DOI: 10.1002/cia2.12096

LETTER TO THE EDITOR

Journal of Cutaneous Immunology and Allergy
WILEY

Poorly differentiated angiosarcoma with few vascular channels

Dear Editor,

Cutaneous angiosarcoma is a rare skin neoplasm with highly aggressive behavior derived from vascular endothelial cells. Because of its rapid clinical course and limited treatment options, timely and accurate diagnosis is essential in order to achieve the good outcome. Here, we report a rare case of poorly differentiated angiosarcoma with very few vascular channels.

An 84-year-old Japanese woman presented with a 3-month history of a mass on the scalp. Clinical examination revealed a solitary 1.5 × 1.5 cm, hard nodule with telangiectasia but not skin color change on the scalp (Figure 1A). No superficial lymph nodes were swollen. She had a history of breast cancer 8 years before and was treated with breast surgery and adjuvant chemotherapy resulting in complete response. Skin biopsy specimen revealed massive invasion of medium-to-large-sized tumor cells in the dermis and subcutaneous tissue without apparent vascular channels in most area (Figure 1B,C). The tumor cells were round to spindle-shaped with vesicular nuclei (Figure 1C). Some parts of the tumor-contained cells with intracytoplasmic vacuoles and centrally placed or compressed nuclei (Figure 1D). Vascular channels partially with erythrocytes

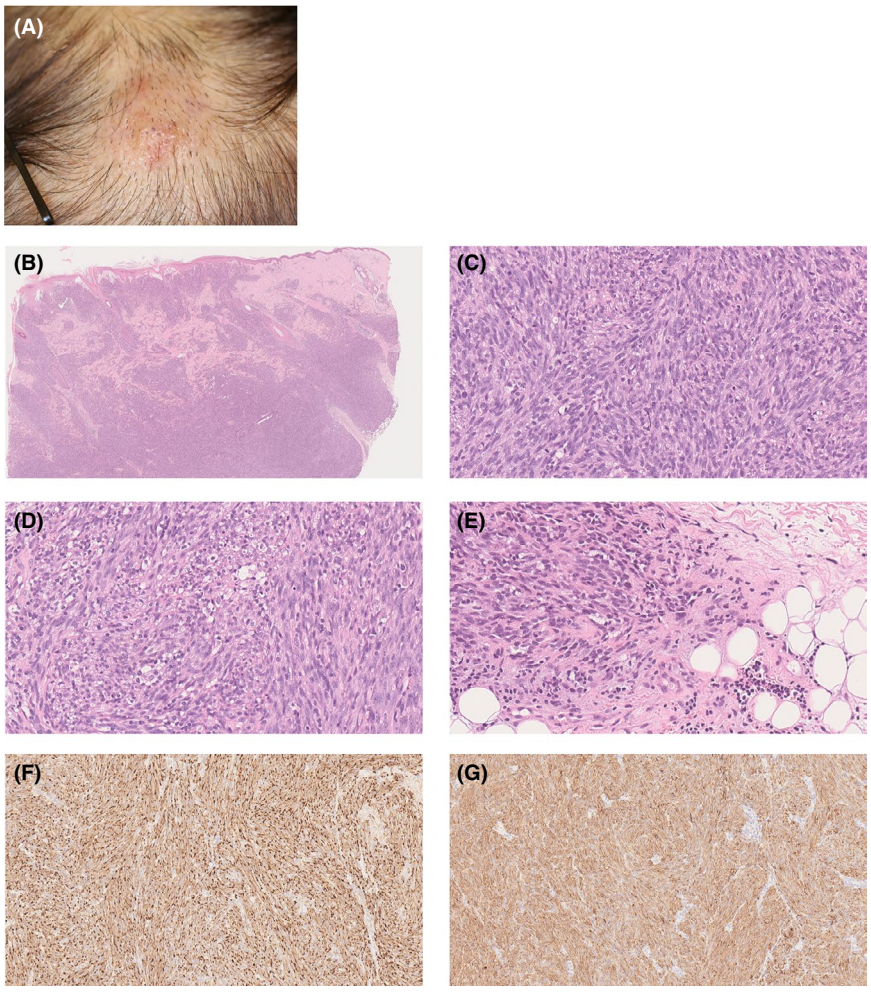

FIGURE 1 A, Clinical presentation shows a solitary 1.5 × 1.5 cm, hard nodule with telangiectasia but not skin color change on the scalp. B, Low-power H&E section shows massive invasion of tumor cells in the dermis and subcutaneous tissue (×25). C, High-power H&E section shows that tumor cells are characterized by medium-to-large size, round to spindle shape, and vesicular nuclei (×400). D, High-power H&E section shows that some parts of tumor cells have intracytoplasmic vacuoles and centrally placed or compressed nuclei (×400). E, High-power H&E section shows vascular channels in very limited area (×400). F, G, Immunohistochemical stain shows that tumor cells are positive for CD31 (F) and D2-40 (G) (F, ×200, G, ×200)

