# An exophytic growth at the genitalia – A rare case report of primary cutaneous apocrine adenocarcinoma

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A 77-year-old Chinese male presented to urology clinic for evaluation of a fungating mass over his suprapubic, penoscrotal region. The mass originated as a nodule located on the proximal penile shaft, progressively enlarging over two years. There was tenderness and contact bleeding with serous discharge from the mass. He had significant weight loss of 5kg over 2months. He denied contact with commercial sex workers, nor application of any creams to the affected area. On examination, a large fungating, exophytic, verruciform growth was noted on the suprapubic region and on the mid-distal penile shaft, along with multiple fleshy, satellite 5-10-millimeter lesions around the suprapubic and scrotum(Figure 1). The testes were normal. Flexible cystoscopy was unremarkable without evidence of bladder involvement. CTTAP revealed multiple pulmonary and liver lesions suspicious for metastatic disease. The patient underwent punch biopsy of the tumor. Histologic evaluation demonstrated densely packed nests of malignant cells centered within the dermis, with occasional pagetoid spread into the epidermis. Lesion cells contained moderate amounts of amphophilic cytoplasm and enlarged oval-shaped nuclei with vesicular chromatin pattern and small but visible nucleoli(Figure 2A). No definite evidence of squamous or basaloid differentiation was observed. The tumor cells showed diffuse expression of CK7 (Figure 2B), negative for CK20, Melan A, SOX10, NKX3.1 or PSA.

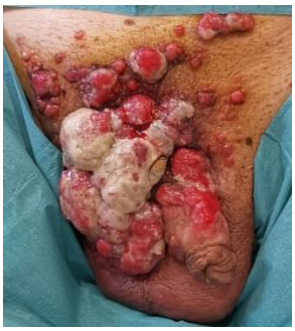

**Figure 1**. multiple fleshy exophytic growth on suprapubic region and on right side of penile shaft, involving penile shaft skin, along with multiple satellite lesions

**Discussion**

The patient was diagnosed with primary cutaneous apocrine gland adenocarcinoma(PAA) of the genital area. PAA is a carcinoma with potential destructive local growth and regional/distant metastasis. It primarily affects patients in their late seventies, without racial or gender predilection. It frequently involves apocrine gland abundant areas such as the axilla (50%) followed by the head and neck (35%). Other affected the locations including auditory canal, nipple and perineum have rarely been reported. Biopsy should be obtained for histologic correlation. The differential diagnoses for this lesion include squamous cell carcinoma, extramammary Paget disease, apocrine hidrocystomas, apocrine cystadenomas and primary cutaneous mucinous carcinoma. PAA are frequently indolent and slowly progressive and may take time to manifest, as was the case in our patient, but can also present rapidly and aggressively. Adnexal carcinomas that develop de novo can be difficult to diagnose clinically. It is important to rule out cutaneous metastasis of adenocarcinomatous origin. GCDFP-15 has been known to stain apocrine rather than eccrine glands in axillary and anogenital skin and CK5/6 and CK7 are useful to differentiate a PAA from a metastasis. CK5/6 positivity has been reported in 80% of PAA but only occurs in 6.66% of metastasis. Adipophilin, mammaglobin can also be useful in distinguishing PAA from metastatic apocrine adenocarcinomas. Metastatic mammary adenocarcinomas are often also positive for estrogen and progesterone receptors.

As PAA are a rare tumor, the available treatments are based on sporadic case reports and there are no guidelines to support management. Multidisciplinary management is critical, and options reported in literature include surgical excision, chemotherapy, and radiotherapy. Surgical excision in the form of wide, local excision with clear margins or without lymph node dissection is the treatment of choice. Chemotherapy regimens include cisplatin, adriamycin and cyclophosphamide[8]. PAA is histologically similar to apocrine subtype of breast cancer and studies have shown increased expression of surface markers like GCDFP-15 and HER-2. RANKL was also noted to be involved in maintaining the microenvironment for tumors of apocrine origin. Immunomodulation can be an option but it is still an experimental method. Radiotherapy could be considered if patient declined surgery, or as adjuvant to surgery in patients with large or poorly differentiated tumors or when margins of resection are positive. In a case report, radiotherapy was used as a primary treatment when the extension of tumor invasion was not optimal for surgical treatment especially with the presence of regional cutaneous metastasis. Close follow up is recommended to detect recurrence.

Our patient was suffering with local complications from the fungating tumor. After a multidisciplinary meeting and prehabilitation, he underwent palliative wide local excision and reconstruction with scrotal advancement flap and split skin graft. Post-operative recovery was uneventful, and the wound healed with secondary intention. The patient has better quality of life after surgery. However, after extensive discussion with medical oncology team, his prognosis was deemed poor and the patient decided against palliative chemotherapy.

Primary cutaneous apocrine adenocarcinoma is a very rare tumor and often presents in an advanced stage. Few case reports have suggested curative treatment in early stage disease, but presentation with metastatic disease has a very poor prognosis. Palliative surgery to alleviate local symptoms and to improve quality of life can be undertaken with careful preoperative planning.

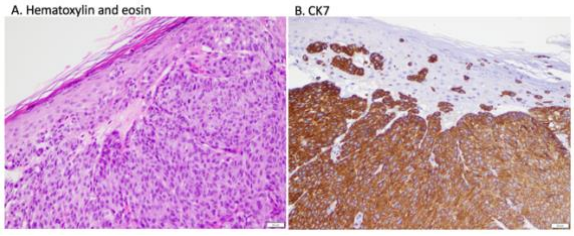


**Figure 2**.

A. Hematoxylin and eosin reveals nests of malignant cells within the dermis, with no definite evidence of squamous or basaloid defferentiation (original magnification x 200)

B. Immunoperoxidase slide of CK7 highlights PAA tumor cell with diffuse expression of CK7 within dermis (original magnification x 200)