SKIN

BRIEF ARTICLE

Multiple Sebaceous Carcinomas in an African American Patient with Muir-Torre Syndrome

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INTRODUCTION

Muir-Torre syndrome (MTS), a variant of hereditary nonpolyposis colorectal cancer (HNPCC) or Lynch Syndrome, is a rare autosomal dominant condition that is seen in adult patients. It is characterized by visceral malignancies and skin tumors, most notably sebaceous neoplasms. Although sebaceous neoplasms are typically slow growing and benign, sebaceous carcinomas can be aggressive and warrant early intervention.[1] There are no reports in the literature describing sebaceous carcinoma in African American patients with MTS. We present a 40-year-old African American patient with multiple sebaceous carcinomas in the setting of MTS.

CASE REPORT

A 40-year-old African American male presented to dermatology clinic with two nontender non-draining “cysts” on the back. The patient reports these lesions appeared and grew rapidly over the previous 1-2 months. Past medical history was notable for end-stage renal disease and hidradenitis suppurativa. Physical exam (Figure 1A – 1C) revealed lesion A, a soft fleshy hyperpigmented papule on the left back with heme crusting and erythema and lesion B, a large 3 x 4 cm broad-based hyperpigmented and pink pedunculated plaque with comedones on the left back.

A Shave biopsy was performed on lesion A and excisional biopsy was performed for lesion B. Initial differential diagnosis included inflamed intradermal nevus, inflamed seborrheic keratosis, cyst, or hidradenitis suppurativa in the setting of history of axillary Hurley stage II disease. Histopathology of lesions A and B revealed sebaceous carcinoma.

Upon further discussion, the patient mentioned a history of colon cancer, diagnosed at 34 years old. Family history was notable for colon and pancreatic cancer in his mother who carried a diagnosis of Lynch Syndrome. Family history further revealed history of pancreatic cancer in the maternal grandmother. The patient’s Mayo Muir-Torre syndrome risk score was equal to 3, suggesting an increased likelihood of MTS.[2] Immunohistochemical stains for mismatch repair gene expression revealed tumor cells positive for MLH1, PMS2, and MSH6 and negative for MSH2 indicating microsatellite instability. The patient underwent Mohs micrographic surgery (MMS) to treat the sebaceous carcinomas on the left back, with both requiring one stage for clearance. At the time of surgery, an additional suspicious skin-colored nodule

was noted in the groin, lesion C (not pictured). A scout biopsy was performed with histopathology revealing sebaceous carcinoma. The patient subsequently underwent MMS with one stage required for clear margins.

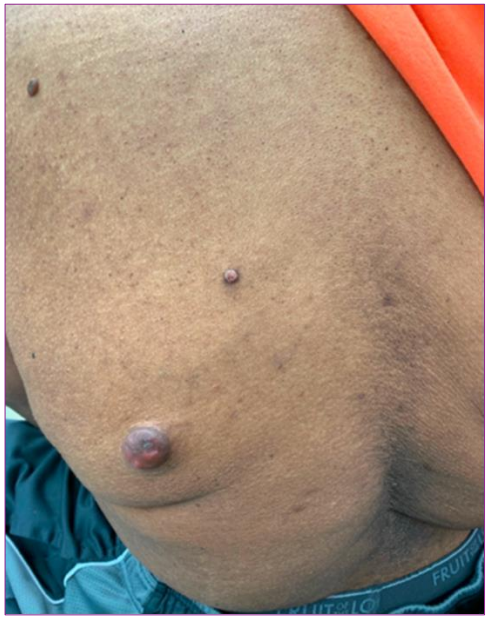

Figure 1A. Left back showing lesions A and B, histologically confirmed to be sebaceous carcinomas

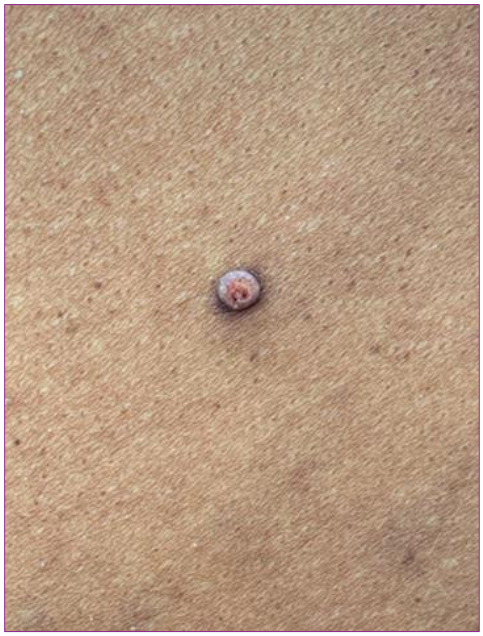

Figure 1B. Lesion A - left mid back. A soft fleshy hyperpigmented papule on the left back with heme crusting and erythema.

