Aggressive Digital Papillary Adenocarcinoma (ADPCA) of heel with Distant Lung Metastasis: A Rare Presentation

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Abstract
Aggressive digital papillary adenocarcinoma (ADPCA) is rare tumour of the sweat glands, 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. Here, we present an interesting case of ADPCA in a 55-year-old male, suspected clinically as pyogenic granuloma, with the initial excision biopsy indicating a malignant tumour in the sweat gland. The histopathological features were studied in detail, along with required markers. The documentation of such malignant tumour is limited in literature. It requires a high index of suspicion and a prudent approach while dealing with all masses arising in the extremities.

Keywords: Digital carcinoma, histopathology, recurrence, metastasis.

Introduction
The aggressive digital papillary carcinoma (ADPC) is a rare category of sweat gland neoplasm characteristically seen on the distal extremities. These tumours are termed so because of their potential for aggressive local growth, resulting in high recurrence rate and predilection for digits. These lesions have predilection for hands and occurs predominantly in males1. However, granulomatous growth on left heel of foot is very rare presentation. These swellings often mimic ganglion cysts or pyogenic granuloma, osteomyelitis, soft tissue infections, haemangiomas, giant cell tumour of the tendon sheath and squamous cell carcinoma, thereby showing signs of a wide variety of lesions2.

Case Report
A 55-year-old male presented with a painless ulcerative lesion of size 4x4 cm at heel of his right foot (Fig 1). The mass was noticed three years back and on gross examination, the nodule
appeared well demarcated and was initially suspected to represent a ganglion cyst. Six months back patient had history of trauma in the same feet and non-healing ulceration developed at the site. The biopsy was planned and that confirmed the diagnosis of ADPAC although, carcinoembryogenic antigen (CEA) was normal. The immunohistochemical stains were performed on paraffin-embedded sections using antibodies against s100 protein, CEA, cytokeratin Cam 5.2, cytokeratin AE1:AE3, and epithelial membrane antigen (EMA). Both epithelioid and spindled cells were diffusely positive for s100 protein, and showed focal positivity for EMA. The immunostains against cytokeratin (Cam 5.2 and AE1:AE3) were immunoreactive, predominantly in the epithelioid cells. Stains against CEA were reactive in the glandular lumens and at the luminal aspect of the epithelioid cells. The MRI right foot showed well defined mass lesion in planter aspect of right heel which is slightly exophytic and nodular of size 1.8x1.9 cm in subcutaneous plane without any muscular and bony extension. The CECT thorax showed multiple variables sized pleural and parenchymal based soft tissue nodules seen scattered in B/L lung fields with largest of size 17x16 mm in postero-basal segment of LLL (Fig 3). Whole body FDG PET scan demonstrated metabolically active soft tissue lesion in right heel region with metabolic active / inactive B/L pulmonary nodules suggestive of pulmonary metastasis. The patient is presently started on anthracycline based chemotherapy and regular follow up in our institute.

Figure 1: Showing an ulcerative lesion on plantar aspect of heel.

Figure 2: Chest X-ray P-A view showing B/L multiple well defined nodular radio-opacities

Figure 3: Lung window axial images arranged cranio-caudally showing multiple well defined nodular opacities in b/l lung fields suggestive of metastasis.

Figure 4 (a): Dermal nodules revealing tumour epithelial cells forming focal cribriform pattern and intra-luminary papillary projections lined by cuboidal to columnar epithelial cells and suggestive of ADPAC. (HNE stain 100X)
Fig 4 (b): Micro photograph revealing dermal nodules without connection to overlying dermis with hyperplasia. (HNE stain, 40X)

Discussion
Digital papillary carcinoma (DPC) is a malignant tumour having eccrine differentiation occurring predominantly in digits. The tumour is rare, slow growing and was originally classified as benign neoplasm. DPC is an uncommon malignant adnexal neoplasm that was first classified by the WHO as a skin tumour in 2006. The condition was documented and established from 1979 until 1987, however, no large-scale study has been conducted due to the rarity of this condition. DPC was formerly classified as adenoma or adenocarcinoma, there were no clinical or histologic characteristics that can differentiate the tumours that are likely to locally recur or distantly metastasize. According to the Armed Forces Institute of Pathology (AFIP), DPC are divided with two low- and high- grade malignant potential groups. A series of 31 cases of DPC were published based on reports from three different institutions. Thirty-one cases were reviewed by at least two authors that identified several histologic features. However, these histopathologic features were not correlated with malignant potential. Clinically, DPC presents as solitary non-tender masses that arise from fingers and toes and/or adjacent skin of the palms and soles. The distal portion of a finger or thumb is frequently documented as a DPC location. The presence on site such as heel has not been reported in literature till date. Cases with minimal atypia that may have previously been classified as ADPA did develop into metastatic disease. Conversely, some cases with histologic features suggestive of malignancy had a benign course. Similarly, clinical parameters such as age at diagnosis, duration of tumour or tumour size, were not correlated with the aggressive behaviour of DPC.

Our patient had a 3-year history of a painless mass followed by trivial trauma to the same area. The biopsy and the systemic examination revealed the diagnosis and the extent of distal metastasis. However, it seems that a thorough examination and scanning is required for the early diagnosis, treatment and to rule out any distant metastasis. In our case, the site of lesion was highly unusual and it silently metastasised to the pulmonary system. This lesion had typical histologic features of ADPC with high mitotic activity and a high proliferation index.

Conclusion
It requires prudent approach and meticulous work up so as to diagnose early signs and symptoms for infrequently presenting lesions on distal extremities.

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