Inflammatory breast carcinoma (carcinoma erysipeloides): an easily overlooked diagnosis

L.J.FINKEL AND C.E.M.GRIFFITHS

Department of Dermatology, University of Michigan Medical Center, Ann Arbor, Michigan 48109-0314, U.S.A. Accepted for publication 2 March 1993

Summary

A 70-year-old woman developed erythema and induration of the right chest wall, and swelling of her right arm. The provisional diagnosis was deep venous thrombosis and/or cellulitis of the right arm. Skin biopsy showed a poorly differentiated adenocarcinoma within lymphatic vessels, and immunohistochemical staining revealed this to be of breast origin.

Inflammatory carcinoma or carcinoma erysipeloides represents <1% of all cases of breast carcinoma. Our case illustrates the importance of considering this entity in the differential diagnosis of unilateral chest wall erythema and induration.

Case report

A 70-year-old white woman developed worsening cough and dyspnoea, for which she was treated with oral geocillin and organidin, with modest improvement. Two months later she developed right-sided pleuritic chest pain, and was noted to have swelling of her right arm, with erythema and induration of her right breast (Fig. 1). She was admitted to hospital, with the provisional diagnoses of deep venous thrombosis of the right arm, cellulitis, and possible respiratory tract infection.

Physical examination revealed an elderly woman in mild respiratory distress, with diffuse rales over the right lung field on auscultation. The skin of the right breast was diffusely erythematous and indurated, but there was no palpable right axillary lymphadenopathy. The right arm was oedematous, but neither erythematous nor indurated. A right lower lobe effusion and interstitial infiltrate were visible on chest X-ray. Diagnoses of pneumonia and cellulitis were made, and intravenous antibiotic therapy begun. After three different antibiotic regimens were unsuccessful, other diagnostic entities were considered. Bronchoscopy was performed to evaluate the right pulmonary infiltrate, and a dermatology opinion about the right chest wall rash was sought.

Investigations

Histology of skin and transbronchial biopsies revealed poorly differentiated adenocarcinoma. Skin biopsy demonstrated lymphangitic spread (Fig. 2), and immunohistochemical staining with GCDFP-15 monoclonal antibody confirmed the breast origin of the tumour (Fig. 3). Computerized tomography of the chest confirmed lymphangitic spread of the tumour. A mammogram did not show any evidence of a mass, and oestrogen and progesterone receptor assays of the tissue were negative. Carcinoembryonic antigen level was raised at 56.4 ng/ml (normal level ≤ 5 ng/ml).

The patient was seen by the oncology service and a diagnosis of inflammatory breast carcinoma with lymphangitic spread to the lung was made. She was subsequently treated with combination chemotherapy consisting of cyclophosphamide, 5-fluorouracil and adriamycin.



Figure 1. Erythema and induration of the right breast; biopsy site is present medially.

Correspondence: Dr C.E.M.Griffiths.

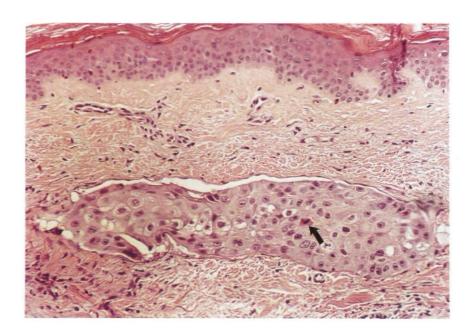


Figure 2. Poorly differentiated adenocarcinoma within dermal lymphatic vessel. Note tumour cell necrosis (arrow) (haematoxylin and eosin).

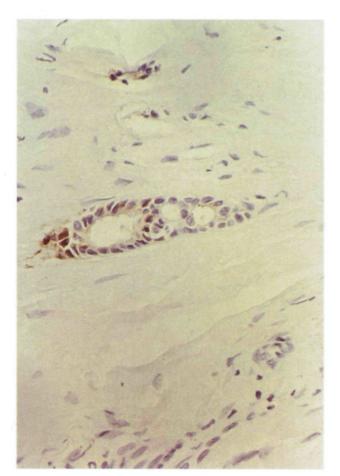


Figure 3. Positive (brown) immunohistochemical staining of tumour with GCDFP-15 antibody, indicative of breast origin.

Discussion

Carcinoma erysipeloides was first described in the early twentieth century, and an excellent review of 28 cases was provided by Lee and Tannenbaum in 1924.1 More recently, this condition has been termed inflammatory carcinoma of the breast. It classically presents as rapidly evolving unilateral chest wall erythema, which may extend to the back, proximal part of the arm, and even across the midline. The inflamed area may show a distinct raised periphery, suggestive of erysipelas, and oedema secondary to lymphatic obstruction may be prominent. Taylor and Metzer² have classified these lesions as primary or secondary. In primary inflammatory carcinoma, signs of inflammation develop simultaneously with carcinoma in a previously normal breast. Although oedema and induration may decrease the sensitivity of mammography, we are left to conclude that our patient's inflammatory carcinoma was primary. Secondary inflammatory carcinoma denotes abrupt inflammation in a breast known to harbour neoplasia. The primary neoplasia may not be in the underlying breast tissue, and inflammatory carcinoma has been reported in association with melanoma, and lung, pelvic, and pancreatic tumours.³⁻⁵ The term 'carcinoma telangiectatica' refers to the presence of purpuric plaques, papules and vesicles, and this should be easy to distinguish from inflammatory carcinoma.

Histology revealed plugging of superficial and deep lymphatics by poorly differentiated adenocarcinoma; the tumour cells contained large, pleomorphic, hyperchromatic nuclei. Deep lymphatic invasion implies an especially poor prognosis. In carcinoma telangiectatica only the more superficial lymphatics are involved.

Immunohistochemistry may prove invaluable, as it did in this case, in establishing the tissue of origin in inflammatory breast carcinoma. Appropriate further diagnostic tests and therapeutic intervention may follow. In our case, the use of a monoclonal antibody raised against a protein found in the fluid of fibrocystic breast disease (GCDFP-15) determined the tissue of origin. GCDFP-15 is found in benign and malignant breast tissue, and in salivary glands, and is produced by all normal apocrine glands. In a series of 562 primary breast carcinomas, 55% of the carcinomas studied stained positively for GCDFP-15.8 The percentage of carcinomas with positive staining was greatest for those subtypes with apocrine histological features (75%). intraductal carcinoma (70%), and infiltrating lobular carcinoma with signet-ring cell differentiation (90%).8 Positive staining was related to a history of fibrocystic disease, but not to age, parity, menopausal status, risk of recurrence, or survival. GCDFP-15 is undetectable in normal lung or gastrointestinal tract tissue.⁷ In our patient, the breast was not removed, because of the presence of metastatic disease, and the exact histological subtype of the tumour could not be ascertained.

Primary inflammatory carcinoma of the breast represents a diagnostic challenge, and delay in diagnosis is common. Erysipelas and cellulitis are the differential

diagnoses most often considered, and our patient had received prolonged intravenous antibiotic therapy. In these circumstances, a biopsy is indicated. The diagnoses of deep venous thrombosis, cellulitis and respiratory infection were all invoked for our patient, to explain arm oedema, sharply marginated chest wall erythema and dyspnoea. Primary inflammatory carcinoma of the breast with lymphatic obstruction and lymphangitic pulmonary spread provided an explanation for all these features. Clinicians should be aware of the various presentations of inflammatory carcinoma of the breast, as prompt diagnosis and therapy are essential.

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