Case Report

Primary cutaneous mucinous carcinoma of the scrotum: A rare tumor at a rare site – A case report and review of literature

Olajide Olusegun Abiola, Gbemi H. Ano-Edward[1], Olutunde A. Oluwumi[2], Mathew E. Lasisi[1]

Department of Surgery, Urology Unit, Bowen University Teaching Hospital, [1]Department of Anatomic Pathology, Bowen University Teaching Hospital, Ogbomoso, Oyo State, [2]Department of Anatomic Pathology, University of Ilorin Teaching Hospital, Ilorin, Kwara State, Nigeria

Abstract

Primary cutaneous mucinous carcinoma (PCMC) is a rare malignant adnexal tumor of eccrine origin with a higher incidence in the head and neck region of the body while scrotal skin involvement is rare. We report a case of a 72-year-old man with ulceration of painless scrotal lesions of 10 years' duration. Histology of the wedge biopsies of the lesions was mucinous carcinoma. Clinical workup excluded noncutaneous primary sites of mucinous carcinoma and distant metastasis. He had wide excision of the scrotal skin lesions with histological findings of nests of malignant cells separated from pool of mucin by fibrocollagenous septae. Periodic acid–Schiff stain was positive; however, immunohistochemical stains for estrogen and progesterone receptors were negative. No local recurrence was observed after 12 months of follow-up. Scrotal PCMC is a rare tumor; this may probably be the first reported case of this tumor in the scrotum. Prognosis was good following surgical excision.

Keywords: Cutaneous lesions, mucinous carcinoma, scrotum

Address for correspondence: Dr. Olajide Olusegun Abiola, Department of Surgery, Urology Unit, Bowen University Teaching Hospital, P.O. Box 15, Ogbomoso, Oyo State, Nigeria.
E-mail: ooabiola2015@gmail.com
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INTRODUCTION

Primary cutaneous mucinous carcinoma (PCMC) is a rare malignant adnexal tumor of the skin with some controversy about its origin from either apocrine or eccrine sweat glands of the skin.[1,2] Recent evidence from immunohistochemical studies and ultrastructural analysis with electron microscope support an eccrine origin.[3] It is also referred to as mucinous eccrine carcinoma and mucinous adenocarcinoma of the skin.[3-5] The prevalence rate is unknown; however, an incidence rate of <0.1 per million people was reported by Breiting *et al.*[6] in a Danish population-based study.

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The tumor commonly arises in the head and neck region with the eyelid being the most common site; involvement of other regions of the body that have been reported are the scalp, face, ear, canthus, neck, axilla, chest/abdominal wall, extremity, groin, vulva, and the penis.[3-5,7]

Diagnosis is by histological examination of the excised skin lesion and clinical workup to exclude primary sites of noncutaneous mucinous carcinoma and metastasis to the skin.[3,4,6] Surgical excision is the mainstay of treatment.

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The disease has a good prognosis, but local recurrence of the tumor is common.[3,4,8]

We report this case due to its rarity, especially in the scrotum; also, this adds to the knowledge of differential diagnosis of malignant scrotal skin lesion.

CASE REPORT

We report a case of a 72-year-old Nigerian with a 10-year history of scrotal skin nodules. Six months before presentation, the nodules ulcerated with copious mucopurulent discharge. On physical examination of the external genitalia, there were fungating solitary masses, measured 4 cm × 6 cm and 2 cm × 4 cm on the left and right hemiscrotum, respectively. The masses were hard in consistency, attached to scrotal skin but free from the underlying scrotal content, and with no contact bleeding. The testicle was palpably normal in each hemiscrotum. There were palpable, tender, and discrete right inguinal lymph nodes. His prostate was enlarged but feels benign on examination.

Initial wedge biopsy of the ulcerated lesions revealed mucinous carcinoma. Negative abdominopelvic computed tomography scan, colonoscopy, chest X-ray, and scrotal ultrasonography ruled out noncutaneous primary sites of mucinous carcinoma with possible metastasis to the scrotum. Prostate-specific antigen was 1.0 ng/dL.

Subsequently, he had wide excision of the masses with skin margin of 10 mm and right superficial inguinal lymphadenectomy [Figure 1]. Histology of the excised masses confirmed mucinous carcinoma with histological findings of nests of malignant cells disposed in glandular pattern and separated from pools of extracellular mucin by fibrocollagenous septae. Periodic acid–Schiff (PAS) stain was positive [Figure 2]. The surgical margins were free of malignant cells. The dissected inguinal lymph nodes show infiltration by inflammatory cells with no malignant cells. Immunohistochemical stains were negative for estrogen receptors (ERs) [Figure 3] and progesterone receptors (PRs) [Figure 4].

A diagnosis of PCMC was made. He was followed up in the urology clinic for over a year with no local recurrence observed.

