JAMA Dermatology Clinicopathological Challenge

Facial Erythema in an Elderly Man

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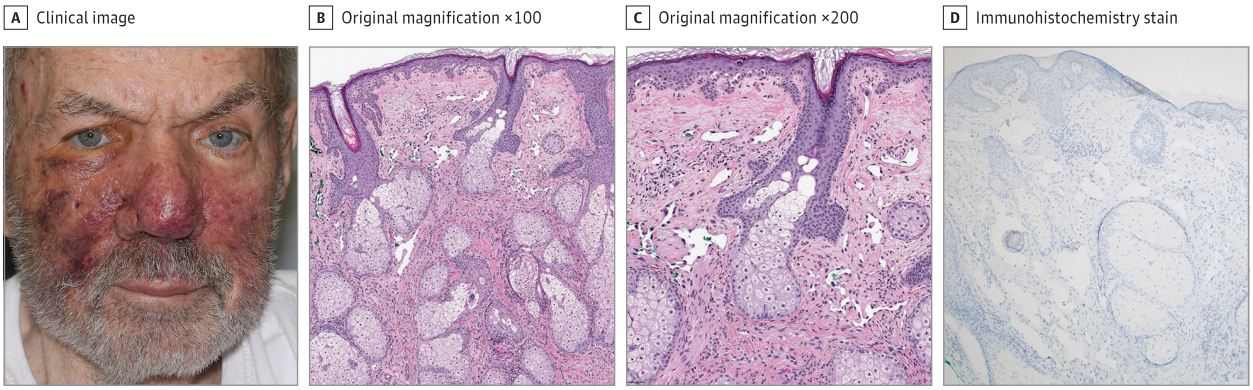

Figure. A, Clinical image showing erythemato-violaceous plaques with ecchymotic areas. B, Histopathologic image showing ectatic vessels with plump endothelia involving the deep dermis and hypodermis (hemotoxylin-eosin). C, On higher magnification, ectatic vascular channels were lined by atypical plump endothelial cells (hemotoxylin-eosin). D, Immunohistochemistry was negative for *Human herpesvirus 8* (original magnification ×100).

A man in his 80s with a pacemaker; a history of congestive heart failure, coronary artery disease, atrial fibrillation, transient ischemic attack, and Parkinson disease; and dependence in all activities of daily living presented to the dermatology department with a 4-month history of new-onset persistent facial eruption. He denied a history of facial flushing. The patient was initially treated for rosacea at an outside hospital with topical 1% metronidazole cream for 1 month without improvement and developed acute facial purpura after 1 day of treatment with oral doxycycline, which was discontinued. Because of the eruption's rapid onset and violaceous appearance, as well as empirical treatment failure, the patient was referred for further evaluation. On examination, the patient had asymmetric, centrofacial, erythematous-violaceous indurated telangiectatic and ecchymotic plaques over a phymatous background (**Figure**, A). A series of punch biopsies were performed (Figure, B-D).

WHAT IS YOUR DIAGNOSIS?

A. Kaposi sarcoma

B. Elder abuse

C. Rosacea-like angiosarcoma

D. Telangiectatic metastatic carcinoma

Diagnosis

C. Rosacea-like angiosarcoma

Discussion

Microscopic examination findings revealed a dissecting vascular proliferation through the entire dermis. Ectatic vascular channels were lined by atypical plump endothelial cells (Figure, B and C). Immunohistochemistry staining results were negative for *Human herpesvirus 8* (Figure, D). These findings were diagnostic of cutaneous angiosarcoma (CA).

Results of a positron emission tomography/computed tomography scan revealed only mildly avid malar subcutaneous thickening consistent with CA (American Joint Committee on Cancer tumor-node-metastasis staging T2aN0M0G2, stage IIB). When feasible, standard treatment entails surgery with or without radiation; however, owing to this patient's comorbidities, poor performance status, and disease extent, reduced-dose intravenous paclitaxel (60 mg/m^2) was initiated as palliative therapy. After completing 6 months of 120-mg paclitaxel weekly treatment, a partial response was obtained. Because of worsening overall health, no additional treatments were added. He was transitioned to home hospice.

Cutaneous angiosarcoma is a rare, aggressive, malignant tumor arising in blood or lymphatic vessels characterized by uncontrolled proliferation of endothelial cells.[1] It typically affects the head and neck areas of older, white, adult men.[2] Cutaneous angiosarcoma typically presents as ecchymosis-like patches or plaques that subsequently expand to form nodules involving larger areas that further ulcerate. Three types of CA have been described: (1) head and neck type (Wilson-Jones angiosarcoma), typically arising on the scalp and termed *idiopathic* with a possible ultraviolet radiation role; (2) postradiotherapy type, typically arising on the breast years after breast cancer treatment; and (3) secondary to chronic lymphedema type or Stewart-Treves syndrome. Rosacea-like angiosarcoma is a rare clinical form of idiopathic CA possibly owing to overlap of concomitant acne rosacea vs a primary rosacea-like CA morphology.

The differential diagnosis may include Kaposi sarcoma (KS), another malignant vascular tumor that can involve the face.[3,4] Kaposi sarcoma most commonly involves the mucosa in the HIV-related type and the distal extremities in the classic type.[4] When it involves the face, KS typically presents as discrete violaceous plaques and nodules.[4] Histologically, KS can resemble CA in its early phases. In later phases, KS presents with spindle cells in the dermis dispersed throughout collagen bundles forming angulated vascular channels.[4] Immunohistochemistry is positive for *Human herpesvirus 8*.

Elder abuse is defined as actions that cause harm or have potential for harm, intentionally or by neglect, or as failure by a caregiver to satisfy the elder's basic needs or to protect from harm.[5] Estimated prevalence in Western countries ranges from 2.2% to 18.4%.[5] Recognition of elder abuse by dermatologists is critical because abuse is associated with increased risk of hospitalization and death.[5,6] Patients can present with bruising, burns, lacerations, traumatic alopecia, external genital trauma, and malnutrition. Traumatic ecchymoses may morphologically resemble this patient's findings. Histopathologic results show noncircumscribed blood extravasation (hemorrhage) and, in later stages, hemosiderin deposition. A high index of suspicion for elder abuse as well as exclusion of other causes is needed.[5]

Cutaneous metastasis can rarely present with a telangiectatic appearance mimicking rosacea-like CA. History of a prior or active cancer can increase the index of suspicion. The present patient had no history of other cancer; however, cutaneous metastasis may be the first clinical sign of an underlying malignant tumor.[7] Dilated blood vessels infiltrated by tumor cells may be seen in histopathological results.[7-10] Metastatic breast cancer can present with this appearance (telangiectatic metastatic breast carcinoma or carcinoma telangiectaticum).[10]

Rosacea-like CA may initially be misdiagnosed as acne rosacea and rhinophymatous rosacea, as in this case, which delays diagnosis and treatment. With the aging worldwide population,[5] dermatologists must maintain a high index of CA suspicion.

ARTICLE INFORMATION

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