Primary cutaneous mucinous carcinoma of the penis

Michael Robinson, MD;[*] John Kelly, MD;[†] Darren Biberdorf, MD;[‡] Iain McAuley, MD[§]

[*]Department of Urologic Sciences, University of British Columbia, Vancouver, BC; [†]Department of Family Practice, Vancouver Island Health Authority, Victoria BC; [‡]Division of General Surgery, Vancouver Island Health Authority, Victoria, BC; [§]Division of Urology, Vancouver Island Health Authority, Victoria, BC.

Cite as: *Can Urol Assoc J* 2014;8(1-2):e89-91. http://dx.doi.org/10.5489/cuaj.1549
Published online February 12, 2014.

Abstract

Primary cutaneous mucinous carcinoma (PCMC) is a rare, slow growing, malignant neoplasm arising from the sweat glands. Lesions are most commonly seen on the head and neck, and frequently mistaken for metastases from more common primary sites, such as the breast, gastrointestinal tract, lung, ovary, or prostate. We present what we believe is the first reported case of PCMC involving the penis.

Introduction

Primary cutaneous mucinous carcinoma (PCMC) is a rare, slow growing, malignant neoplasm arising from the sweat glands.[1] Lesions are most commonly seen on the head and neck, and frequently mistaken for metastases from more common primary, sites such as the breast, gastrointestinal tract, lung, ovary or prostate. First described by Lennox and colleagues in 1952,[2] PCMC is also known as mucinous adenocystic carcinoma, primary mucinous carcinoma of the skin and colloid carcinoma.[3] We present what we believe is the first reported case of PCMC involving the penis.

Case report

A 65-year-old healthy male presented to our urology clinic with recurrence of a penile lesion that had been excised 8 years earlier by his family doctor. The initial lesion was described as roughly a 5-mm soft, round nodule. Unfortunately, the pathology report for this specimen was not available in the patient's medical record. The patient was otherwise healthy, but did have a strong family history for gastrointestinal cancers (mother, maternal grandparents, maternal aunt). In light of this history alone, he underwent unremarkable upper endoscopy and colonoscopy in 2009. Additional history of potential relevance is of a herpetiform exanthem involving the penis in 1987, which resolved with no further issue following antiviral treatment. The patient also described a chemical burn to his penis in 1990 after an organic solvent spill to his groin. This particular agent contained a number of aromatic petrochemicals, including toluene and benzene. This exposure was apparently quite painful, but resolved spontaneously over several days.

In January 2013 we excised this penile lesion from the dorsal aspect of his proximal penile shaft. Grossly the appearance of this lesion was consistent with a keloid or hypertrophic scar. Surgical pathology confirmed a central, pale tan nodule measuring 2.4 × 1.4 × 0.5 cm. Microscopic evaluation demonstrated positive lateral and deep margins with histology typical of mucinous carcinoma. Immunohistochemical analysis demonstrated E receptor positivity and was negative for CDX2 and TTF1, supporting the diagnosis of PCMC. These findings were consistent on independent review by another experienced pathologist at the hospital. A small minority of cells were also positive for CK20, prohibiting the exclusion of a primary source from GI or other organ system. Due to the inherent difficulty in differentiating in PCMC from metastatic mucinous carcinoma, the patient underwent metastatic workup with computed tomography of the chest, abdomen and pelvis, as well as a bone scan. These investigations were negative for metastatic disease. Hematologic, renal and hepatic profiles were within normal limits.

Following the negative metastatic workup, we brought the patient back for excision of his scar. A 1-cm margin was taken around the scar and the deep margin extended to Buck's fascia with sparing of neurovascular structures. Surgical margins at the lateral aspect returned positive with identical histology as previously reported (Fig. 1). The patient was then brought back for a third procedure, and again, a 1-cm margin was taken around the previous scar. Margins for this final specimen were negative.

