CLINICAL VIGNETTE

Carcinoma en cuirasse

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Case Report

A 70-year-old woman with history of triple-negative breast cancer presented with an extensive rash on the chest. Eight years prior, the patient developed intralobular carcinoma of the left breast treated with lumpectomy and radiation. Five years later, her breast cancer recurred, and was refractory to bilateral mastectomy and multiple chemotherapy agents. Throughout this time she developed an enlarging, indurated rash circumferentially across her chest. The rash was both painful and pruritic.

On exam, along her double mastectomy scar, there were large confluent hyperpigmented brown/black plaques with areas of necrosis. The lesions were rock-hard on palpation.

A punch biopsy was done, which showed metastatic breast adenocarcinoma. Tumor was positive for cytokeratin 7 and GATA3, negative for estrogen receptor and progesterone receptor. Of note, at the time of punch biopsy, lidocaine was unable to be infiltrated due to rock-hard induration of lesions. The punch biopsy was performed without lidocaine, and the patient felt no pain as likely the local nerves were damaged from infiltrating tumor.

CT of the chest, abdomen, and pelvis showed extensive local and regional chest wall cancer recurrence, axillary adenopathy, pleural carcinomatosis and pleural effusion. There was also peripheral encasement involving the left lung lobe predominantly. Adnexal and distal pelvic floor carcinomatosis was seen resulting in obstruction of the distal right ureter and marked hydronephrosis, and large malignant ascites.

The patient was enrolled in a clinical trial with pembrolizumab and imprime by hematology/oncology. Despite this, serial scans showed significant disease progression. Over the next five months, the patient decompensated with recurrent malignant ascites, pleural effusions, and renal failure that ultimately lead to her death.

Discussion

Cutaneous metastases occur in 1-10% of patients with cancer, though incidence varies widely depending on the type of cancer.[1] Breast cancer is the most common type in women to metastasize to the skin. Breast cancer also has the most variable expression of cutaneous metastases, including papulonodular lesions, erysipeloid inflammatory plaques, alopecia neoplastica, and carcinoma en cuirasse.[2]

Carcinoma en cuirasse (CEC) is a rare type of aggressive cutaneous cancer metastasis, most commonly seen with breast cancer but also seen in primary lung, gastrointestinal, and genitourinary cancers. In breast cancers that metastasize to the skin, CEC makes up about 3 to 6% of cases.[3] Initially CEC presents as erythema and pitting edema of the skin. Over time, the lesions gradually become firm and indurated.[4] CEC was first described in 1838 and was aptly named "en cuirasse" which means "breastplate armor," because CEC results in woody induration of skin that mimics armor.[5] The pathogenesis of CEC is unknown. Pleiotrophin, an extracellular signaling molecule, has been hypothesized to act as a tumor promoter by inducing angiogenesis and activating stromal fibroblasts, resulting in sclerotic carcinoma.[6]

Differential diagnosis for CEC include morphea, chronic radiation dermatitis, and keloids. Histology is helpful in distinguishing between these entities. In morphea, histology shows thickened hyalinized collagen bundles and loss of adventitial fat. In chronic radiation dermatitis, collagen is swollen and hyalinized, as in morphea, however there are also large stellate radiation fibroblasts. Keloids display whorls of thickened collagen and fibroblasts, as well as pink hyalinized collagen that resemble “bubble gum.” In CEC, histology shows fibrotic stroma and tumor cells lining up in single rows within collagen bundles. The malignant cells appear similar to fibroblasts, and as a result, may be overlooked.

CEC portends a terrible prognosis given that CEC itself represents an enormous tumor burden and thus late stage cancer. Furthermore the fibrotic nature of CEC increases resistance to chemotherapy. Treatment is aimed at treating the underlying cancer and providing symptomatic relief. Topical anti-pruritic agents, topical steroids, intralesional chemotherapy, and radiation have been used with limited and varying success.[7] In one report, VRCTC-310, a combination of snake venoms crotoxin and cardiotoxin, was used in a woman with CEC, and achieved >80% reduction in tumor mass after 6 weeks of treatment.[8] In another study with twelve patients with cutaneous metastasis of breast cancer, bleomycin was administered followed by brief electric pulses to tumor nodules, and achieved 75% complete response.[9]

In summary CEC is a rare, distinct type of cutaneous cancer metastasis. It is important for internists to recognize this condition and promptly refer patients to Dermatology for evaluation and biopsy. Diagnosis of CEC can provide valuable information in the staging and prognosis of cancer.

Figures

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