CLINICAL VIGNETTE

Young Man with Periareolar Skin Nodule Positive for Breast Cancer

Dorcas Chi, MD

Case Report

A 45-year-old man presented with a raised erythematous left periareolar skin nodule present for at least five years. He recalls similar lesion in the right inferior breast which was excised twenty years prior in the Philippines but apparently not sent for pathology review. He noticed the left skin nodule growing bigger over the past year but denied any pain or bleeding. The nodule was dark red and pea sized. Skin punch biopsy revealed adenocarcinoma infiltrating into deep dermis and subcutaneous fat. Differential diagnosis included primary eccrine tumor, adnexal carcinoma or metastatic disease. Tumor cells were very close to a nerve bundle but without definite perineural invasion. Tumor was 100% ER/PR positive, HER2 negative, and with low Ki67 index. He underwent left chest wide local excision including elliptical skin excision and subcutaneous fatty tissue measuring 5 x 3 x 4 cm. No discrete solid mass lesion was grossly evident. Final pathology revealed only focal minute residual microscopic tumor adjacent to previous surgical scar. The surgical margins were free of tumor with closest margin 2mm inferiorly. Staging PET/CT scan showed only localized postsurgical changes in the left breast. There was no FDG avid lymphadenopathy in the axillary or distant sites. Expanded immunostaining studies supported diagnosis of primary breast cancer over skin eccrine carcinoma or neuroendocrine tumor. Bilateral diagnostic mammogram showed only benign appearing postsurgical changes in the left lumpectomy site. Breast MRI showed post-surgical changes with no suspicious subareolar enhancement. Myriad MyRisk gene panel was negative for any deleterious mutation. He elected to proceed with left total mastectomy and sentinel node biopsy months later. Final pathology showed marked fibrosis and biopsy site reaction. There were isolated tumor cells in the sentinel node. His final staging was pT1aN0(i+). Oncotype Recurrence Score was 5. Repeat PET scan was negative and he proceeded with tamoxifen treatment.

Discussion

The appearance of cutaneous metastases from breast carcinoma is variable and can present as nodules or inflammation of the skin, mimicking benign skin conditions. Occult breast cancer (OBC) was first described in 1907 as "cancerous axillary glands with non-demonstrable cancer of the mamma".[1] OBC is defined as a clinically recognizable metastatic carcinoma from an undetectable primary breast tumor, accounts for 0.3–1% of all breast cancers, and can involve lymph node, bone, and skin.[2] It is thought that OBC is secondary to micro-invasive breast cancer.[3]

This is a rare case of a healthy young male with isolated periareolar skin nodule containing metastatic breast cancer. The primary breast cancer remained clinically and radiologically occult despite staging PET scan and mastectomy. Histological and immunohistochemical findings supported its origin as breast primary. This represents CUP (cancer of unknown primary) from occult breast cancer. A multimodality approach was taken and he received surgery and remains on antiestrogen endocrine treatment. The isolated tumor cells (ITCs) in his sentinel node are not defined as a true pathological positive node based on current AJCC TNM classification. However, the clinical importance of ITCs has been debated as some argue its having true metastatic potential rather than just artifacts from benign transportation after tumor manipulation.[4] This is an interesting and yet concerning finding as there is no evidence of primary tumor in his mastectomy specimen. Given his Oncotype score was low, he did not get systemic chemotherapy and remains on tamoxifen treatment. He is now 18 months from his diagnosis and remains with no evidence of disease progression.

As diagnostic techniques continue to improve, the incidence of occult breast cancer has decreased and is rare. Patients with such presentation should have a complete physical examination, mammography, ultrasound, and MRI of the breasts. There is a lack of consensus for the treatment of cancer of unknown primary syndrome from occult breast cancer. After multidisciplinary team evaluation, our patient underwent multimodality treatment including mastectomy and antiestrogen therapy. The prognosis for this complex oncological entity remains guarded although data does support long term disease control.[5]

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