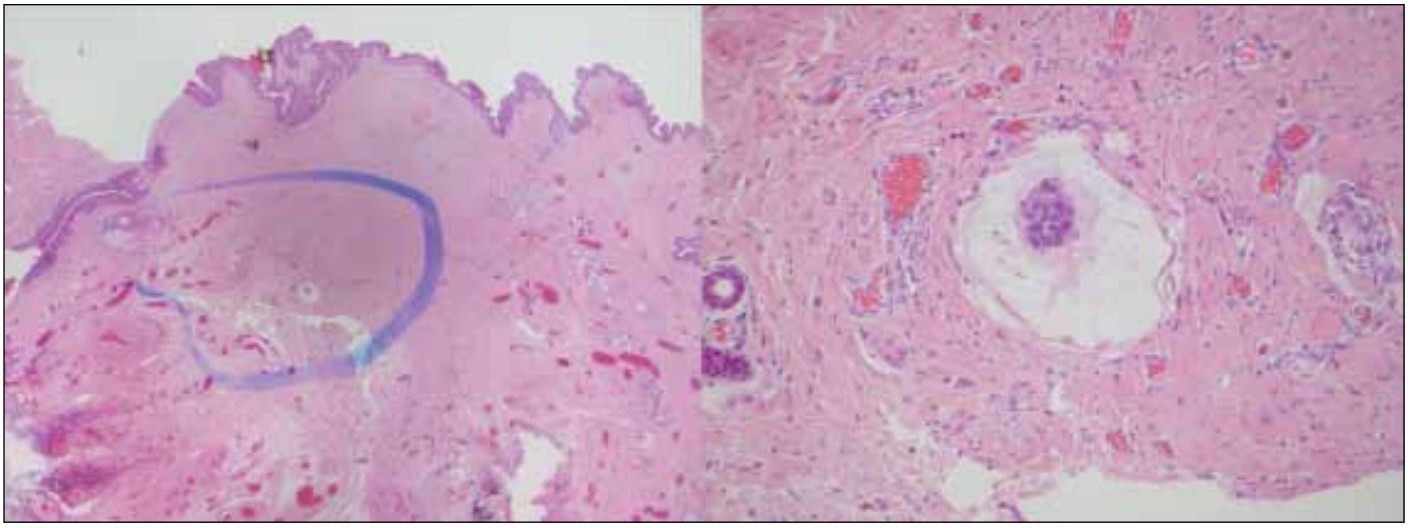

Fig. 1. Photomicrograph hematoxylin and eosin stain (H&E) stained sections of residual disease at low and high power magnification demonstrating an island of uniform appearing epithelial cells floating in a pool of mucin.

Discussion

The incidence of PCMC is about 0.07 per million and most often occurs during the 7th decade of life.[4] Local recurrence is as high as 33%, but metastasis is rare.[4] Only 3 cases of death attributed to metastatic disease have been reported.[5-7] In a previous review of the literature that identified 206 published cases, 150 cases involved the head (eyelids, face, scalp, neck), while 7 involved the vulva.[8] There were no cases of PCMC involving the penis identified in our search of the PubMed database limited to publications in English.

On histology, PCMC is typically described as nests of epithelial cells floating in lakes of extracellular diastase-resistant, periodic-acid Schiff (PAS)-positive mucin.[3,9] PCMC also expresses low-molecular weight cytokeratins (cytokeratin-7, CAM 5.2), CEA antigen, epithelial membrane antigen, gross cystic disease fluid protein, S-100 protein, human milk factor globulins, alpha-lactalbumin, TTF1 and 3, and estrogen and progesterone receptors.[10]

In spite of helpful immunohistochemical characteristics, misdiagnosis as a cutaneous metastasis from more common sites of mucinous adenocarcinoma, such as breast, lung, gastrointestinal tract, prostate and ovary, is common. Many tumour cells stain positively for site specific markers of these other organs.[3,9,11] Final diagnosis, therefore, can only be made through clinical investigation to exclude a primary tumour elsewhere.[4] Recommended treatment includes standard to wide local excision.[2,3,9,12] Other approaches include Mohs surgery,[13] the addition of regional node dissection,[4] and the use of adjuvant hormone therapy with antiestrogenic agents, such as tamoxifen, to reduce the risk of recurrence.[10] Recurrent PCMS appears to be resistant to both chemotherapy and radiation.[14] Some groups suggest yearly follow-up after excision to monitor for recurrence and exclude the rare case where a tumour lacking in situ elements actually represents metastasis from another site.[4]

Conclusion

PCMS is a rare malignant neoplasm with low propensity for metastasis. PCMC most commonly involves the head, and we report the first case to our knowledge involving the penis. While characteristic immunohistochemical features aid in the diagnosis, exclusion of a primary tumour elsewhere remains essential. Surgical excision is the standard for treatment and patients should be followed to monitor for recurrence.

