignant sweat gland neoplasm, with surgical margin being positive for tumor deposits was made. The patient got discharged against medical advice and was lost to follow-up.

After 6 months, the patient presented again, with recurrence of the same swelling, which was solitary, dome shaped, with irregular borders, and intact overlying skin, measuring 1.2 × 1 cm. X-ray of the hand showed a soft tissue growth, with mild cortical thickening. Magnetic resonance imaging (MRI) showed a soft tissue growth with tendon involvement. Recurrence, fast growth, previous histopathological examination report and considering the chances of no follow-up, the distal phalanx was amputated after the patient's consent. Gross specimen of the amputated finger after surgery showed a grey white tumor at distal end (Figure 1). Histopathological examination of the specimen showed a malignant neoplasm in the dermis (Figure 2a). The tumor cells were arranged in varied pattern, including large cystic spaces (Figure 2b), with few showing papillae, solid sheets, small cords and nests of cells infiltrating the dermis and adjacent tissue. The cells exhibited moderate pleomorphism, had eosinophilic cytoplasm and bland vesicular nuclei. Brisk mitotic activity was noted (3–5/hpf). Areas of tumor necrosis and desmoplasia were seen. The distal stump of amputation was negative for tumor deposits. Though immunohistochemistry confirmed the epithelial origin by being positive for epithelial membrane antigen (EMA) and cytokeratin (CK) myoepithelial markers were also positive (calponin and p63) (Figure 3: Immunohistochemistry (IHC) showing p63 positivity in the myoepithelail cells lining the

tubules). The final diagnosis was given as aggressive digital papillary adenocarcinoma based on the clinicopathological and histological findings.

The patient was subjected to a full body work up, including computed tomography (CT) to rule out lung and other metastatic focus from the tumor in the digit, and primary tumor in any other site and was discharged on the 5th post-operative day and was lost to follow up. Repeated attempts to contact the patient for further follow up yielded no responses till date.

Discussion

ADPCA tumors are rare sweat gland neoplasms, first described by Helwig in 1984 and later by Kao GF in 1987. They characteristically occur on the hands, fingers and toes. These tumors were so named because of their potential for aggressive local growth, resulting in high recurrence rate and prelidiction for digits (2).

This lesion is found more commonly on the hands and occurs predominantly in males (3), but the patient in our case was a female. These swellings often masquerade ganglion cysts (4) or pyogenic granuloma as in our case (1). They can also be mistaken for paronychia or present with an appearance similar to osteomyelitis and soft tissue infection, haemangioma, giant cell tumor of the tendon sheath and squamous cell carcinoma, thereby showing signs of a wide variety of lesions (5).

Histologically ADPCA has solid and cystic spaces, with or without papillary projections in the cystic spaces. The cysts are formed either by central degeneration/necrosis of solid areas or amassed secretory material, which appeared to be mucin (6). The close differential diagnosis indicated papillary eccrine carcinoma, which in our case was ruled out as the ducts and cysts of this lesion were much smaller than that of ADPCA.

Ruling out a primary in the breast is very important in females as the histopathological features of malignancy such as hyperchromatic nuclei, brisk mitotic activity; areas of tumor infiltration into the surrounding stroma and necrosis, seen in ADPCa closely resemble a metastatic infiltrating carcinoma of the breast (7). Some authors have advised amputation as definitive surgery rather than local excision because of the propensity of this neoplasm to recur and metastasize to distant sites, particularly lungs (1). Wide excision and partial digital amputation significantly reduces the chances of metastasis and recurrence (8). Sentinel lymph node mapping

