

Carcinoma en cuirasse

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ABSTRACT

Carcinoma en cuirasse (CeC) is an extremely rare form of cutaneous metastasis of breast cancer. Guidelines for diagnosis and treatment of CeC are limited due to the small number of reported cases. It can be difficult to distinguish CeC from benign etiologies on initial presentation, but CeC can be easily distinguished by histopathology. Treatment of CeC focuses on palliation with no consensus on therapy guidelines. Treatment modalities that have been explored include chemotherapy, radiotherapy (with or without local hyperthermia), and hormonal antagonists. Here we present a 62-year-old woman with recurrent triple-negative breast cancer manifesting as CeC to the chest wall.

KEYWORDS Breast cancer; carcinoma en cuirasse; cutaneous metastasis

Carcinoma en cuirasse (CeC) is a rare form of breast cancer metastasis, with an incidence of approximately 6%.¹ It tends to appear after initial treatment,² and skin metastases are often associated with breast carcinomas in women.³ CeC first appears as nodules that coalesce, forming diffuse sclerodermoid indurations invading the chest wall and abdomen.^{4,5} Anatomist Alfred Velpeau used the term *cuirasse*, because he thought that the clinical presentation resembled a medieval breastplate or a *cuirass*.⁶ Due to the small number of documented cases, data on this disease are limited. This makes distinguishing CeC from a rash or other forms of cutaneous metastases difficult. Here we report a case of a patient who developed CeC and discuss her progression on treatment.

CASE REPORT

A 62-year-old black woman presented in August 2015 with a mass in the right breast that had been growing for a month. She was nulliparous with a 30-pack-year smoking history and a family history of breast cancer, including a niece with a *BRCA1* gene mutation. A diagnosis of grade III triple-negative infiltrating ductal carcinoma was made. She received several cycles of dose-dense chemotherapy followed

by bilateral mastectomy and lymph node dissection; 10 of 13 lymph nodes had disease present. Upon completion of radio- and chemotherapy, she began breast reconstruction 6 months postmastectomy. In September 2016, she developed a pruritic, erythematous, nodular rash bilaterally over her chest wall. A punch biopsy showed a 7-mm focus of high-grade invasive ductal carcinoma in the dermis similar to the prior tumor. The current tumor had large, pleomorphic, atypical cells with increased nuclear-to-cytoplasmic ratios; large prominent nucleoli; open chromatin; cytoplasmic vacuoles; and scattered karyorrhectic debris, with many mitotic figures, like the original tumor (*Figure 1a*). Bone and computed tomography scans of the chest, abdomen, and pelvis showed no distant disease, only disease in the chest wall and lymph nodes. She received palliative treatment with carboplatin and gemcitabine, but her cancer progressed and became CeC. The chest wall and abdomen were diffusely indurated with firm nodules coalescing into diffuse sclerodermoid plaques (*Figure 1b*). They were hyperpigmented in a background of erythema (*Figure 1b*). After poor response to the first palliative treatment, she began another regimen with eribulin, which softened her hard nodules. Progress stagnated after five treatment cycles. She switched to liposomal doxorubicin but the cancer progressed. She lived 20 months with CeC.

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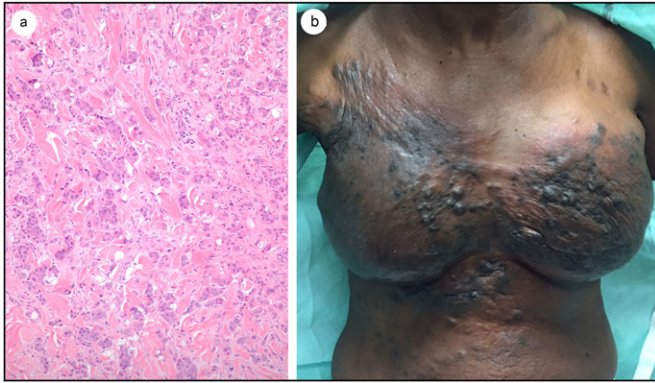


Figure 1. (a) Hematoxylin and eosin–stained section of the tumor in the reticular dermis of the skin ($\times 100$ magnification). (b) Clinical appearance of recurrence of triple-negative breast cancer patient. There are cutaneous metastases to the skin overlying the breast skin, with extensive purple to black nodularity with diffuse sclerodermoid indurations.

DISCUSSION

Breast cancer is not only the most common malignancy in women⁷ but is the primary tumor in about 70% of cutaneous metastases.^{8,9} Cutaneous disease may be the first presentation, because widespread internal metastases can accompany cutaneous disease, resulting in a poor prognosis of only months.¹⁰ This differs with cutaneous metastases in breast cancer patients, because median survival was 13.8 months with a 3.1% 10-year survival rate.¹¹ Despite the extensive cutaneous involvement, our patient had only local disease but died 20 months after her rash developed.

CeC comprises 3% to 6% of cutaneous metastases in breast cancer patients.¹² CeC presents clinically in two stages. The first is erythema and pitting edema of the skin, often confused with a benign dermatitis. Next is formation of thick leathery skin that hardens.⁵ Sclerodermoid chest wall metastases like CeC have a distinctive histologic appearance with fibrotic stroma formed secondary to tumor deposition.³ Eventually, this fibrosis overtakes the area, leaving few identifiable tumor cells. Tumor cells present are often seen in an “Indian-filing” pattern with cells lined up in single rows within collagen bundles of the dermis.¹³ The presence of fibrosis makes it difficult for chemotherapy to penetrate and be effective.¹⁴

The differential diagnosis for patients with CeC includes radiation-induced morphea, inflammatory breast cancer, radiation dermatitis, and other cutaneous metastases. These can be discerned by histology, which often resembles the primary cancer. Unlike radiation dermatitis and inflammatory breast cancer, after the original stage CeC is not associated with inflammatory changes.¹⁵ Morphea and postirradiation morphea grossly resemble CeC, but sclerosis in morphea is characterized by thickened collagen bundles, decreased fat and vascularity, and atrophic eccrine glands.¹⁶ This is distinct from the Indian-filing pattern and fibrosis in CeC. To assess disease metastasis, imaging modalities can be used, such as computed tomography and bone scans, as well as positron emission tomography/computed tomography.¹⁷

There is no consensus on treatment for CeC due to the small number of cases, but modalities used include chemotherapy, snake venom, radiotherapy, hyperthermia, and hormonal antagonists.^{2,18–20} One patient with primary CeC improved with only tamoxifen therapy, suggesting that hormonal therapy may be sufficient to control estrogen receptor–positive disease.² After treatment with eribulin, our patient had softening of her nodules, which appears to be the first documented case in CeC. Yet, treatment remains focused on symptom palliation and improving quality of life, because it is never curative.

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Avocations



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