Competing interests: Dr. Robinson, Dr. Kelly, Dr. Biberdorf and Dr. McAuley all declare no competing financial or personal interests.

This paper has been peer-reviewed.

References

1. Mendoza S, Helwig EB. Mucinous (adenocystic) carcinoma of the skin. *Arch Dermatol* 1971;103:68-78. htt p://dx.doi.org /10.1001/archderm .197 1.04000130070011
2. Lennox B, Pearse AG, Richards HG. Mucin-secreting tumours of the skin with special reference to the so-called mixed-salivary tumour of the skin and its relation to hidradenoma. *J Pathol Bacteriol* 1952;64:865-80. htt p://dx.doi.org /10.100 2/path.1700640418
3. Martinez SR YS. Primary mucinous carcinoma of the skin: A review. *Int J Oncol* 2005;2:June 24, 2013.
4. Breiting L, Christensen L, Dahlstrom K, et al. Primary mucinous carcinoma of the skin: A population-based study. *Int J Dermatol* 2008;47:242-5. http://dx.doi.org/10.1111/j.1365-4632.2008.03558.x
5. Wright JD, Font RL. Mucinous sweat gland adenocarcinoma of eyelid: A clinicopathologic study of 21 cases with histochemical and electron microscopic observations. *Cancer* 1979;44:1757-68. http://dx.doi.org /10. 1002/1 097-0142(197911)44:5<1757::AID-CNC R2820440533>3.0.C 0;2-8

6. Rao KV, Tikku I, Kapur BM, et al. Invasive primary mucinous carcinoma of the skin. *Int Surg* 1978;63:168-70.
7. Jih MH, Friedman PM, Kimyai-Asadi A, et al. A rare case of fatal primary cutaneous mucinous carcinoma of the scalp with multiple in- transit and pulmonary metastases. *J Am Acad Dermatol* 2005;52:S76-80. http://dx.doi.org/10.1016/j.jaad.2004.06.047
8. Breiting L, Dahlstrom K, Christensen L, et al. Primary mucinous carcinoma of the skin. *Am J Dermatopathol* 2007;29:595-6. http://dx.doi.org/10.1097/DAD.0b013e318158d878
9. Kazakov DV, Suster S, LeBoit PE, et al. Mucinous carcinoma of the skin, primary, and secondary: A clinicopathologic study of 63 cases with emphasis on the morphologic spectrum of primary cutaneous forms: Homologies with mucinous lesions in the breast. *Am J Surg Pathol* 2005;29:764-82. http://dx.doi.org/10.1097/01.pas.0000159104.02985.6b
10. Hanby AM, McKee P, Jeffery M, et al. Primary mucinous carcinomas of the skin express TFF1, TFF3, estrogen receptor, and progesterone receptors. *Am J Surg Pathol* 1998;22:1125-31. http://dx.doi.org/10.1097/00000478-199809000-00012
11. Carson HJ, Gattuso P, Raslan WF, et al. Mucinous carcinoma of the eyelid: An immunohistochemical study. *Am J Dermatopathol* 1995;17:494-8. http://dx.doi.org/10.1097/00000372-199510000-00011
12. Bellezza G, Sidoni A, Bucciarelli E. Primary mucinous carcinoma of the skin. *Am J Dermatopathol* 2000;22:166-70. http:// dx.doi.org/10.1097/00000372-200004000-00014
13. Ortiz KJ, Gaughan MD, Bang RH, et al. A case of primary mucinous carcinoma of the scalp treated with mohs surgery. *Dermatol Surg* 2002;28:751-4. http://dx.doi.org/10.1046/j.1524-4725.2002.02007.x
14. Snow SN, Reizner GT. Mucinous eccrine carcinoma of the eyelid. *Cancer* 1992;70:2099-104. http://dx.doi.org/10.1002/1097-0142(19921015)70:8<2099::AID-CNCR2820700815>3.0.CO;2-C

Correspondence: Dr. Michael Robinson, Department of Urologic Sciences, University of British Columbia, Vancouver, BC; m_rob13@hotmail.com