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Digital Papillary Adenocarcinoma of Phalanx: A rare cutaneous malignancy

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Abstract: Digital papillary adenocarcinoma is a rare sweat gland neoplasm of eccrine differentiation affecting the distal aspect of digits. It is a slow growing tumour and may invade the underlying soft tissues and bone. The tumour is of aggressive nature with a propensity to metastasize and recur. We report a rare case of digital papillary adenocarcinoma of middle phalanx in a 45 years old male with a short history of six months duration. Histopathology and immunohistochemistry confirmed the diagnosis. Partial amputation of the digit was carried out and no signs of recurrence and metastasis were noted after two years of follow up. Thus, digital papillary adenocarcinoma requires a high index of suspicion in conditions where the lesion is arising on the distal aspects of the digits because early detection and treatment can prevent its further progression.
Keywords: Digital Papillary Adenocarcinoma, Phalanx, Eccrine gland, Neoplasm, immunohistochemistry

INTRODUCTION:

Digital papillary adenocarcinoma is an uncommon cutaneous adnexal tumour of eccrine sweat gland origin. It presents as a painless, solitary, cystic nodule that usually affects the digits of both upper and lower extremities. The tumour is highly aggressive as it can locally invade the underlying tissues and can metastasize. Clinically, it simulates other lesions of the skin thereby, requiring histopathological examination for its precise identification. Elementary detection and prompt management is the key to reduce the risk of metastasis.

CASE REPORT:

A 45 years old healthy male presented in the department of skin with a solitary nodule on the distal aspect of his right middle phalanx for the past 6 months. On local examination, the nodule was painful, erythematous, tensed, and small measuring approximately 1.5 X 1.5 cms. X-Ray revealed a soft tissue growth with no involvement of bone. Routine haematological and biochemical investigations were within normal limits. Chest x-ray was normal. The nodule was excised and sent for histopathological examination with a clinical impression of capillary hemangioma of the middle phalanx.

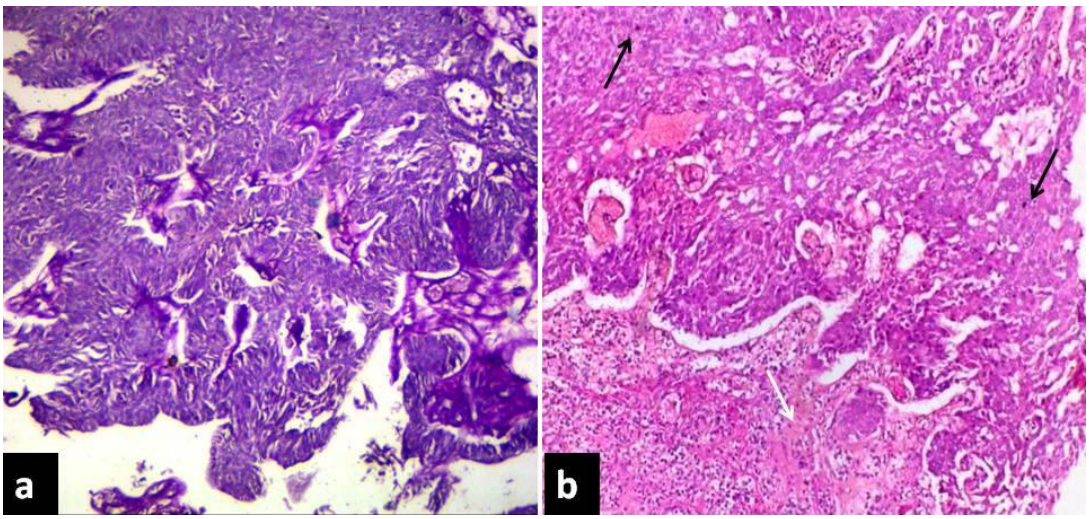

Fig-1: Digital Papillary Carcinoma (A) showing papillary epithelial projections (PAS, 100X) (B) Showing epithelial projections within the cystic spaces and abundant necrosis (marked with white arrow) and mitosis (marked with black arrows) (H&E, 100X)

