LETTER TO THE EDITOR

Case of lymph node primary perivascular epithelioid cell tumor associated with leukoderma

Dear Editor,

Perivascular epithelioid cell tumor, also known as PEComa, is a type of mesenchymal neoplasm.\(^1,2\) Here, we encountered an interesting case of primary PEComa in a cervical lymph node (LN) that was associated with leukoderma.

A 63-year-old Japanese man was referred to us with a 3-month history of a swollen LN in the left neck. The patient was a white collar worker belonging to an advertising agency unassociated with chemical agents. The patient had not used a cosmetic containing rhododendrol. Physical examination, including otological examination, revealed a swollen, movable, non-tender LN in the left neck, and several white macules measuring less than 40 mm in diameter on the forehead and neck (Fig. 1a). The areas of leukoderma had been increasing in number for 30 years. The patient had not been diagnosed with disease of the thyroid gland or anemia. Computed tomography detected a swollen LN measuring 16 mm × 13 mm (Fig. 1b), but no other lesions. Histopathological examination of the resected swollen LN revealed that tumor cells occupied the majority of the LN (Fig. 1c). The tumor consisted of epithelioid

Figure 1. (a) Cutaneous manifestations of leukoderma are evident on the head and neck. (b) Computed tomography of the head and neck revealed a swollen oval lymph node in the left neck (white arrowheads). (c) Tumor cells occupy the majority of the lymph node (hematoxylin–eosin [HE], original magnification ×40). (d) The tumor consists of epithelioid cells with abundant cytoplasm and pleomorphic nuclei with distinct nucleoli and occasional mitoses (HE, ×400). (e) Immunohistochemical staining using anti-Melan-A/MART-1 antibodies. The antibodies are reactive with the tumor cells (×400). (f) Immunohistochemical staining using anti-MUM1 antibodies. The antibodies are not reactive with the tumor cells, but are reactive with some lymphocytes (×400).

Correspondence: Yoshimasa Nobeyama, M.D., Ph.D., Department of Dermatology, The Jikei University School of Medicine, 25-8 Nishi-shimbashi 3-chome, Minato-ku, Tokyo 105-8461, Japan. Email: nobederm@jikei.ac.jp

© 2017 The Authors. The Journal of Dermatology published by John Wiley & Sons Australia, Ltd on behalf of Japanese Dermatological Association. This is an open access article under the terms of the Creative Commons Attribution-NonCommercial License, which permits use, distribution and reproduction in any medium, provided the original work is properly cited and is not used for commercial purposes.
cells with abundant cytoplasm and pleomorphic nuclei containing distinct nucleoli, which occasionally showed mitoses (Fig. 1d). Anti-S100 protein antibodies (Agilent Technologies/DAKO, Santa Clara, CA, USA), anti-HMB-45 antibodies (Agilent Technologies/DAKO) and anti-Melan-A/MART-1 antibodies (Agilent Technologies/DAKO) were all reactive with the tumor cells (Fig. 1e). In contrast, neither anti-MUM1 antibodies (Agilent Technologies/DAKO; Fig. 1f) nor anti-α-smooth muscle actin antibodies (Agilent Technologies/DAKO) were reactive with the tumor cells. Consequently, the diagnosis of PEComa was made. No additional therapy was performed. As of June 2017, 60 months after the resection of the LN, there has been no evidence of recurrence. Since the resection, no new areas of leukoderma have appeared, and the existing ones have not grown in size.

Tumors with positive immunophenotypic reactivity to melanocytic markers, including S100 protein, HMB-45, Melan-A/MART-1, tyrosinase and microphthalmia transcription factor, represent melanoma, clear cell sarcoma (CCS) and PEComa. Most PEComa show a clinically benign course in contrast to the unfavorable courses of melanomas and CCS.

Recently, Ferenczi et al. reported that MUM1 immunostaining is useful for distinguishing PEComa from melanoma and CCS.3 When MUM1 immunostaining score was calculated by multiplying the staining intensity from no staining (score 0) to strong staining (score 3) by positive tumor cell percentage for each individual specimen, the average score was 6.8 in 8 PEComa, 35.9 in 11 CCS, 270.4 in 11 primary melanomas and 114.2 in 11 metastatic melanomas.3 The completely negative staining and a favorable outcome in this case indicate the diagnosis of PEComa.

Naveh et al. suggested that immunohistopathological examination shows no difference between melanoma-associated leukoderma and vitiligo vulgaris.4 Therefore, immunohistopathological findings of PEComa-associated leukoderma may be similar to those of vitiligo vulgaris. Heretofore, PEComa- or CCS-associated leukoderma has not been reported, even though both PEComa and CCS share immunoreactivity for melanocytic markers. The leukoderma may arise in association with the occurrence of PEComa within an LN, where tumor cells are in close contact with immunocompetent cells.

**CONFLICT OF INTEREST:** None declared.

Sachiko TAJIMA-KONDO, Yoshimasa NOBEYAMA, Hidemi NAKAGAWA
Department of Dermatology, The Jikei University School of Medicine, Tokyo, Japan
doi: 10.1111/1346-8138.14019

**REFERENCES**

1. Hornick JL, Fletcher CD. PEComa: what do we know so far? Histopathology 2006; 48: 75–82.
2. Bonetti F, Pea M, Martignoni G, Zamboni G. PEC and sugar. Am J Surg Pathol 1992; 16: 307–308.
3. Ferenczi K, Lastra RR, Farkas T et al. MUM-1 expression differentiates tumors in the PEComa family from clear cell sarcoma and melanoma. Int J Surg Pathol 2012; 20: 29–36.
4. Naveh HP, Rao UN, Butterfield LH. Melanoma-associated leukoderma - immunology in black and white? Pigment Cell Melanoma Res 2013; 26: 796–804.