

Hook and Terminus-Digital Papillary Adenocarcinoma

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Digital papillary adenocarcinoma emerges as a malignant adnexal tumour. The neoplasm exhibits a significant predilection for and appears confined to acral sites. Neoplasm is additionally designated as digital papillary eccrine adenoma, aggressive digital papillary adenoma or digital adenocarcinoma.

Initially scripted by Helwig as an eccrine acrospiroma in 1979, tumefaction is commonly confined to fingers and toes. The palm and soles are infrequently involved.

Clinically, tumefaction may be misinterpreted as a cystic lesion. Neoplasm is constituted of basaloid, cuboidal or low columnar epithelial cells admixed with myoepithelial cells. Tumour cells may display mild to moderate cytological atypia.

A male preponderance is observed with male to female proportion of ~4:1. Neoplasm emerges between 14 years to 83 years with average age of disease occurrence at 52 years. Frequently, the Caucasian population is implicated [1,2].

Commonly, neoplasm arises within digits as fingers and toes. In contrast to the feet, lesion frequently occurs within the hands (> 80%). Exceptionally, sites as the palm, sole, facial region or thigh may be implicated [1,2].

As digital papillary adenocarcinoma configures as an eccrine tumour, involved acral sites display an abundance of eccrine glands.

Certain neoplasms may arise on account of trauma to site of disease emergence [1,2].

Digital papillary adenocarcinoma emerges as a gradually progressive, asymptomatic tumour nodule varying from 0.4 centimetres to 4.3 centimetres diameter with average magnitude of 1.7 centimetres. Tumefaction may be misinterpreted as a cystic lesion [2,3].

Ulceration of superficial cutaneous surface is exceptionally encountered. Tumours invading subjacent bone, joint or superficial nerves may be associated with pain. Exceptionally, distant metastasis into pulmonary parenchyma or infiltration of regional lymph nodes may ensue.

Tumour cells depict BRAFV600E genetic mutations along with overexpression of FGFR2 gene [2,3].

Upon microscopy, a well circumscribed, multinodular tumefaction appears confined to dermis or superficial subcutaneous tissue. Neoplasm is constituted of an amalgamation of dual solid and cystic segments. Besides, papillary projections, tubular articulations or ductal structures may be discerned. Occasionally, tumefaction may depict an infiltrative neoplastic perimeter.

Infrequently, morphological features as epidermal hyperplasia, superficial cutaneous ulceration or focal contiguity with superimposed epidermis may be discerned [3,4].

Tumour cells appear as basaloid, cuboidal or low columnar epithelial cells admixed with myoepithelial cells. Mild to moderate cytological atypia may be encountered. Severe cytological atypia or tumour necrosis is exemplified in a subset of neoplasms.

Mitotic figures may be inconspicuous or frequently discerned and vary from 1 to 15 mitosis per mm².

Foci of spindle shaped cells, clear cell alterations or squamoid differentiation may be observed [3,4].

Ultrastructural examination depicts specific categories of neoplastic cells denominated as clear cells, dark cells or myoepithelial cells. Few dark cells are impregnated with dense granules [3,4].

Primary tumour

- TX: Primary tumour cannot be assessed.
- T0: No evidence of primary tumour.
- Tis: Carcinoma *in situ*.
- T1: Tumour ≤ 2 centimetres in greatest dimension with < 2 high risk features.
- T2: Tumour > 2 centimetres in greatest dimension along with or in the absence of singular additional high risk feature OR tumour of variable magnitude along with ≥ 2 high risk features.
- T3: Tumour demonstrating invasion of orbit, maxillary, mandibular or temporal bone.
- T4: Tumour delineates invasion of axial skeleton or appendicular skeleton or perineural invasion within nerves confined to base of skull.

Regional lymph nodes

- NX: Regional lymph nodes cannot be assessed.
- N0: Regional lymph node metastases absent.
- N1: Metastasis confined to singular ipsilateral lymph node ≤ 3 centimetres in greatest dimension.
- N2: Metastasis confined to singular ipsilateral lymph node >3 centimetres and ≤ 6 centimetres in greatest dimension OR within multiple ipsilateral lymph nodes ≤ 6 centimetres in greatest dimension OR within bilateral or contralateral lymph nodes ≤ 6 centimetres in greatest dimension.
- N2a: Metastasis confined to singular ipsilateral lymph node >3 centimetres and ≤ 6 centimetres in greatest dimension.
- N2b: Metastasis confined to multiple ipsilateral lymph nodes ≤6 centimetres in greatest dimension.
- N2c: Metastasis within bilateral or contralateral lymph nodes ≤ 6 centimetres in greatest dimension.
- N3: Metastasis confined to a lymph node > 6 centimetres in greatest dimension.