DISCUSSION

PCMC is a rare tumor of the sweat glands with a higher incidence in the head and neck region of the body.[3,9] The occurrence of the lesion in other parts of the body such as the axilla, chest/abdominal wall, extremity, groin, vulva, and the penis has been reported;[3-5,7] however, the

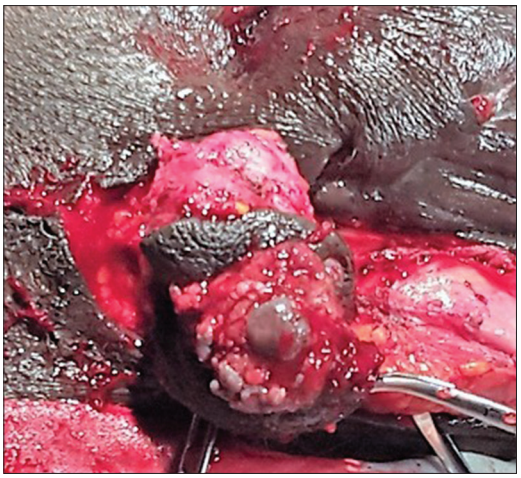

Figure 1: Wide excision of the scrotal lesion

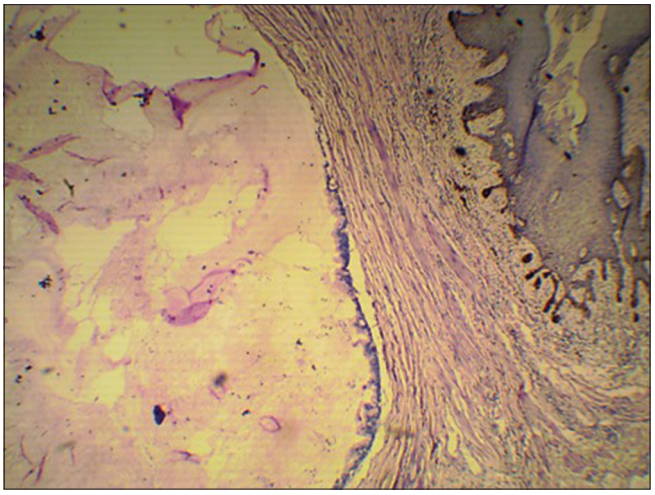

Figure 2: Histograph of mucinous carcinoma of the scrotum Periodic acid–Schiff ×40

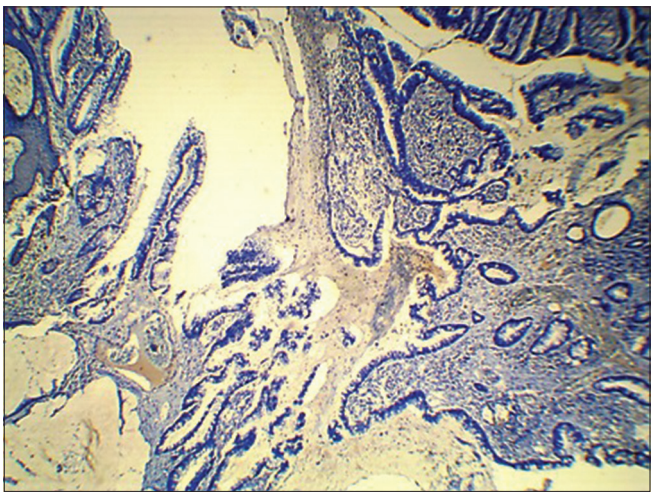

Figure 3: Histograph of immunohistochemical stain of estrogen receptors in mucinous carcinoma of the scrotum ×40

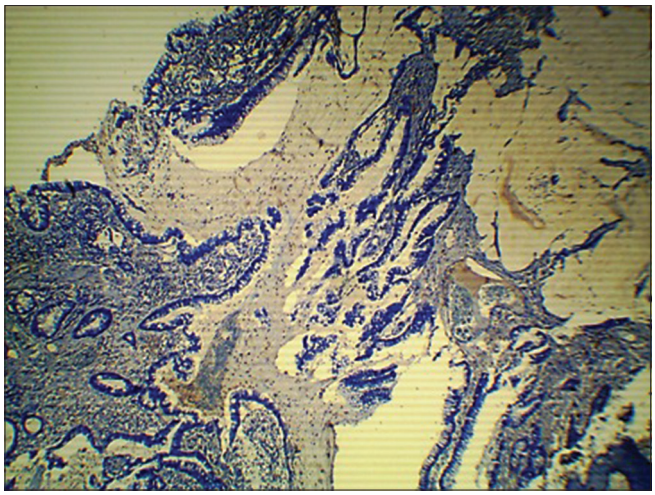

Figure 4: Histograph of immunohistochemical stain of progesterone receptors in mucinous carcinoma of the scrotum ×40

authors found no documented involvement of the scrotum in the literatures.

PCMC is a slow-growing/indolent tumor which rarely metastasizes except to regional lymph nodes, and local recurrence is common.[3,4,8-10] However, there has been a reported case of distant metastasis to the bone from the primary site of the tumor.[11]

In a review of literature by Martinez and Young,[3] the tumor was observed to occur more in the elderly with an average age of 63 years. Furthermore, its occurrence was found more among Caucasians compared to African-Americans and Asians.

