



Carcinoma en cuirasse caused by pleomorphic lobular carcinoma of the breast in a man

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INTRODUCTION

Breast carcinoma, the most common malignancy in women, is rarely diagnosed in men. Approximately 1,910 new cases of breast carcinoma in men are reported annually¹ compared with more than 230,000 new cases in women.² Most male cases (90%) are of the invasive ductal subtype.³ The lifetime risk of breast cancer for men in the general population is 0.1%.¹ Genetic predisposition significantly heightens this risk, with mutations in *BRCA2* increasing the lifetime risk to an estimated 5% to 10%.¹ Inactivation of other tumor suppressors, including *BRCA1*, *PTEN*, *p53*, and *CHEK2*, is also found to confer an increased risk in men.^{1,3} Congenital disorders such as Klinefelter's syndrome contribute to at least 3% of the incidence of all male breast cancer.¹

We report a 77-year-old man presenting with a painful right axillary mass diagnosed as metastatic carcinoma of unknown primary. Despite multiple treatment modalities, his disease progressed to the brain, lungs, and skin as carcinoma en cuirasse. Histopathologic diagnosis at autopsy was pleomorphic lobular carcinoma (PLC), an uncommon subtype of breast cancer that is reported rarely in men.

CASE REPORT

A 77-year-old man with a 50-pack-year smoking history presented to our institution with an enlarging, painful, right axillary mass. Fine-needle aspiration and excisional biopsy were consistent

Abbreviations used:

CTH2:	caudal type homeobox gene 2
ER:	estrogen receptor
PLC:	pleomorphic lobular carcinoma
PR:	progesterone receptor
PSA:	prostate-specific antigen
TTF1:	thyroid transcription factor 1



Fig 1. CT scan of the chest shows right axillary and subpectoral adenopathy.

with metastatic carcinoma of unknown primary. Histology testing found infiltrates of tumor nodules with large, pleomorphic, poorly differentiated epithelial cells extending into the surrounding fibrofatty tissue and lymphatics. Immunohistochemical stains were positive for cytokeratin 7 and epithelial-membrane antigen but negative for lymphocytic markers (CD3, CD20, CD45), caudal type homeobox gene 2 (CTH2), prostate-specific antigen (PSA), progesterone receptor (PR), estrogen receptor (ER), and

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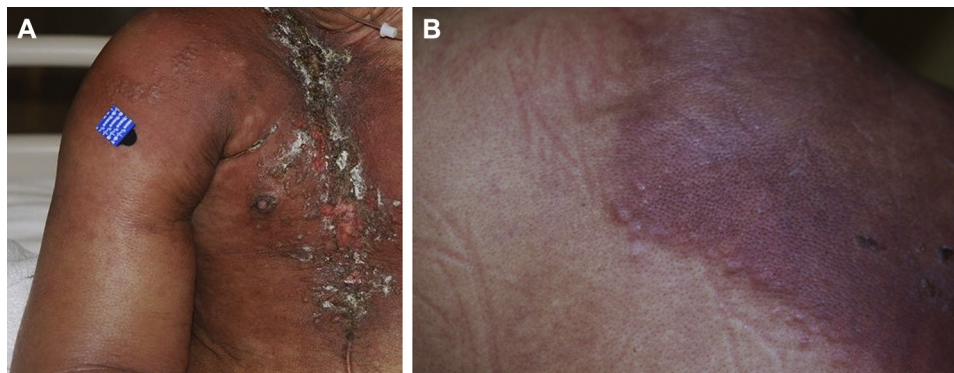


Fig 2. Carcinoma en cuirasse secondary to pleomorphic lobular carcinoma of the breast. **A**, The anterior chest wall, shoulder and right neck were encased in an erythematous, sclerotic plaque, composed of approximately 0.5- to 1.0-cm firm papules, sparing the nipples. **B**, The affected area also demonstrated a peau d'orange appearance with papulonodular margins.

thyroid transcription factor 1 (TTF1). Computed tomography (CT) scan of the chest found right axillary and subpectoral adenopathy (Fig 1). Positron emission tomography/CT scan found fluorodeoxyglucose uptake in the right neck and chest consistent with a neoplastic process.

The patient received multiple treatment modalities, complicated by intermittent periods of poor follow-up. Therapy included 5 cycles of carboplatin and paclitaxel, 1 cycle of docetaxel, and palliative radiotherapy. Fourteen months after initiating treatment, a workup for dyspnea found malignant effusion and brain metastases.