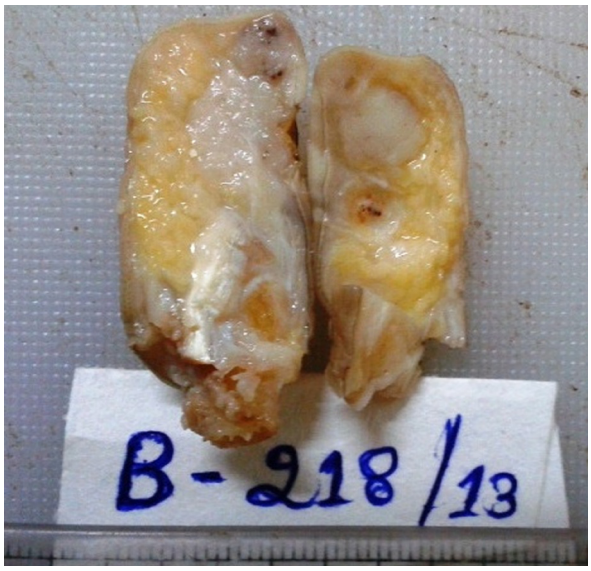

Figure 1: Cut section of the amputated digit showing a grey white tumor at the distal end.

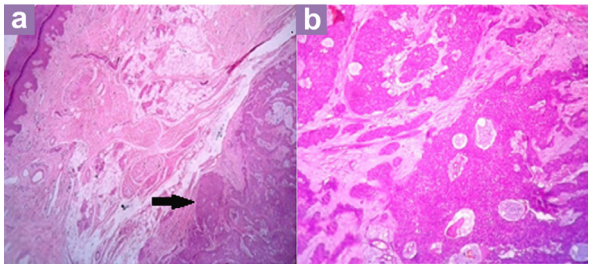

Figure 2: a) Hematoxylin and eosin (H&E) stain showing the arrow pointing at the tumor in the dermis. b) H&E stain showing cystic spaces and cells in solid sheets.

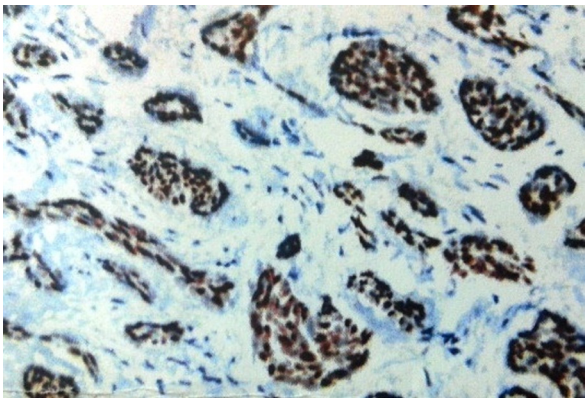

Figure 3: Immunohistochemistry (IHC) slide showing p63 positivity in the myoepithelial cells lining the tubules.

is useful in detecting early metastatic spread, and is recommended when possible (9).

Immunohistochemical work up of 31 cases of ADPACa done by Suchak et al showed the tumor cells are positive for EMA, CEA and CK and also surprisingly, myoepithelial markers (desmin, calponin and p63) were also found to be positive. Their study concluded the presence of myoepithelial cells histologically and immunohistochemically was not synonymous with benignity as many of them developed lung metastasis (8). In our case also we found positive for both epithelial and myoepithelial markers, and because of its recurrence and propensity for metastasis, the diagnosis of carcinoma was made. No effective treatment for metastatic disease has yet been developed.

Conclusion

ADPCA is an uncommon malignant adnexal tumor of the skin, rare in females, with prelidiction for the digits, masquerading other benign neoplastic, and non-neoplastic lesions. Hence a high index of suspicion is needed for early diagnosis and treatment. The tumor cells may express myoepithelial markers in addition to epithelial markers. However, such expression does not indicate their benign nature. Wide excision or amputation if clinically indicated, and long-term follow-up for recurrence and metastasis must be considered for these tumors.

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Conflict of interest

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Authors' Contributions

Conception and design: GV
Analysis and interpretation of the data, drafting of the article, collection and assembly of data: AS
Critical revision of the article for the important intellectual content and final approval of the article: GV, VC

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