JAMA Dermatology Clinicopathological Challenge

An Elderly Woman With Violaceous Swelling With Telangiectasia and Necrosis on the Face

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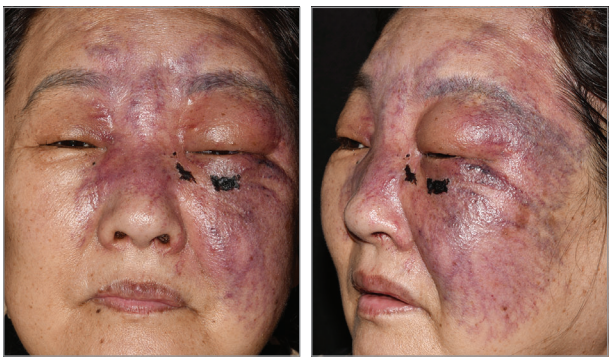

Figure 1. A, A full image and B, a three-quarters image of the face, showing telangiectatic, swelling erythema mainly involving the left side of the forehead, eyelids, nose, and cheek.

A woman in her 60s presented to the clinic with a 3-month history of pain, swelling, and erythema on her face. Eleven years earlier, she had been diagnosed with left breast carcinoma and had undergone a mastectomy with subsequent radiotherapy and chemotherapy. Seven years later, she had been found to have left supraclavicular lymph nodes metastasis and had undergone a surgical excision. On presentation for the current problem, the patient experienced pain on palpation, but denied fever, chills, or other symptoms. A physical examination revealed violaceous swelling with telangiectasia and necrosis on the forehead, eyelids, nose, and cheek, predominantly involving the left side of the face (Figure 1). A biopsy of the lesion was performed for histopathological analysis (**Figure 2**).

WHAT IS YOUR DIAGNOSIS?

- **A.** Cutaneous angiosarcoma
- **B.** Radiation dermatitis
- **C.** Wolf isotopic response
- **D.** Telangiectatic metastatic breast carcinoma
- **E.** Herpes zoster

Diagnosis

D. Telangiectatic metastatic breast carcinoma

Discussion

Histopathological examination revealed hyperkeratosis, epidermal atrophy, and dilated vessels in the dermis. One vessel had nests of highly atypical cells (**Figure 2**A). On immunohistochemical examination, epithelial cells in the dilated vessels were positive for CD31, and the atypical cells were strongly positive for GATA3 (Figure 2B). A diagnosis of telangiectatic metastatic breast carcinoma (TMBC) was made.

Breast carcinoma is the most prevalent cancer and the second most common cause of cancer death in women.[1] Metastatic breast carcinoma most frequently involves the lymph nodes, lung, liver, and bone marrow; cutaneous metastases are relatively uncommon, with an incidence of 2.42%.[2] Clinical manifestations can include nodules (47%-80%), alopecia neoplastica (2%-12%), telangiectatic carcinoma (8%-11%), carcinoma erysipeloides (3%-6%), and melanoma-like metastases (6%).[1,3] Common sites of cutaneous metastases are the chest and abdomen; the face is rarely affected.[4,5] To our knowledge, 2 cases of metastatic breast carcinoma involving the face have been reported, simulating cutaneous angiosarcoma: one with fa-

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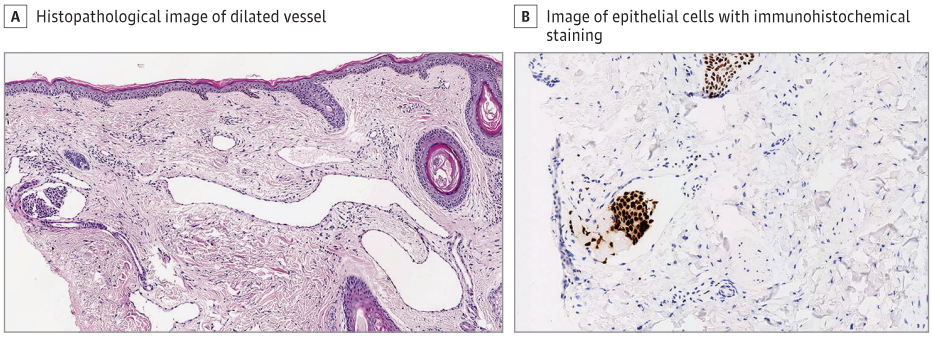

Figure 2. A, Highly atypical cells in a dilated vessel in the dermis (hematoxylin-eosin stain; original magnification, ×10). B, Atypical cells in dilated vessels (streptavidin-peroxidase stain; original magnification, ×20).

cial telangiectasia and erythema[4] and the other with a large red-purplish plaque and stony, hard nodules.[5]

Cases of TMBC have variable clinical appearances and usually present as fleshy to erythematous nodules on the chest; they rarely involve the face. Cutaneous angiosarcoma usually begins as bruise-like patches on the skin, as well as erythematous or violaceous macules, patches, papules, and nodules on the face and scalp of elderly people.[6] Lesions may be solitary or multiple; facial swelling may occur. The clinical difference might be the lesion spreading pattern. Cutaneous angiosarcoma usually spreads centrifugally and involves both sides of the face, whereas metastases from a left breast carcinoma mainly affect the left side of the face, as in this case. Histopathologically, cutaneous angiosarcoma is lined by atypical endothelial cells that exhibit apparent nuclear pleomorphism and high mitotic activity, as opposed to those in TMBC, which typically displays tumor cells within dilated vessels in the superficial dermis that can be seen in cutaneous metastases of other carcinomas. Panels of immunohistochemical markers can help determine primary sites of metastasis, including estrogen, progesterone, cytokeratin 7, GATA binding protein 3, and gross cystic disease fluid protein–15.

Other differential diagnoses include radiation dermatitis, Wolf isotopic response, and herpes zoster. Radiation dermatitis usually exhibits poikilodermatous changes on the skin surface, including erythema, telangiectasia, hyperpigmentation, and hypopigmentation. Since the patient underwent radiotherapy 11 years earlier, this lesion may be confused with chronic radiation dermatitis, which also presents as painful erythema with telangiectasia and necrosis and displays vascular dilation histopathologically.[7] However, other histopathological features of chronic radiation dermatitis did not exist, including pigmentary alteration in the epidermis, dermal fibrosis, irregular shaped fibroblasts, and loss of adnexal structures. It is also important to suspect herpes zoster when a painful, edematous erythema with unilateral dermatomal distribution occurs in an elderly woman who had received immunosuppressive therapy. Histopathologically, herpes zoster is characterized by intraepidermal vesicles, ballooning degeneration of enlarged keratinocytes, and eosinophilic intranuclear inclusion bodies in multinucleated giant cells.[8] Also, Wolf isotopic response is the subsequent occurrence of a new skin condition at the site of another unassociated, previously healed disease. The most common primary disease is herpes zoster, which has been reported to be followed by a variety of conditions.[9] Thus, the cutaneous angiosarcoma–like lesion in zosteriform distribution raised the possibility of Wolf isotopic response. However, the patient did not recall a varicella-zoster infection or any skin condition on her face. The presence of atypical cells in dilated vessels excluded these diagnoses.

The 5-year survival rate is reported[10] to be 38% in patients with cutaneous metastases vs 99% in those with localized breast cancer. Recognition is crucial for early diagnosis and management, and suspicion for metastatic spread should be raised in patients with breast cancer who have telangiectatic facial swelling. Unfortunately, the patient was in poor health and could not undergo or afford further treatment. She was still alive at 5-month follow-up.

ARTICLE INFORMATION

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