Distant metastasis

- M0: Distant metastasis absent.
- M1: Distant metastasis present into soft tissue or various and specific sites [3,4].

Stages of non melanocytic cutaneous carcinoma

- Stage 0: Tis, N0, M0.
- Stage I: T1, N0, M0.
- Stage II: T2, N0, M0.
- Stage III: T3, N0, M0 or T1, T2, T3, N1, M0.
- Stage IV: T1, T2, T3, N2, M0 or any T, N3, M0 or T4, any N, M0 or any T, any N, M1 [3,4].

Digital papillary adenocarcinoma is diffusely immune reactive to pan-cytokeratin, CK7, S100 protein, oestrogen receptors (ER), progesterone receptors (PR) or androgen receptors (AR).

Intrinsic cellular layer appears immune reactive to epithelial membrane antigen (EMA) or carcinoembryonic antigen (CEA) which highlights luminal border of the tubules [4,5].

Extraneous myoepithelial cell layer appears immune reactive to smooth muscle actin (SMA), calponin, p63 or focally immune reactive to D2-40 and CK14 [4,5].

Digital papillary adenocarcinoma necessitates segregation from neoplasms as hidradenoma, papillary eccrine adenoma, apocrine hidrocystoma or apocrine cystadenoma, microcystic adnexal carcinoma, hidradenocarcinoma, epithelial-myoepithelial carcinoma or adenomyoepithelial tumour and distant metastasis from various primary adenocarcinomas [4,5].

Clinically, emergence of digital papillary adenocarcinoma may be indicated by site of origin of the neoplasm and associated biological course.

Histological assessment of surgical tissue samples may be appropriate for definitive neoplastic discernment [5,6].

Comprehensive surgical extermination of the neoplasm or amputation of implicated digit is an optimal mode of therapy.

Sentinel lymph node biopsy may be beneficially adopted in order to achieve decimated localized tumour recurrence or distant metastasis. Adoption of adjuvant chemotherapy remains non-beneficial and therapeutic outcomes remain debatable.

Conventional radiation therapy appears efficacious upon employment of palliative doses.

Gene therapy targeting FGFR gene or BRAF gene appears advantageous [5,6].

Morphological features appear non-concurrent with prognostic outcomes.

Distant metastasis into pulmonary parenchyma or regional lymph node metastasis are exceptionally observed. Localized tumour recurrence appears in up to 21% neoplasms and distant metastasis occurs in up to 50% lesions. Disease-associated mortality occurs within 2.1% subjects.

Extensive, meticulous long-term follow-up appears mandatory as the neoplasm demonstrates a protracted clinical course and delayed emergence of distant metastases [5,6].

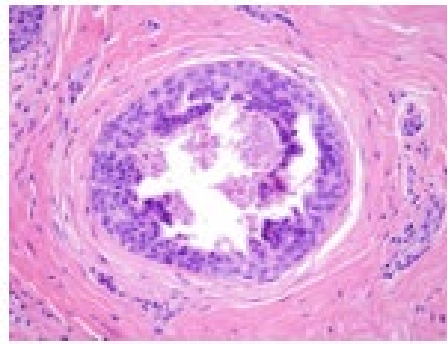


Figure 1: Digital papillary adenocarcinoma demonstrating dual component of cystic and solid areas and ductal structures layered by cuboidal to columnar epithelial cells admixed with myoepithelial cells. Surrounding stroma is dense and fibrotic [7].

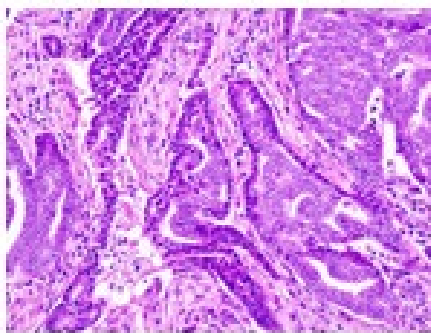


Figure 2: Digital papillary adenocarcinoma delineating dual component of cystic and solid areas and ductal structures lined by cuboidal to columnar epithelial cells admixed with myoepithelial cells. Surrounding stroma is dense and fibrotic [8].

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7. Image 1 Courtesy: Dermatopathology.com.
8. Image 2 Courtesy: Pathology outlines.

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