The patient was referred to hospice care; however, 3 months later, he presented with worsening dyspnea. At this time, the right anterior chest wall and neck were encased in an erythematous, sclerotic plaque, composed of approximately 0.5- to 1.0-cm firm papules, sparing the nipples (Fig 2, A). The affected skin showed a peau d'orange appearance with peripheral papulonodules and pseudovesicular papules (Fig 2, B). The ipsilateral arm was exquisitely tender with 4 + pitting edema. CT scan found complete collapse of the right lung with massive pleural effusion; irregular, nodular, soft-tissue densities were seen on the right anterior chest wall, suspected to be the primary source. On brain magnetic resonance imaging, metastases had enlarged and several new enhancing lesions were noted. Skin biopsy found epidermal spongiosis with interstitial fibrosis and thickened collagen bundles in the dermis. Atypical cells infiltrating in a single file pattern and abundant mucin deposition were found (Fig 3). Immunohistochemistry was positive for cytokeratin 7, and negative for cytokeratin 20, TTF1, PSA, and CTH2, consistent with carcinoma. Shortly thereafter, he died of a septic episode.

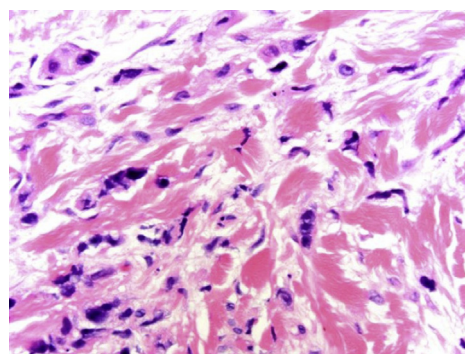


Fig 3. High-power view shows atypical cells infiltrating in a single-file pattern and abundant mucin deposition. (Hematoxylin-eosin stain.)

At autopsy, the pericardial sac and pleura were studded with white, firm nodules, and the skin showed metastatic carcinoma. Immunohistochemistry was positive for pan cytokeratin, gross cystic disease fluid protein 15, and mammaglobin and negative for E-cadherin, ER, PR, and *HER2/neu*. The final pathologic diagnosis was pleomorphic lobular carcinoma of the breast, apocrine subtype, with metastases to skin, brain, lungs, pleura, pericardium, diaphragm, chest wall muscle, and lymph nodes.

DISCUSSION

Invasive lobular carcinoma of the male breast is rare, owing to the absence of lobules and acini in male breast tissue, representing only 1% of male breast cancer.³ PLC is an uncommon highly aggressive subtype, with few reported cases in men.⁴ PLC often presents at an advanced stage, is likely to be larger with more positive lymph nodes at time of diagnosis, and carries an increased risk of lymphovascular invasion and distant metastases.⁵ Cytologically, it is

characterized by enlarged nuclei, prominent nucleoli, increased hyperchromasia, frequent mitotic figures, and eosinophilic cytoplasm.^{5,6} Histologically, PLC shows loss of E-cadherin expression and no expression of ER and PR and has a high rate of *HER2/neu* gene amplification.⁶ However, *HER2* gene amplification appears to be variable in male PLC cases.⁴ In this patient, the apocrine subtype was suspected, associated with significantly more genomic alterations than the nonapocrine type.⁶

Breast cancer is a common malignancy to metastasize to the skin. Carcinoma en cuirasse, a distinct clinicopathologic type of cutaneous metastasis, was coined by Velpeau in 1838 to describe a thick, leathery breast plate of cancer involving the chest and abdominal wall.⁷ Primary carcinoma en cuirasse is rare, presenting as extensive skin involvement in the setting of untreated malignancy; secondary disease is more common, presenting as a local chest wall recurrence after mastectomy, radiation, or chemotherapy.⁸ In this case, carcinoma en cuirasse was caused by occult breast cancer.

The pathogenesis of carcinoma en cuirasse remains largely unknown. It is speculated that pleiotrophin, an extracellular signaling molecule, acts as a multifunctional tumor promoter leading to rapid tumor growth and progression to the scirrhous subtype of invasive carcinoma.⁹ Clinically, this carcinoma begins as scattered, firm, papulonodules overlying an erythematous skin surface progressing to brawny hardness, which eventually coalesces into a sclerodermoid plaque.^{7,8} Histologically, there is extensive dermal fibrosis with single filing of atypical malignant cells; tumor cells may be overlooked because of similarity in appearance to fibroblasts.¹⁰ Treatment options are limited. Because the tumor cells cause extensive reactive fibrosis and decreased

vascularity, chemotherapeutic agents may be unable to obtain tumoricidal concentrations locally.^{8,10} The goal is palliative; local irradiation, skin graft, and nonsteroidal anti-inflammatory drug therapy have shown some success.

To our knowledge, this is the first reported case of carcinoma en cuirasse caused by male breast cancer, and the fifth reported case of PLC in a man. Our case serves to highlight the rapid progression of this rare malignancy and the challenges associated with early detection, diagnosis, and treatment.

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