SKIN CANCER (OTHER THAN MELANOMA)

LYMPHANGIOSARCOMA AS A DISTINGUISHABLE VARIANT OF ANGIOSARCOMA

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Introduction: An angiosarcoma is defined as a rare and aggressive malignant vascular tumor; its clinical features generally involve purpuric macules and hemorrhagic nodes or ulcerations with prominent extravasation of red cells. However, ill-defined edematous lesions with lymphorrhea and non-hemorrhagic nodes without extravasation of red cells have been observed in rare cases. Such rare cases are considered as “lymphangiosarcomas” because they have more lymphatic characteristics than common angiosarcomas.

Objective: We investigated the clinicopathological differences between lymphangiosarcoma (LAS) and hemangiosarcoma (HAS), which represented common angiosarcoma.

Material and Methods: We conducted a retrospective study of 119 angiosarcoma patients treated at our institution from 1986 to 2017. Twenty of these cases were diagnosed as LAS. Immnohistochemical stains were performed in primary tissue samples for CD31, CD34, D2-40, Prox-1, LYVE-1, and claudin-5. Using the angiosarcoma cell line (ISO-HAS) and lymphangiosarcoma cell line (MO-LAS) that we had established, Prox-1, LYVE-1 and claudin-5 levels were measured by flow cytometry (FC).

Results: On immunohistopathological analysis, 40.4% and 71.9% of LAS and HAS patients, respectively, tested positive for CD34. Except a few, all patients (both LAS and HAS) tested positive for D2-40 and CD31. Prox-1, LYVE-1, and claudin-5 showed diffusely positive expression in 100%, 94%, and 94% of LAS patients and 65.2%, 69.6%, and 56.5% of HAS patients, respectively. Prox-1 and claudin-5 levels were significantly higher in MO-LAS cells than in ISO-HAS cells, and the both cells showed similar levels of LYVE-1, which were measured using FC.

Conclusions: LAS is difficult to diagnose clinically and histopathologically owing to the presence of more lymphatic features and lack of hemorrhagic features; LAS is also very rarely observed, which contributes to the difficulty in diagnosis. Thus, Lymphangiosarcoma should be recognized as a variant of angiosarcoma because it has clear lymphatic features.