Grossly, the nodule was well defined and greyish-white in colour. Microscopically, both solid and cystic areas were identified with papillary epithelial projections within the cystic spaces associated with fibrovascular cores in some areas. Multi-nodular epithelial aggregates were present. The epithelium was made up of low columnar cells surrounded by a basal layer of myoepithelium and the cysts were filled with eosinophilic decapitation secretions. Mitoses and necrosis were abundant [Figure 1]. The luminal cells revealed strong positivity for CEA while the basal cells expressed CK5/6.

On the basis of above findings and clinic-radiological correlation, a histopathological diagnosis of digital papillary adenocarcinoma was made. Partial amputation of the concerned digit was performed. The patient was kept under close follow up for two years with no signs of recurrence observed during this period.

DISCUSSION

Digital papillary carcinoma (DPA) is regarded as an uncommon malignant adnexal neoplasm of sweat gland origin with eccrine differentiation. It was first described by Helwig [1] in 1984 and later by Kao et al [2] in 1987. It presents almost exclusively on the fingers, toes, palms, and soles. Hands are involved more frequently than feet [3]. There is a male predilection, and most affected individuals are adults in the fifth and sixth decades of life **[**2]**.** It has also been reported in a 15 years young female [4, 5].

In the past, it was classified as aggressive digital papillary adenoma and aggressive digital papillary adenocarcinoma on the basis of histological features. The criteria for malignancy were poor glandular differentiation, necrosis, cytological atypia, high mitotic count and invasion of underlying structures and blood vessels [2]. However, WHO (2006) recommends to avoid the term aggressive digital papillary adenoma as these tumours tend to grow aggressively resulting in local recurrence and metastasis. The commonest site of metastasis is lung and lymph nodes [2]. Therefore, it is essential to perform a routine chest x-ray examination once the diagnosis of digital papillary adenocarcinoma is established. In the present case, there were no signs of metastasis in the lungs and lymph nodes.

Histologically, the tumour is composed of multi-nodular epithelial aggregates with cystic spaces in the dermis. Papillary epithelial projections are common within cystic spaces associated with fibrovascular cores in some areas. The epithelium is composed of low columnar or cuboidal cells. Cytologic atypia is usually not marked. Mitoses and necrosis are frequent findings. Cysts contain either necrotic debris or eosinophilic secretory material. Immunohistochemically, the tumour cells express diffuse immunoreactivity for EMA and CEA [6]. The myoepithelial layer shows positivity for calponin, S100, basal cytokeratins and smooth muscle actin. The differential diagnosis comprises of apocrine adenocarcinoma, adenoid cystic carcinoma of the sweat glands, and mucinous eccrine carcinoma.

Digital papillary adenocarcinoma is often clinically mistaken as other common primary lesions of skin like osteomyelitis, soft tissue infections, hemangiomas, giant cell tumour of tendon sheath, ganglion cysts, and sometimes squamous cell carcinoma [7]. Histopathology and immunohistochemistry helps in resolving the grey zone and confirms the diagnosis. Wide local excision with partial digital amputation has been offered as the treatment of choice as it decreases the risk of recurrence and metastasis [6].

In the current case, patient presented to us early because of intense pain and erythema associated with digital mass. As DPA is very rare in general practise, it was clinically misdiagnosed as capillary hemangioma. Therefore, histopathology is essential in the lesions of digits and immunohistochemistry supports the diagnosis. Prompt surgical management was offered and our patient showed no recurrence or metastasis after two years of follow up.

CONCLUSION:

We present this case as it is an extremely rare tumour with limited cases described in the literature. Thus, digital papillary adenocarcinoma requires a high index of suspicion in conditions where the lesion is arising on the distal aspects of the digits because early detection and treatment can prevent its further progression.

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