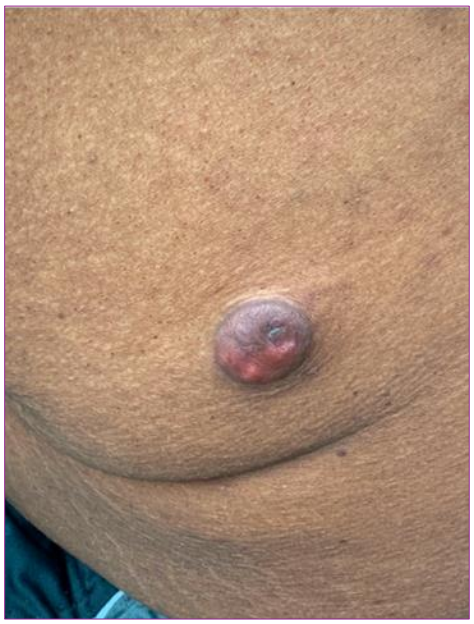

Figure 1C. Lesion B - left inferior back. A broad-based hyperpigmented and pink pedunculated plaque with comedones.

Mismatch Repair Gene Expression

MLH1: Expressed in tumor cells (Positive)
PMS2: Expressed in tumor cells (Positive)
MSH2: Not expressed in tumor cells (Negative)
MSH6: Expressed in tumor cells (Positive)

DISCUSSION

To our knowledge, there are currently only 3 cases of sebaceous neoplasms in African American patients described in the literature.[3,4,5] Of these, one case report details an African American patient with a diagnosis of MTS and a biopsy-proved sebaceous adenoma of the back.[3] Moon et al. reported one patient with a pruritic, erythematous, mildly lichenified plaque on the abdomen with biopsy revealing sebaceous carcinoma; this patient did not have an associated MTS.[4] Another author theorized the occurrence of sebaceous neoplasms in African Americans without MTS and a potential relationship between the immunosuppressive state in end-stage renal failure.[5,6] As noted in this patient, concomitant hidradenitis suppurativa disease could delay diagnosis if sebaceous neoplasms present within typical areas of HS disease (axilla, groin, buttocks, inframammary chest) or with similar clinical features, particularly follicular occlusion lesions.[7]

Clinicians should obtain a careful medical and family history when cyst-like lesions present with unusual features or in patients with known MTS. It is also important to consider differences in clinical appearance of sebaceous neoplasms in skin of color. Dermatologists should have a low threshold to sample suspicious lesions or refer for excisional biopsy if not amenable to simple shave removal. Specific staining patterns of MLH1, PMS2, and MSH6 positivity and MSH2 negativity are helpful to identify tumors with microsatellite instability and patients with risk of MSH2 germline mutation necessitating further work-up. In patients with diagnosed sebaceous neoplasms, the Mayo Muir-Torre syndrome risk score should be calculated.[2] For scores of 2 or greater, patients may have an increased likelihood of MTS and referral to appropriate subspecialities is pertinent.

Conflict of Interest Disclosures: None

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References:

1. John AM, Schwartz RA. Muir-Torre syndrome (MTS): An update and approach to diagnosis and management. J Am Acad Dermatol. 2016;74(3):558-566. PMID: 26892655
2. Roberts ME, Riegert-Johnson DL, Thomas BC, et al. A clinical scoring system to identify patients with sebaceous neoplasms at risk for the Muir-Torre variant of Lynch syndrome. Genet Med. 2014;16(9):711-716. PMID: PMID: 24603434
3. Wargo JJ, Plaza JA, Carr D. Sebaceous neoplasia leading to the diagnosis of Muir-Torre syndrome in an African American man. JAAD Case Rep. 2021;11:72-73. Published 2021 Mar 23. PMID: 33937470
4. Moon TC, Cassler NM, Lackey JN. Sebaceous carcinoma on the abdomen in an African-American male patient. Indian Dermatol Online J. 2015;6(Suppl 1):S27-S29. PMID: 26904445
5. Kaminska EC, Iyengar V, Tsoukas M, Shea CR. Borderline sebaceous neoplasm in a renal transplant patient without Muir-Torre syndrome. *J Cutan Pathol*. 2013;40(3):336-340. PMID: 23174034
6. Descamps-Latscha B. The immune system in end-stage renal disease. *Curr Opin Nephrol Hypertens*. 1993;2(6):883-891. PMID: 7922228
7. Sabat R, Jemec GBE, Matusiak Ł, Kimball AB, Prens E, Wolk K. Hidradenitis suppurativa. Nat Rev Dis Primers. 2020;6(1):18. Published 2020 Mar 12. PMID: 32165620