CASE REPORT

Grenz zone in a case of post-radiation cutaneous angiosarcoma

Hasbani D. J., MD, Kurban M., MD, Abbas O., MD

Department of Dermatology, American University of Beirut Medical Center, Lebanon

ABSTRACT

Cutaneous angiosarcoma is a well-described but rare complication of breast cancer treatment, which is associated with a poor prognosis. Post-radiation angiosarcoma tends to develop around 5-6 years after radiation and clinically presents as asymptomatic large erythematous plaques, patches, or nodules. The lesions may be multicentric and almost always involve the previous surgical scar. Histologically, angiosarcoma exhibits vasoformative elements always involving the dermis. But, to the best of our knowledge, a grenz zone has not been previously described. In this article, we present the case of a radiation-associated cutaneous angiosarcoma exhibiting a grenz zone.

KEY WORDS: Grenz Zone, Angiosarcoma

INTRODUCTION

Cutaneous angiosarcoma is a well-described but rare complication of breast cancer treatment, which is associated with a poor prognosis. It may develop either on the arm, in the setting of lymphedema after mastectomy and axillary lymph node dissection. Or, over the chest wall, in the area of prior radiation therapy after breast-conserving therapy.[1] Post-radiation angiosarcoma tends to develop around 5-6 years after radiation and clinically presents as asymptomatic large erythematous plaques, patches, or nodules, that may be multicentric and almost always involve the previous surgical scar.[1,2] Histologically, angiosarcoma may lie in a spectrum between well- and poorly-differentiated. It consists of vasoformative elements always involving the dermis,[1] but, to the best of our knowledge, a grenz zone has not been previously described. In this article, we present the case of a radiation-associated cutaneous angiosarcoma exhibiting a grenz zone.

REPORT OF A CASE

A 76-year-old woman presented with a 3-month history of progressive erythematous lesions over her right breast. She had a history of infiltrating lobular carcinoma post-partial mastectomy and radiation treatment of the left and right breasts, 8 and 5 years prior to presentation, respectively. On physical examination, dark red to violaceous well-defined firm plaques, as well as bruise-like cutaneous changes, were observed diffusely over the right breast and extending across the intramammary folds to involve the medial aspect of the left breast (Fig. 1). A punch biopsy was obtained,

Correspondence: *Dr. Ossama Abbas, Professor, Department of Dermatology, American University of Beirut Medical Center, Riad El Solh/ Beirut 1107 2020, Beirut, Lebanon, P.O.Box 11-0236, Email : ossamaabbas2003@yahoo.com, Tel: +961-1-350000, ext. 7915, Fax : +961-1-745320*

and it exhibited irregularly shaped anastomosing vascular channels and sheet-like highly infiltrative poorly demarcated dermal growth of atypical CD31-positive cells with nuclear atypia and scattered mitoses (Fig. 2). Given the clinical picture, these findings were consistent with post-radiation cutaneous angiosarcoma. Interestingly, a prominent grenz zone was noted overlying the dermal proliferation.

Fig. 1 Cutaneous angiosarcoma presenting as firm red to violaceous plaques, as well as ecchymotic areas over the right breast.

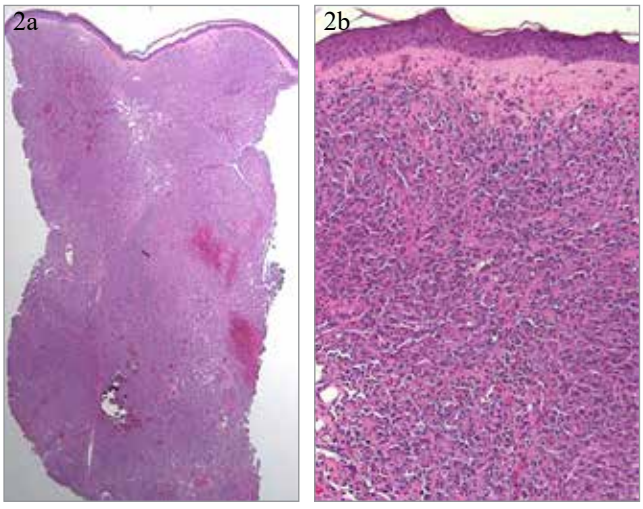

Fig. 2 A&B, Punch biopsy featuring a grenz zone overlying a poorly demarcated dermal sheet-like highly-infiltrative growth of atypical cells with nuclear atypia and scattered mitoses, as well as irregularly shaped anastomosing vascular channels (hematoxylin-eosin x20 and x400, respectively).

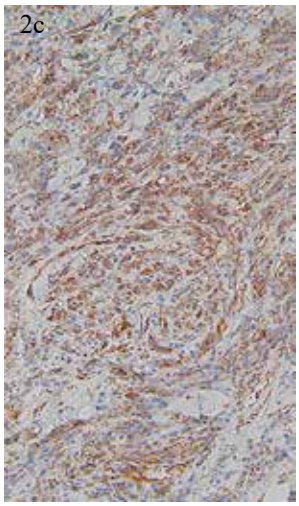

Fig. 2 C, Immunohistochemical staining for CD31 showing positivity in the tumor cells (x400).

DISCUSSION

Grenz zone is a term used in dermatopathology to refer to a narrow uninvolved zone of papillary dermis lying between the epidermis and an inflammatory or neoplastic infiltrate beneath. The observation of a grenz zone is quite particular in nature. It is a defining histologic characteristic of certain inflammatory conditions, namely granuloma faciale and erythema elevatum diutinum, infectious entities like leprosy, and neoplastic ones like dermatofibroma. Nevertheless, it less often embodies an uncommonly encountered feature of select neoplastic entities, such as metastatic melanoma and atypical fibroxanthoma.[3] The underlying pathogenesis for the development of a grenz zone has not been well defined to date, but it has been postulated to be related to the deposition of newly formed collagen at the site of ultraviolet light-induced elastosis in the upper dermis. The B lymphocytes exhibit specific adhesive properties that, under the influence of certain chemokinetic and chemotactic signals, lead to their migration towards and up to this specific zone in the dermis; hence, this may explain the grenz zone observed in B-lymphocyte rich cutaneous infiltrates.[3] However, the potential for grenz zone formation is elusive in

the case of vascular neoplasms. The only other vascular proliferation that has previously been reported to show a grenz zone is acquired elastotic hemangioma.[4]

To our knowledge, no previous observations of a grenz zone occurring in cutaneous angiosarcoma have been reported in the literature to date. This reinforces the non-specific nature of this peculiar histologic feature and allows us to add angiosarcoma to the histologic differential diagnosis of emerging entities exhibiting a grenz zone. Although the underlying etiology is not clear, further investigation into the incidence of this finding should be conducted to better characterize its potential implications.

REFERENCES

1. Brenn T, Fletcher CDM. Radiation-Associated Cutaneous Atypical Vascular Lesions and Angiosarcoma: Clinicopathologic Analysis of 42 Cases. Am J Surg Pathol. 2005; 29(8).
2. Ronen SA-O, Ivan DA-O, Torres-Cabala CA-O, Curry JA-O, Tetzlaff MA-O, Aung PP, et al. Post-radiation vascular lesions of the breast. J Cut Pathol. 2019; 46(1):52-58.
3. Abbas O, Mahalingam M. The grenz zone. Am J Dermatopathol. 2013; 35(1):83-91.
4. Cohen PR, Hinds BR. Acquired elastotic hemangioma: case series and comprehensive literature review. Cureus. 2017; 9(12).