were found in very limited area (Figure 1E). Immunohistochemical studies showed that the tumor cells were positive for CD31, CD34, and D2-40, but were negative for cytokeratin AE1/AE3, epithelial membrane antigen, S-100, synaptophysin, alpha-smooth muscle actin, and human herpesvirus 8 latent nuclear antigen-1 (Figure 1F,G and data not shown). Based on the findings above, we diagnosed her as having poorly differentiated angiosarcoma. She died of unknown cause soon after diagnosis.

The pathologic diagnosis of angiosarcoma is easy when tumor cells form vascular channels. On the other hand, in rare cases of poorly differentiated angiosarcoma with spindle or epithelioid differentiation, the tumor shows solid growth pattern and overt vasoformative elements are hard to recognize.[1-3] In such cases, accurate diagnosis is challenging with a wide differential diagnosis, including carcinoma, melanoma, epithelioid sarcoma, and mesenchymal tumors such as smooth muscle tumors, dermatofibrosarcoma protuberans, and atypical fibroxanthoma. Helpful histological features are the identification of areas of better-differentiated angiosarcoma, although such area was found in very limited area after careful search in previous cases similar to our case.[2,3] In addition, intracytoplasmic vacuoles, indicating the early phase of angiogenesis, can also be clue of poorly differentiated angiosarcoma as shown in our case, while they are not specific for angiosarcoma.[2,3] Finally, immunohistochemical analysis on endothelial cell antigens is needed on the diagnosis of angiosarcoma, particularly poorly differentiated angiosarcoma. In our case, coexpression of vascular endothelial markers, CD31 and CD34, and a lymphatic endothelial marker, D2-40, and lack of differentiation markers other than endothelial markers led us to the diagnosis of angiosarcoma. However, loss of expression of one or more endothelial markers is common in angiosarcoma and endothelial marker expression is also found in a subset of nonvascular malignancies.[4] In addition, cytokeratin and epithelial membrane antigen are also expressed in some cases of epithelioid angiosarcoma.[4,5] Staining for multiple markers indicating endothelial differentiation and various lineage markers is important in diagnosing poorly differentiated angiosarcoma.

CONFLICT OF INTEREST

The authors declare no conflict of interest.

INFORMED CONSENT

Informed consent was obtained from the patient.

DECLARATIONS

Approval of the research protocol: No human participant was involved in this study.

Registry and the Registration No: N/A.

Animal Studies: N/A.

Arisa Kishi MD
Tomomitsu Miyagaki MD, PhD
Takafumi Kadono MD, PhD

Department of Dermatology, St. Marianna University School of Medicine, Kawasaki, Japan

Correspondence
Tomomitsu Miyagaki, MD, PhD, Department of Dermatology, St. Marianna University School of Medicine, 2-16-1 Sugao, Miyamae-ku, Kawasaki, Kanagawa 216-8511, Japan.
Email: asahikari1979@gmail.com

ORCID

Tomomitsu Miyagaki https://orcid.org/0000-0003-1879-3128

REFERENCES

1. Shustef E, Kazlouskaya V, Prieto VG, Ivan D, Aung PP. Cutaneous angiosarcoma: a current update. J Clin Pathol. 2017;70:917–25.
2. Wood A, Mentzel T, van Gorp J, Flucke U, Huschka U, Schneider J, et al. The spectrum of rare morphological variants of cutaneous epithelioid angiosarcoma. Histopathology. 2015;66:856–63.
3. Terada T. Fatal poorly differentiated angiosarcoma of the scalp. Int J Clin Exp Pathol. 2010;3:541–4.
4. Rao P, Lahat G, Arnold C, Gavino AC, Lahat S, Hornick JL, et al. Angiosarcoma: a tissue microarray study with diagnostic implications. Am J Dermatopathol. 2013;35:432–7.
5. Al-Abbadi MA, Almasri NM, Al-Quran S, Wilkinson EJ. Cytokeratin and epithelial membrane antigen expression in angiosarcomas: an immunohistochemical study of 33 cases. Arch Pathol Lab Med. 2007;131:288–92.