Case Report: Sebaceous Carcinoma of the Areola in an Immunosuppressed Patient

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Introduction

Sebaceous carcinoma (SC) is a rare, aggressive skin cancer derived from the epithelium of the sebaceous glands typically found in a periocular distribution.1-4 Few cases have reported extracutaneous locations, to date. We present an unusual case of extracutaneous SC on the areola of the breast. Our case report is unique, given the rarity of the neoplasm, uncommon location, mode of occurrence, and chronic immunosuppressed state of our patient, secondary to renal transplantation. Moreover, an association with sporadically occurring SC in both renal transplant recipients (RTTs) and chronic immunosuppression has been reported in a limited number of cases.5,6

Case Report

- A 65-HD immunosuppressed Asian male with a medical history significant for polycystic kidney disease, with two subsequent renal transplants, presented complaining of a wart-like growth on his right nipple. (Figure A)
- The lesion had been present for approximately six months, and had been bleeding and pruritic as well as significantly grown.
- An attempt was made to treat the lesion at home with OTC wart liquid remover, without resolution.
- PMH was remarkable for HTN, and PCKD. Surgical history included renal transplant x 2 in addition to a removal of benign bladder growth and oral pharyngeal lesion.
- Chronic immunosuppressive treatment for approximately 10 years, with a regimen of tacrolimus, mycophenolic acid, prednisone, and recently, IVIG.
- Past exposure to routine x-rays, CT scans, and dental x-rays.
- Denial history of skin or any other cancer. Family history significant only for two daughters with PCKD and a sister deceased from ovarian cancer.
- The skin exam revealed a 3 cm friable, erythematous and pustular, well-circumscribed mass on the right areola, clinically concerning for SCC versus pyogenic granuloma.
- A shave biopsy of the entire demonstrated lobules of enlarged atypical sebaceous tissue emanating from the dermoepidermal junction and transected broadly at the deep surgical edge.
- Multiple mitotic figures, including atypical radial forms were apparent. (Figure B, C) Immunostains were positive for adipophilin. Given the large number of mitotic figures and apparent size of the process, there was reasonable concern for a superficial sampling of SC.
- Upon referral to general surgery, it was noted that the biopsy performed 2 weeks earlier which had amputated the entire growth on the surface had since returned, growing even larger than it was originally. (Figure D) Clearly, clinically it was behaving as an aggressive, rapidly growing process.
- The lesion was excised via a right mastectomy with radical dissection and sentinel lymph node (SLN) staging with lymph node biopsy x4.
- All four of the right SLNs of the axilla were negative for metastatic SC by morphology, together with EMA and pan-cytokeratin immunohistochemistry (IHC) exams to rule out Muir-Torre Syndrome.
- Therefore, the staging was without evidence of stage III metastasis. (Figure E)
- The patient was then referred to Oncology even though there is neither literature clearly supporting a clear-cut role for adjuvant radiation or chemotherapy, nor are there randomised prospective trials to guide the treatment.
- Given the patient's chronic immunosuppression and kidney disease, and the potential additional risk with any therapy, it was determined that our patient would forego adjuvant therapy.
- The patient then began additional testing and follow-up for continued surveillance of recurrence or metastases.
- He is currently one year post-op, and is in remission in regards to his renal transplant rejection, with no metastases or new lesions found on subsequent follow-up visits.

Clinical and Histologic Findings

- 1. Lesion of right areola or initial presentation prior to shave biopsy.
- 2. Right areola. Multiple mitotic figures including atypical radial forms and clear cell differentiation.
- 3. Right areola biopsy. Multiple mitotic figures including atypical radial forms and clear cell differentiation.
- 4. Lesion of right nipple on initial presentation prior to shave biopsy.
- 5. Right nipple biopsy. Five-generations photo prior to surgical excision.

Muir-Torre Syndrome (MTS)

- A rare syndrome known to arise in association with internal malignancy.
- MTS is characterized by sebaceous carcinoma, particularly in the inner canthus, colon associated adenocarcinoma, cutaneous keratoacanthomas, and parenchymal organ-associated granulomatosis. Skin ulcers require one or more sebaceous neoplasms in association with one internal malignancy.
- colon, renal, endometrial, ovarian, and breast cancers have been reported in 30-40%.
- The patient may have a personal history of a family history of a first-degree relative with MTS.
- Our patient demonstrated: the use of immunohistochemistry(IHC) with adipophilin, focality, and immunofluorescence with PCKD as well as chromosomal analysis of regions 8q, to determine MTS.
- The results of the IHC and MSI indicate whether the patient likely has a sporadic tumor rather than an inherited syndrome-associated tumor like MTS.

Novel Follow-Up Protocol for Our Case

- Clinical follow-up every 6 months, PET/CT scan to determine if there is any evidence of metastatic disease.
- General Surgery follow-up every 6 months
- Dermatology follow-up every 3 months for full body exam

Conclusion

- Extracutaneous SC is a rare entity. Given the general behavior of ocular SC, it is not clear as of yet if extracutaneous SC behaves similarly and requires the same treatment protocol and surveillance. It is important for dermatologists, dermatopathologists, and pathologists to have a low threshold for identifying these neoplasms, especially in immunosuppressed RTR patient populations. More research must be conducted to better determine the risk factors and treatment options for patients in this population, as well as for patients with extracutaneous SC in general, since there are virtually no treatment protocols to follow. To date, we must draw on what we currently know about SC overall until more is known.

References