The lesion usually presents as painless, papular, or nodular lesions of varying sizes; but, it may be seen as an isolated lesion reddish/pinkish/grayish/bluish or purplish in color. Telangiectasia may or may not be present. The surface may appear smooth, ulcerated, or crusted.[3,4,12] The lesion being a slow-growing lesion over a period of years may be mistaken for a benign lesion.[4] The patient reported presented with scrotal skin nodules of a period of 10 years which was slow growing until 6 months before presentation when ulceration of the nodules with mucopurulent discharge was observed.

Differential diagnosis of PCMC includes sebaceous cyst, hemangioma, adenoid cystic carcinoma, lipoma, melanoma, epidermal inclusion cyst, Kaposi's sarcoma, squamous cell carcinoma, and metastatic tumors to the scrotal skin.[3,4,6]

In diagnosis of PCMC, it is important to exclude the possibility of noncutaneous visceral mucinous carcinoma with metastasis to the skin.[6,10,13] Mucinous carcinoma has been reported to occur in other viscera such as the breast, gastrointestinal tract, ovary, lung, salivary gland, paranasal sinuses, and lacrimal glands.[3,9,10] Hence, a thorough workup to identify possible primary sites of metastasis to the scrotum is important. Common primary tumor sites which may metastasize to scrotal skin are colon/rectum, prostate, lungs, and testicles.[14,15] Therefore, a complete clinical history, physical examination, investigations such as colonoscopy or barium enema, scrotal ultrasound, prostate-specific antigen, trans-rectal ultrasonography, computed tomography scan of the chest and abdominopelvic region are of great value.

Diagnosis of mucinous carcinoma is by excisional biopsy with histological findings of nests of tumor cells separated from pools of mucin by fibrocollagenous septae.[3-6,9] The malignant cells may line the wall of cystic degeneration with the cells mostly cuboidal to oval with pleomorphic hyperchromatic nuclei, prominent nucleoli, abundant eosinophilic cytoplasm that stains magenta with PAS, and few mitotic figures;[3,4] a similar finding was found in the case reported.

Primary lesions histologically have more organized epithelial cells, fewer mitoses, and less hyperchromasia in individual cells compared to metastatic lesions.[12] However, it is difficult to distinguish primary mucinous carcinoma from noncutaneous metastatic mucinous carcinomas of the skin by routine histological examination.[16] Immunohistochemical staining has been found to be helpful in differentiating primary from metastatic mucinous carcinoma of the skin.[16,17] Immunohistochemical markers which are often expressed by the tumor include cytokeratin (CK)-7, epithelial membrane antigen, ER, and PR.[3,17,18] A similarity between cutaneous mucinous carcinoma and mammary mucinous carcinoma in expression of ER and mucus-associated peptides of the trefoil factor family (TFF1 and TFF3 mRNA) was observed by Hanby *et al*.[17] The absence of expression of CK-20 may exclude the diagnosis of metastatic colorectal mucinous carcinoma which often expressed CK-20.[16] However, in the index case reported, immunohistochemical stains for ER and PR were negative.

Evidence-based treatment guideline for PCMC is yet to be available; however, like other skin tumors, surgical excision remains the mainstay of treatment.[8] Complete excision of the lesion is essential for cure as incomplete excision can lead to local recurrence which is resistant to both radiation and chemotherapy.[4,8,13] Like other malignant skin lesions, wide excision with sufficient skin margin to achieve a tumor-negative margin is of essence.[8] Although there is yet to be a recommended surgical margin for wide excision of the lesion, a margin of at least 10 mm could be advocated

based on the findings of Martinez and Young[3] in which in a review of cases of PCMC, an average margin of 12.5 mm in the least dimension was observed. In the case reported, wide excision with a 10 mm skin margin was done, and a surgical free margin was observed histologically.

Moh's micrographic surgery may be an alternative to wide excision for smaller superficial lesion, especially in areas where wide excision may be difficult considering anatomic or cosmetic restraint.[3,19]

In the index scrotal PCMC, the superficial inguinal lymph nodes which drain the scrotal skin was enlarged and tender; hence, lymphadenectomy was done. However, palpable inguinal lymph nodes in such situations may be an inflammatory reaction as it was observed in the case reported. Medical therapy with antiestrogen has been suggested based on the shared similarity with mammary tumor in expression of ER and mucosal repair-related trefoil factor protein (TFF1).[4,17]

In a systematic review and meta-analysis of outcomes of PCMC after surgery, a better surgical outcome was observed in older patient and Asians while lesions on the trunk were associated with worse outcome.[8] The surgical outcome of the index case was good; no local recurrence of the lesion was observed after wide excision with a follow-up period of more than a year despite being of a black race and location of the lesions on the trunk (scrotum). However, further follow-up of the patient is pertinent for prompt detection of local recurrence of the lesion.

CONCLUSION

PCMC of the scrotal skin is a very rare tumor; this may probably be the first reported case of PCMC of the scrotum. Prognosis in PCMC is favorable, but local recurrence following surgical excision is a frequent complication, especially in the absence of a surgical free margin like most skin malignant lesions.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient has given his consent for his images and other clinical information to be reported in the journal. The patient understands that name and initials will not be published and due efforts will be made to conceal identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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